Masses of the Nose, Nasal Cavity, and Nasopharynx in Children

Diana P. Rodriguez, MD
Emily S. Orscheln, MD
Bernadette L. Koch, MD

Abbreviations: ABC = aneurysmal bone cyst, FB = foreign body, FDG = fluorine 18 fluorodeoxyglucose, IH = infantile hemangioma, JNA = juvenile nasopharyngeal angiofibroma, NGH = neuroglial heterotopia, NLC = nasolabial cyst, NLD = nasolacrimal duct, NPC = nasopharyngeal carcinoma, PG = pyogenic granuloma, RMS = rhabdomyosarcoma

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From the Department of Radiology, Nationwide Children’s Hospital, 700 Children’s Dr, Columbus, OH 43205 (D.P.R.); and Department of Radiology, Cincinnati Children’s Hospital, Cincinnati, Ohio (E.S.O., B.L.K.). Presented as an education exhibit at the 2015 RSNA Annual Meeting. Received March 23, 2017; revision requested June 28 and received July 15; accepted July 27. For this journal-based SA-CME activity, the author B.L.K. has provided disclosures (see end of article); all other authors, the editor, and the reviewers have disclosed no relevant relationships. Address correspondence to D.P.R. (e-mail: Diana.Rodriguez@nationwidechildrens.org).

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SA-CME LEARNING OBJECTIVES
After completing this journal-based SA-CME activity, participants will be able to:

- Understand the embryologic development of the frontonasal region and recognize developmental nasal midline masses that result from anomalies of the anterior neuropore.
- List common inflammatory lesions of the nose, nasal cavity, and nasopharynx in children and describe the characteristic imaging findings.
- Describe the relevant neuroimaging findings that can be used to distinguish benign from malignant neoplasms of the nose, nasal cavity, and nasopharynx in children.

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Introduction

A wide range of masses develop in the nose, nasal cavity, and nasopharynx in children. These lesions may arise from the nasal ala or other structures of the nose, including the mucosa covering any surface of the nasal cavity, the cartilaginous or osseous portion of the nasal septum, the nasal turbinates, and the nasal bones. Lesions may also arise from the nasopharynx or adjacent structures and involve the nose by way of direct extension. The causes of nasal masses in children include congenital and developmental disorders such as congenital nasolacrimal duct mucocele, dermoid cyst, cephalocele, and nasal neuroglial heterotopia; inflammatory and infectious processes such as mucocele, polyp, and pyogenic granuloma; benign neoplasms such as infantile hemangioma and juvenile nasopharyngeal angiofibroma; malignant lesions such as rhabdomyosarcoma and nasopharyngeal carcinoma; and masses related to prior trauma such as septal hematoma. Although direct visualization, without imaging, is frequently sufficient to diagnose pediatric nasal conditions, in many cases imaging has a key role in the treatment of the affected child. Some of these lesions have characteristic computed tomography and/or magnetic resonance imaging findings, some of them are diagnosed on the basis of the location and imaging findings combined, and others demonstrate nonspecific imaging findings. However, imaging is important for better defining the total extent of the lesion and guiding the clinician in determining whether medical and/or surgical intervention is required. In this article, the authors review the imaging findings of the most common causes—and many of the not-so-common causes—of nasal masses encountered in the pediatric population.

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Selection of the imaging modality depends on various factors such as the patient’s age, the availability of some modalities, the location of the mass, and the need for sedation or anesthesia. CT and MR imaging are complementary examinations, and although MR imaging is now the preferred examination for evaluating many of these lesions, CT may be necessary to further assess bone changes. However, caution is always recommended when considering the use of CT in children owing to the potential increased risk of carcinogenesis from ionizing radiation. CT evaluation of most head and neck lesions is performed with the administration of contrast material, and images are best evaluated when they are reformatted to include three planes in both soft-tissue and bone algorithms. MR imaging sequences that are ideal for assessing nasal lesions include multiplanar thin-section (ie, high-resolution) T1-weighted imaging, T2-weighted imaging with fat saturation, contrast material–enhanced T1-weighted imaging with fat saturation, and diffusion-weighted imaging.

Nonneoplastic Disorders

Congenital Nasolacrimal Duct Mucocele

Congenital nasolacrimal duct (NLD) mucocele refers to cystic dilatation of the NLD that is caused by obstruction of the duct’s distal opening at the valve of Hasner (ie, plica lacrimalis). Tear fluid secreted by the lacrimal glands is drained by the NLD from the medial canthus to the inferior meatus of the nasal cavity. Distal obstruction of the NLD results in the classic triad of a dacrocystocele (ie, dilated nasolacrimal sac), which appears as a medial canthal mass; a dilated NLD with expansion of its osseous canal; and an intranasal mass (ie, mucocele) below the inferior turbinate, which is the inferior extension of the dilated duct (1). On the other hand, proximal NLD obstruction results in a dilated nasolacrimal sac (classic dacrocystocele) owing to failure of the canalization that occurs during the 6th month of fetal life.

Obstruction of the NLD manifests at birth or shortly after birth as a firm bluish medial canthus mass (dacrocystocele) with excessive lacrimation and/or discharge. Nasal mucoceles can be large enough to obstruct the nasal cavity. Neonates with bilateral lesions (in 10% of cases) can present with severe respiratory distress, especially during feedings. A similar clinical presentation is seen with bilateral choanal atresia and pyriform aperture stenosis (2). The inability to pass a nasoenteric tube raises suspicion for these conditions. Other lesions that cause obstruction of the nasal passages in neonates include cephalocele, dermoid cyst, and nasal neuroglial heterotopia (NGH) (Table). A secondary infection—namely, dacrocystitis—may occur as a complication.

Imaging evaluation can be performed with CT or MR imaging. However, nonenhanced CT is the preferred imaging modality, as it depicts the soft-tissue and bone detail of the NLD mucocele triad. A dacrocystocele appears as a unilateral or bilateral low-attenuating, well-circumscribed round cystic lesion with a thin rim in the medial canthus. There is expansion of the bony NLD canal, with a low-attenuating dilated duct, and a well-circumscribed intranasal cyst. Large cysts can cause nasal septal deviation. An NLD mucocele can be detected prenatally with ultrasonography (US) or MR imaging (3) (Fig 1). At MR imaging, it appears as a well-defined tubular fluid structure extending from the medial canthus into the inferior nasal cavity, with a hyperintense signal on T2-weighted images and a hypointense signal on T1-weighted images. In the setting of dacrocystitis, contrast enhancement may be useful for further assessment of complications such as periorbital cellulitis and abscess formation (Fig 2).

Noncomplicated congenital dacrocystoceles are treated conservatively with massage and manual decompression. For dacrocystitis, systemic antibiotic treatment and surgical intervention with use of a probe to decompress the sac are required. The surgical decompression procedure is most often performed in the operating room. In the presence of an intranasal mucocele, probing is often unsuccessful, and cyst marsupialization is frequently necessary (2).
## Nonneoplastic and Neoplastic Masses of the Nose, Nasal Cavity, and Nasopharynx in Children

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Note.—ABC = aneurysmal bone cyst, FB = foreign body, IH = infantile hemangioma, JNA = juvenile nasopharyngeal angiofibroma, NLC = nasolabial cyst, NPC = nasopharyngeal carcinoma, PG = pyogenic granuloma, RMS = rhabdomyosarcoma.

**Figure 1.** Congenital NLD mucoceles in a fetus at 32 weeks gestation, who had bilateral cysts in the region of the lacrimal sacs at prenatal US (not shown). Coronal T2-weighted fetal MR image shows bilateral tubular hyperintense NLDs (arrows) extending into the inferior meatus.

### Congenital and Developmental Nasal Midline Masses

Developmental nasal midline masses result from neural tube closure anomalies of the anterior neuropore (ie, primitive frontonasal region), which forms and closes between the 3rd and 4th weeks of gestation. A complex sequence of events dictated by molecular changes coordinates and controls the morphologic development of the craniofacial structures in this region between the 3rd and 10th weeks of gestation (4).
The nose forms from the frontonasal processes and two nasal placodes that develop dorsal to the stomodeum in the 4th week of gestation (4,5). The nose originates from ectoderm, mesoderm, and a deep cartilaginous capsule. Neural crests form the sphenoid and ethmoid bones and the posterior nasal septum. Mesenchymal structures form the skull base and nose and then fuse and ossify. During this process, at 8 weeks gestation, a temporary fontanelle (fonticulus nasofrontalis) forms between the nasal bones and inferior frontal bones, and then later fuses to form the frontonasal suture (Fig 3a). Another transient space, the prenasal space, is located between the posterior aspect of the frontal and nasal bones and the anterior aspect of the nasal cartilage (Fig 3a). This space extends from the foramen cecum, a midline opening just anterior to the crista galli of the ethmoid bone, to the osteocartilaginous junction. A dural projection extends through the foramen cecum and comes in close contact with the subcutaneous region (Fig 3b). This projection then involutes and obliterates as the nasal process of the frontal bone grows (Fig 3c). Abnormal regression of these structures may result in midline developmental anomalies such as nasal dermoid cysts, cephaloceles, and nasal NGH (6).

**Nasal Dermal Sinus Cyst, Tract, or Fistula.**—A nasal dermoid is an anomaly of the anterior neuropore that is believed to result from failed involution of the neuroectoderm within the prenasal space, as just described. This congenital anomaly is rare, with an incidence ranging from one in 20000 to one in 40000 births. A nasal dermoid may manifest as a tract or fistula, with or without dermoid or epidermoid cysts. Dermoids are lined with stratified squamous epithelium and may contain adnexal structures such as hair or sebaceous glands (7).

The clinical manifestation includes a mass at the tip of the nose, nasal dorsum, or glabella. A sinus may be present with a sebaceous discharge and/or hair. Dermoid cysts may be superficial or intranasal. Intracranial extension cannot be determined clinically. Nasal dermoid infection may result in complications such as meningitis, cavernous sinus thrombosis, and periorbital cellulitis (8).

CT and MR imaging are useful and have complementary roles in the evaluation of nasal dermoids. CT images depict the bone detail and can show a large (>3 mm in diameter) foramen cecum and/or a bifid or deformed crista galli in the case of intracranial extension. A dermoid cyst, which has the density of fat, or an epidermoid cyst, which has the density of water, can be found along the dermal tract, from the tip of the nose or nasal cavity to the foramen cecum. CT and MR imaging are performed with contrast material, which aids in delineating the cyst and sinus tract and evaluating for possible infection.

High-resolution MR imaging aids in identifying an enhancing dermal sinus tract and enables better evaluation of possible intracranial extension (Fig 4a). Dermoid cysts manifest as well-circumscribed round lesions that are bright on T2-weighted MR images and of variable signal intensity on T1-weighted MR images, depending on the cyst contents. There is minimal rim enhancement at contrast-enhanced MR imaging. Diffusion-weighted MR images show epidermoid cysts—and sometimes dermoid cysts—that have high signal intensity, with corresponding low signal intensity on apparent diffusion coefficient maps (Fig 4b, 4c). To decrease the artifact from

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**Figure 2.** Acute dacrocystitis in a 3-week-old male neonate with congenital NLD mucoceles who presented with left eye swelling and redness. (a) Axial contrast-enhanced CT image shows enlarged lacrimal sacs (arrows) bilaterally, with surrounding soft-tissue stranding, which is greater on the left side. (b) Axial contrast-enhanced bone window CT image obtained at the level of the inferior meatus shows nearly complete nasal obstruction by the intranasal component of the mucoceles (arrows).
adjacent structures, non–echo-planar diffusion-weighted techniques are preferred (8).

Treatment consists of complete excision of the cyst and sinus tract to avoid recurrence and infection. The surgical approach varies according to the location of the cyst and tract, as well as the presence or absence of intracranial extension. Treatment techniques include direct excision, minimally invasive techniques with endoscopic resection, a rhinoplasty approach, and a transcranial approach (7,8).

**Anterior Cephalocele.**—The term cephalocele refers to the herniation of intracranial contents through a skull defect, which may involve the meninges and cerebrospinal fluid (meningocele) or the meninges, cerebrospinal fluid, and brain (encephalocele and meningoencephalocele). Cephaloceles result from failed neural tube closure and are classified on the basis of their location. Anterior cephaloceles include basal, frontoethmoidal, and nasopharyngeal types and are more frequent in the Asian population. Posterior cephaloceles are more frequent in the western hemisphere.

Basal cephaloceles (10% of cases) result from failed ossification, with extension of neural crest cells through the osseous defect, and can contain pituitary tissue, optic nerves, or vascular (arterial and venous) structures. Examples of midline basal cephaloceles include transethmoidal, sphenoethmoidal, and sphenopharyngeal types.
Nasopharyngeal cephaloceles are uncommon and occult and can extend through the ethmoid bone, sphenoid bone, or basiocciput into the nasal cavity or nasopharynx (Fig 5) (9).

Sincipital, or frontoethmoidal, cephaloceles are anterior herniations that manifest with an external mass and result from failed detachment of the cutaneous ectoderm and neuroectoderm of the anterior neuropore in the 3rd week of fetal life. There are three types of frontoethmoidal cephaloceles: frontonasal cephaloceles (40%–60% of cases), which protrude through the fonticulus nasofrontalis into the glabella (Fig 6); nasoethmoidal cephaloceles (30% of cases), which protrude through the foramen cecum into the nasal cavity; and naso-orbital cephaloceles, which protrude into the orbit through the lacrimal bone (9).

Although most cephaloceles are apparent at clinical examination, occult nasopharyngeal cephaloceles may manifest with airway obstruction (secondary to nasal or nasopharyngeal obstruction), cerebrospinal fluid leakage, or meningitis. Other congenital midline abnormalities are frequently seen in association with cephaloceles and include callosal dysgenesis, optic nerve hypoplasia, midline facial anomalies, cleft lip and palate, and hypothalamic-pituitary axis dysfunction.

MR imaging is the imaging modality of choice for evaluating cephaloceles. MR images show the extension of the malformation, the cephalocele contents, and the associated intracranial congenital anomalies. MR venography and MR arteriography help to identify arteries, veins, and dural venous sinuses in the cephalocele. CT is
helpful in demonstrating the osseous anatomy and defects for preoperative planning. However, the radiologist should be aware that portions of the anterior cranial fossa are not yet ossified in children younger than 1 year and should not be mistaken for bone defects. Current preferred treatment methods include a transnasal endoscopic approach and/or endoscopic transcra-niotomy approach (9).

**Nasal NGH.**—Nasal NGH, an anterior neuro-pore anomaly, refers to sequestered dysplastic, nonneoplastic neurogenic brain tissue that is not connected to the subarachnoid space. This tissue consists of astrocytes and neuroglial fibers with fibrovascular connective tissue stroma.

As with other anomalies of the anterior neuropore, NGH results from the extension of ectodermal tissue through the foramen cecum into openings in the nasofrontal region. This tissue is sequestered when the sutures close and appears as a soft-tissue mass that is most often midline, intranasal (30% of cases) or extranasal at the nasal dorsum (60% of cases), or both intra- and extranasal. Other sites where NGH may be seen include the pharynx, ethmoid bone, and palate.

MR imaging facilitates excellent soft-tissue characterization and has become the preferred imaging modality for evaluating NGH. Most often no intracranial connection with the mass is seen; however, a fibrous stalk is present occasionally. Recommended MR imaging examinations include high-resolution (ie, thin-section) multiplanar sequences such as T2-weighted imaging with fat saturation, T1-weighted imaging, non–echo-planar diffusion-weighted imaging, and contrast-enhanced multiplanar T1-weighted imaging with fat saturation. However, sedation or anesthesia is frequently required and can be a limiting factor.

NGH usually appears as a nonenhancing mass that is iso- to hyperintense to the brain parenchyma on T2-weighted MR images, without a corresponding decrease in diffusivity (ie, restricted diffusion). These findings distinguish NGH from dermoid and epidermoid cysts, which frequently demonstrate restricted diffusion. Although the use of CT in children has decreased owing to the potential increased risk of carcinogenesis from ionizing radiation, CT is frequently used in combination with MR imaging to assess anterior skull base lesions, with better demonstration of the adjacent osseous structures preoperatively. On CT images, NGH appears as a large, well-defined, nonenhancing soft-tissue mass that is intranasal or over the nasal dorsum (Fig 7). Treatment consists of complete surgical resection (10).

**Developmental Infectious and Inflammatory Disorders**

**Nasolabial Cyst.**—NLCs are developmental, nonodontogenic cysts that manifest in the nasolabial fold along the anterior maxilla. Histologically, these cysts are lined with different types of epithelium, including respiratory, pseudostratified ciliated, stratified squamous, and cuboidal epithelia, with goblet cells (11). The cause of NLCs is still unclear, and two theories have been proposed. One theory suggests that the cyst is derived from epithelial cells that are retained in the mesenchyme after fusion of the medial and lateral nasal processes during the 4th week of fetal life. The second theory suggests that NLCs arise from remnants of embryonic NLD tissue. NLCs manifest in individuals at any age, with a female-to-male ratio of 4:1. Approximately 90% of these cysts are unilateral, and they are more common on the left side (11).

The clinical presentation includes nasal obstruction, nasolabial swelling, and/or pain if there is superimposed infection. However, NLCs can be asymptomatic and found incidentally at imaging performed for other reasons. At physical examination, they can be fluctuant or mobile.

CT is the imaging technique most often used to evaluate NLCs. It should be performed after intravenous administration of contrast material, which enables better delineation of the mass lesion, identification of secondary changes in the adjacent bone, and demonstration of surrounding inflammatory changes. At CT, NLCs appear as well-circumscribed round cystic lesions with minimal rim
enhancement, in the submucosa of the anterior nasal floor, between the upper lip and the nasal aperture (Fig 8). Mucoid or serous cyst contents show fluid attenuation or slight hyperattenuation at CT (12). NLCs are extraskeletal lesions, but long-standing cysts may cause bone remodeling or erosion of the anterior maxilla (11).

An NLC may appear as a simple cyst at MR imaging, with a hypointense signal on T1-weighted images, a hyperintense signal on T2-weighted images, and minimal, if any, thin-rim enhancement. However, because the signal intensity of these cysts varies, depending on the presence of proteinaceous contents, they may appear hyperintense on T1-weighted MR images and hypointense on T2-weighted MR images (11,13). When infected, NLCs may show thick rim enhancement and perilesional inflammatory changes.

The preferred treatment for NLCs is intraoral sublabial surgical excision; however, endoscopic transnasal marsupialization is another approach used by some surgeons. A similar low recurrence rate has been reported with these two techniques. Other treatment options include aspiration with enucleation, and cryotherapy-assisted sublabial excision (11,12).

**Tornwaldt Cyst.**—Tornwaldt cysts are common nasopharyngeal cystic lesions lined with respiratory epithelium. They are believed to develop
secondary to occlusion of the opening of the pharyngeal bursa. The pharyngeal bursa represents an embryonic communication between the anterior tip of the notochord and the pharyngeal roof, which usually resolves after the 6th week of fetal life. Tornwaldt cysts rarely occur in the 1st decade of life and most likely develop secondary to inflammation or trauma (14). Incidental detection of Tornwaldt cysts at imaging has been reported in approximately 6% of cases (14,15). Location has a key role in recognizing a Tornwaldt cyst, as this cyst develops in the midline posterior wall of the nasopharynx, between the longus capitis muscles, without lymphoid tissue interposed between them. Although patients with large infected Tornwaldt cysts are mainly asymptomatic, they can present with persistent nasopharyngeal drainage, halitosis, and fluid-filled middle ear cavities (due to eustachian tube obstruction) (14).

On CT and MR images, Tornwaldt cysts appear as well-defined round or oval fluid-filled lesions with a thin rim (14). At MR imaging, there is typically a hypointense signal on T1-weighted images and a hyperintense signal on T2-weighted images (Fig 9). Fluid-fluid levels within the cyst may be present owing to layering proteinaceous or hemorrhagic contents and can exhibit a hyperintense signal on T1-weighted MR images, a hypointense signal on T2-weighted MR images, and isoattenuation relative to muscle on CT images. No enhancement is expected at contrast-enhanced imaging unless infection is present (14,15).

Treatment is not required for asymptomatic cysts, but it is indicated in symptomatic cases. Transoral excision or marsupialization of the cyst is the surgical technique of choice.

Adenoid Mucous Retention Cyst.—Adenoid mucous retention cysts may develop when inflammation leads to obstruction and subsequent cystic dilatation of the adenoid crypts. These cysts are lined with granulation tissue and inflammatory cells and contain lymphoid tissue and mucus, in contrast to Tornwaldt cysts, which have an epithelial lining (15). Adenoid mucous retention cysts are typically located eccentrically within the adenoid tissue in the posterior nasopharynx (14,15). These are small round or oblong cysts that range in size from a few millimeters to 20 mm. Adenoid cysts are usually asymptomatic and are found incidentally in approximately 6%–10% of patients (14,15). Affected patients do not require treatment.

At CT and MR imaging, adenoid cysts exhibit typical simple cyst characteristics and a thin nonenhancing rim. At MR imaging, these cysts typically exhibit a hypointense signal at T1-weighted imaging and a hyperintense signal at T2-weighted imaging, with no contrast enhancement (14) (Fig 10).

Mucocele of the Paranasal Sinuses or Concha Bullosa.—Mucoceles are benign cystlike masses lined with respiratory epithelium that develop secondary to obstruction of the ostium of a paranasal sinus. The chronic accumulation of mucus results in expansion of the sinus, with bone remodeling and sometimes erosion (16). In adults, mucoceles are often secondary to inflammation, trauma, or surgery. In children, mucoceles are rare and commonly manifest in association with cystic fibrosis. While mucoceles are primarily a condition of the paranasal sinuses, they can result in nasal obstruction secondary
to sinus expansion and are most common in the frontal sinus, followed by the ethmoid sinuses (16). The rare occurrence of mucocles in the nasal turbinates or nasal septum also can cause nasal obstruction (17). The clinical presentation depends on the location of the mucocele and its mass effect on adjacent structures. Complications include intracranial and intraorbital extension, which more frequently involves the frontal sinus, and occasionally infection (mucopyocele). Clinical symptoms include headache, maxillofacial pressure, nasal congestion, increased nasal drainage, and cranial nerve impingement (16).

At imaging, a mucocele appears as an expanded opacified sinus or turbinate (18) (Fig 11). The internal attenuation of a mucocele at CT is variable and depends on the nature of its contents; inspissated mucus may show hyperattenuation. Likewise, the signal intensity of these contents is highly variable at MR imaging: if there is uniform watery content within the mucocele, the lesion will be hypointense on T1-weighted images and hyperintense on T2-weighted images. However, as the mucocele contents become dehydrated or if there is internal hemorrhagic or proteinaceous material, the signal may be hyperintense on T1-weighted images and hypointense on T2-weighted images. Contrast-enhanced MR images may show mild rim enhancement but no internal enhancement.

The preferred surgical treatment is endoscopic marsupialization. This technique allows recovery of normal sinus function and mucociliary clearance (16).

**Nasal and Antrochoanal Polyps and Sinonasal Polyposis.**—Polyps consist of thickened redundant mucosal protrusions in the paranasal sinuses or nasal cavity, and they are believed to result from chronic inflammation. Polyps can be seen in association with allergic or atop respiratory epithelia (19,20). The most commonly encountered symptoms include nasal obstruction, rhinorrhea, facial pain, and a chronic cough in children (19).

Polyps can manifest as sinonasal polyposis, a solitary nasal polyp, or antrochoanal polyps (21) and can be differentiated by their imaging characteristics and location. In the setting of allergic rhinosinusitis, sinonasal polyposis consists of multiple polypoid masses that expand and completely fill the sinonasal cavities. Polyposis in the ethmoid cells may cause bulging of the lamina papyracea into the orbits, as well as expansion and thinning of the ethmoid septa and/or nasal septum, with or without bone dehiscence (Fig 12a). At MR imaging, polyps generally demonstrate a hyperintense signal on T2-weighted images and a hypointense signal on T1-weighted images owing to their high water content, with thin peripheral contrast enhancement (Fig 12b). Allergic sinusitis is seen in association with asthma in one-third of patients with polyps. Individuals who have polyps in association with allergic asthma, an aspirin allergy, and aggressive polyposis—known as the Samter triad—are considered a subset of persons who have chronic sinusitis with polyps. A number of genes have been identified in patients with this association. Complications with allergic polyposis are rare but include intracranial extension of disease.

In patients with cystic fibrosis, the prevalence of nasal polyps ranges between 6% and 48% and can be as high as 86% in patients who undergo functional endoscopic sinus surgery. New genes that may help to explain the development of nasal polyps in the setting of cystic fibrosis are under investigation (22,23). Cystic fibrosis is a recessive disease caused by mutations of the cystic fibrosis transmembrane conductance regulator gene, which acts as a cAMP (cyclic adenosine 3',5'-monophosphate)-regulated chloride channel. This genetic mutation results
in high concentrations of chlorine in secretions, increased mucus thickness, and impaired mucociliary clearance, predisposing affected persons to inflammation and chronic infection of the respiratory tract, including the sinonasal cavities. Bilateral nasal polyps with nasal lateral wall bulging are a frequent endoscopic finding. *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *Streptococcus viridans* are the most common organisms that colonize the sinonasal cavities in persons with cystic fibrosis.

A solitary polyp in the nasal cavity is rare and is confined to the nasal cavity alone. This polyp can arise from any location, such as the septum, turbinate, or cribiform plate. An antrochoanal polyp, a type of solitary polyp, arises from the maxillary sinus, fills the sinus, and prolapses through the maxillary ostium to result in its characteristic dumbbell shape (24). As this polyp grows, it extends into the nasal cavity and posteriorly to the choana (Fig 13)—hence, the term *antrochoanal polyp*. Because of its large stalk, an antrochoanal polyp is susceptible to complications such as torsion, strangulation, and even autoamputation and expulsion (24). A solitary polyp also can be seen in association with allergic processes.

**Nasal PG.**—PG, also referred to as lobular capillary hemangioma, is a benign capillary proliferation that can arise from the skin or the mucosa of the oral and nasal cavities. In the most recently revised classification of vascular anomalies, PG is categorized as a benign vascular tumor (25). Therefore, *pyogenic granuloma*—not *lobular capillary hemangioma*—is the preferred term.

The term *pyogenic granuloma* was coined in the early 1900s, when this lesion was believed to represent nonspecific granulation tissue that developed in response to the presence of a pyogenic agent. In 1980, Mills et al (26) introduced the term *lobular capillary hemangioma* on the basis of this lesion’s histologic appearance and described the typical lobular arrangement of the capillaries: usually localized to the deep portion of the lesion and surrounded by dense fibrous stroma with infiltration of inflammatory cells. More recently, in a study involving nasal PGs, Puxeddu et al (27) described two distinct histologic areas: a lobular area consisting of capillary proliferation with lobular architecture, and an ulcerative area consisting of inflammatory granulation tissue beneath an ulcer, with neutrophilic infiltrates and irregular dilated blood vessels.

The cause of PG is still unclear, and many etiologic factors have been suggested. Trauma and hormonal changes that occur during pregnancy or

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**Figure 12.** Sinonasal polyposis in an 11-year-old boy with sinus disease, headaches, and asthma. (a) Coronal contrast-enhanced bone window CT image shows complete paranasal sinus opacification and expansion (arrow), with nearly complete opacification of the nasal cavity. (b) Axial T2-weighted MR image shows pansinus mucosal thickening (arrows) with high signal intensity.

**Figure 13.** Antrochoanal polyp in a 6-year-old girl with a history of snoring. Axial nonenhanced CT image shows complete opacification of the right maxillary sinus. A soft-tissue mass (arrows) extends from the right maxillary antrum to the right nasal cavity and posteriorly to the nasopharynx and oropharynx (not shown). There was also expansion of the medial maxillary ostium (not shown).
with the use of hormonal contraceptives are believed to have a role in the development of this entity. However, it often manifests de novo, without a clear cause, and it occurs in both female and male persons. PG during pregnancy, termed *pyogenic granuloma gravidarum*, has been shown to regress spontaneously during the postpartum stage. It can occur in individuals at any age and has a slight predilection for males in the pediatric population and females in the adult population (28).

PG in the nasal cavity is rare but arises most commonly from the anterior nasal septum, followed by the turbinates and roof of the nasal cavity. The clinical presentation typically includes recurrent epistaxis, nasal obstruction, rhinorrhea, and pain (27,29). At endoscopy, nasal PG appears as a red or purple polypoid mass that bleeds easily when touched. It can manifest with or without superficial ulceration (27,28). In children, the differential diagnosis of nasal PG includes other masses such as dermoid cysts, NGH, meningocele and encephalocele, angiomaticus polyp, and JNA (30).

On nonenhanced CT images, nasal PG appears as a well-circumscribed, hypodense mass in a characteristic location, without intrinsic calcification. It can cause bone remodeling. The average size of nasal PGs is 1–2 cm, but lesions as large as 8 cm have been reported. On contrast-enhanced CT images, there is usually intense diffuse enhancement. However, the pattern of enhancement varies and includes diffuse homogeneous enhancement, somewhat inhomogeneous enhancement with spotty or linear areas of relative hypoattenuation, and central enhancement with a hypoattenuating rim (31) (Fig 14).

At MR imaging, nasal PG appears homogeneously isointense to the cerebral gray matter on T1-weighted images and heterogeneously hyperintense on T2-weighted images, with a hypointense rim. Yang et al (29) described the MR imaging appearance as a lesion with marked contrast enhancement and a nonenhancing rim. In some patients, they observed flow voids and intralesional foci of hemorrhage that appeared hyperintense on T1-weighted MR images.

The preferred treatment for nasal PG is surgical excision. However, various methods such as electrocoagulation, cryotherapy, laser therapy, excisional surgery, and (rarely) excisional surgery preceded by endovascular embolization have been used. The recurrence of these lesions varies among reported series, from 0% to 42% (32).

**Nasal Septal Hematoma.**—Hematoma of the nasal septum results from the rupture of small blood vessels in the nasal septum secondary to trauma and develops between the bony or cartilaginous septum and the respective overlying mucoperichondrium and mucoperiosteum. The nasal septum receives its blood supply from the branches of the internal and external carotid arteries. A rich vascular network arising from the overlying mucosal membrane traverses the mucoperichondrium through vascular channels located at the condromaxillary junction. Therefore, disruption or separation of the cartilage from its mucoperichondrium impairs the blood supply and may result in ischemia and necrosis of the cartilage (33).

Nasal septal hematoma may result from minor or major trauma with or without nasal fractures. A thorough nasal examination with anterior rhinoscopy is mandatory to exclude septal hematoma. There is a strong male predominance, and early diagnosis and treatment consisting of incision and drainage are critical to avoid complications such as abscess formation, septal necrosis and perforation, and “saddle-nose” deformity (34).

On CT images, a septal hematoma most often appears as an iso- to hypoattenuating collection centered in the nasal septum, but it may also be heterogeneous (Fig 15). CT is also useful for depicting associated fractures.

**Nasal Septal Abscess.**—Nasal septal abscess typically results from trauma with subsequent infection of a nasal septal hematoma. The infection can occur within 3 days of the trauma and may result in ischemia, avascular necrosis, and/or resorption of the septal cartilage. Less commonly, nasal septal abscess can occur in the setting of sinonasal or dental infection or in immunocompromised patients. In neonates, an infected underlying mass...
such as a dermoid or epidermoid cyst may be the cause of the abscess.

The most common causative organism is *S. aureus*; however, other common aerobic bacteria such as *Haemophilus influenzae* and group A β-hemolytic streptococci, and *Streptococcus pneumoniae* also can be involved. Bacteria produce collagenases that may contribute to the necrosis and liquefaction of the cartilage (33).

The most common symptom at presentation is bilateral nasal obstruction, although patients may also present with pain, fever, and/or headache 5–7 days after the injury. If a nasal septal abscess is not recognized and treated in a timely fashion, potential complications such as necrosis of the nasal septal cartilage with a resultant saddle-nose deformity may occur. In addition, hematogenous or direct spread can result in orbital cellulitis, intracranial abscess, meningitis, and/or cavernous sinus thrombosis.

On contrast-enhanced CT images, a nasal septal abscess appears as a peripherally enhancing fluid collection centered within the nasal septum (Fig 16). Adjacent inflammatory change or gas within the fluid collection also may be present. CT is useful for the detection of underlying sinus or dental infection and facial fractures, as well as orbital, intracranial, and vascular complications. At MR imaging, a nasal septal abscess typically demonstrates a hyperintense signal on T2-weighted images and a hypointense signal on T1-weighted images, with characteristic diffusion restriction. However, the signal may be altered in the presence of an underlying hematoma or septal mass.

Treatment includes drainage of the abscess to release the pressure and reestablish the blood supply to the septum, and debridement of the infected cartilage. In children, treatment is also directed at preventing the loss of septal cartilage, for which adequate reconstructive surgical therapy is required to prevent functional and aesthetic problems of the nose and midface (33).

### Nondevelopmental and Noninflammatory Nasal Lesions

**Foreign Body.**—Young children commonly insert FBs into their nose; the highest incidence of these cases occurs in persons 3–5 years of age (35,36). In older children, nasal FBs are more frequently reported in those with special needs or a developmental delay (36). Caretakers frequently witness children introducing an object into the nose, or the incident is reported by the child. Less frequently, nasal FBs are found incidentally at diagnostic imaging or when complications occur.

Nasal FBs are most often located anterior to the middle turbinate, or anterior and beneath the inferior turbinate (36). In two recently published large series (35,36), the most common FBs removed were hard spherical or globular objects such as beads, toy fragments, dried vegetables, and seeds and nuts. Other frequently removed objects included foam, paper, and disk-shaped objects such as button batteries and magnets. Inorganic objects were encountered more frequently than were organic objects.
The signs and symptoms of a nasal FB include nasal obstruction, epistaxis, a foul odor from the nasal region, and halitosis (36). Purulent rhinorrhea occurs when infection develops. Severe injury and complications such as mucosal necrosis and septal perforation have been reported in association with inserted button batteries, with débridement in the operating room required (36). Pressure between two magnets also can result in nasal septal perforation. Therefore, the identification of batteries or magnets should prompt quick intervention.

The imaging characteristics depend on the FB material and the length of time it has been lodged. Occasionally, in chronic cases, a rhinolith may develop. Radiographs may be adequate for identifying radiopaque FBs (Fig 17). Chest and abdominal radiography should be considered if aspiration or ingestion of a nasal FB is suspected. CT can depict the soft-tissue attenuation of FBs such as plastic and organic material, including seeds, food, and sometimes wood. In the immediate setting, dry wood mainly contains air, which shows low attenuation at CT and can be easily mistaken for normal nasal cavity air on soft-tissue window images. Therefore, wood is best detected at CT by using wider window settings, such as bone window and lung window, and appears as a linear or cylindrical focus of slightly higher attenuation than air surrounded by low-attenuating inflammatory soft tissue. A window width of 3000 and window level of 500 are recommended for detection of intranasal wooden FBs. In the chronic stage, wood and other FBs can develop calcification and/or a soft-tissue encasement, and rarely can they cause osseous erosion. Although MR imaging is rarely necessary, it is indicated for the evaluation of intracranial complications.

Nasal FBs are more often removed in the emergency department by either emergency department staff or otolaryngologists, depending on their level of expertise and the complexity of the case. Investigators from a single pediatric hospital recently reported a high rate of otolaryngology involvement in the removal of FBs (35). In their experience, approximately 59% of patients were referred to the otolaryngology clinic and required operating room intervention; these cases more frequently involved disk-shaped FBs and children older than 5 years (35).

Benign Neoplasms and Neoplasm-like Lesions

Hemangioma

Hemangiomas are benign vascular tumors, and two types occur early in life: IH, which is also known as hemangioma of infancy, and congenital hemangioma. According to the 2014 revised classification by the International Society for the Study of Vascular Anomalies, which was recently updated to achieve a uniform classification system, these two lesion types are distinctly different (25).

IH is the most common tumor of infancy, with an incidence of 4%–10%. It is more frequent in females and manifests predominantly in the face and neck (37). IH manifests during the 1st weeks of life and first undergoes a proliferating phase characterized by rapid growth over months. It then progressively regresses during a spontaneous involution phase. The subclasses of IH include multifocal, segmental, and indeterminate types, which are based on the extent and distribution of the lesion. Segmental IHs are
large, usually greater than 5 cm in diameter, and seen in association with other anomalies such as PHACE syndrome. The original acronym, **PHACE**, refers to posterior fossa anomalies, hemangioma, arterial lesions, cardiac abnormalities and/or aortic coarctation, and abnormalities of the eye (38). Anomalies of ventral development, such as partial or complete agenesis of the sternum or sternal cleft, also are seen with PHACE syndrome (38) and account for the acronym **PHACES** that is used by some authors. Histologically, IH consists of proliferating endothelial cells and pericytes that have unique immunohistochemical markers, such as glucose transporter 1 (25). There are frequently multiple IHs, and their clinical appearance can be variable. They may manifest as a typical strawberry-red skin lesion, which is often the tip of the iceberg of an extensive and deep lesion. On the other hand, segmental hemangiomas may have a telangiectatic appearance or appear as only a faint erythematous area.

Compared with IHs, congenital hemangiomas are much less common, and they are present and usually fully developed at birth, after having completed the proliferating phase in utero. There are three types of congenital hemangiomas: rapidly involuting, noninvoluting, and partially involuting. Rapidly involuting lesions usually involute quickly during the first year of life. Congenital hemangiomas consist of capillary lobules and can be associated with large veins, arteries, and lymphatics. Unlike IHs, congenital hemangiomas lack expression of the glucose transporter 1 immunohistochemical marker (25).

At ultrasonography (US), CT, and MR imaging, IH appears as a well-circumscribed, lobulated hypervascular soft-tissue mass. Color Doppler US images obtained during the proliferating phase typically show high vessel density (more than five vessels per square centimeter) and a high systolic Doppler shift (>2 kHz), with low resistive arterial tracings but without arterIALIZED veins to suggest an arteriovenous shunt. During the involutional phase, there is a progressive decrease in vessel density and an increase in resistive indexes. IH has soft-tissue attenuation at nonenhanced CT, is isointense to muscle at T1-weighted MR imaging, and is hyperintense at T2-weighted MR imaging (Fig 18a). IHs show diffuse intense homogeneous contrast enhancement at CT and MR imaging (Fig 18b), and intralesional flow voids, which are usually large and numerous in younger patients, can be seen on MR images. As the IH lesion involutes, there is a progressive decrease in its size, with an increased fibrofatty matrix that results in a more heterogeneous appearance, and decreased contrast enhancement. Congenital hemangiomas have a similar lobulated appearance but typically exhibit a heterogeneous and less well-defined appearance at all imaging examinations, and they may contain areas of calcification, hemorrhage, or necrosis.

The majority of IHs spontaneously involute and do not require treatment. However, when lesions compromise adjacent structures such as cranial nerves, airways, and/or intraorbital structures, or are large and compromise the overlying skin, the first-line treatment is propranolol (39) or other β-blocker therapy. Excision is reserved for ulcerated lesions, residual lesions, and large periorbital lesions that can affect vision and are nonresponsive to medication therapy.
Juvenile Nasopharyngeal Angiofibroma

JNA is a highly vascular benign neoplasm that exhibits local aggressive and invasive behavior and occurs in adolescent boys. After extensive debate, it is now most commonly accepted that JNA originates at the sphenopalatine foramen in the posterolateral aspect of the nasal wall. The cause of JNA remains unclear; however, the Wnt signaling pathway and angiogenic and hormonal factors are believed to have a role in the pathophysiology of this neoplasm, especially in patients with associated familial adenomatous polyposis (40). JNA can spread locally in all directions from its origin, but it frequently extends laterally into the pterygopalatine fossa and posterior nasal cavity, or it can spread farther, into the infratemporal fossa by way of the pterygomaxillary fissure (41). It can also expand posteriorly and laterally to the pterygoid process and/or between its plates (42), or superiorly to the sphenoid sinus and orbit. Intra- or extradural intracranial invasion, and involvement of the cavernous sinus, sella, and chiasm can occur.

The typical clinical presentation of JNA includes nasal obstruction (90% of cases) and epistaxis (60% of cases), which is due to its highly vascular nature. Other clinical manifestations include hyponasal speech, proptosis, and cranial neuropathies, which can manifest with diplopia in advanced cases.

JNA has both fibroblastic and vascular components and appears on CT images as a soft-tissue mass with well-marginated lobulated contours in the characteristic location, with intense contrast enhancement. CT best depicts bone erosion and bowing of the adjacent paranasal sinus walls, orbits, and skull base (Fig 19a).

At MR imaging, the mass is relatively iso- or hypointense to muscle on T1-weighted images, is iso- to slightly hyperintense on T2-weighted images, with intrinsic areas of hypointensity owing to its fibrous components (Fig 19b), and has marked diffuse contrast enhancement (Fig 19c). Intralesional flow voids can often be seen. MR imaging is the imaging technique of choice for identifying intraorbital or intracranial extension and cavernous sinus and paranasal sinus involvement. It also aids in differentiating tumor involvement of the sinuses versus fluid retention in the sinus secondary to obstruction by the tumor (43).

Digital subtraction angiography has an important role in preoperative planning: it enables identification of the tumor’s vascular supply and thereby yields information regarding potential invasion.
routes for preoperative embolization (Fig 20a, 20b) (43). Such planning helps to reduce bleeding during surgical resection. Surgery should be performed within 48 hours after the embolization to avoid recanalization or neovascularization. Angiography depicts a tumor blush, which is usually arising from branches of the external carotid artery; however, recruitment of additional vessels may occur with tumor growth. Up to 30% of cases may involve an internal carotid artery blood supply, usually in the setting of intracranial extension. Biopsy is contraindicated in patients with this highly vascular tumor.

**Nasal Pleomorphic Adenoma**

Pleomorphic adenoma is the most common benign salivary gland tumor, and it mainly arises from the major salivary glands. Approximately 75% of these neoplasms are located in the parotid gland, 15% of these lesions are located in the submandibular gland, and less than 10% are located in the minor salivary glands of the palate (44). Rare locations include the nasal cavity, pharynx, larynx, trachea, and lacrimal glands (45).

Nasal pleomorphic adenoma is very rare and more common in young and middle-aged women; however, it also occurs in children. This tumor most frequently originates from the nasal septum; however, other sites include the lateral nasal wall, nasal turbinates, and nasopharynx (45). The histologic features of nasal pleomorphic adenoma are similar to those of major salivary gland pleomorphic adenoma. However, nasal pleomorphic adenoma characteristically demonstrates greater cellularity and a predominance of epithelial rather than stromal elements. This lesion is a well-circumscribed lobulated mass that usually has a thick fibrous capsule. It often remodels the adjacent bone but rarely causes bone destruction. The clinical presentation is typically painless unilateral nasal obstruction and occasionally epistaxis and/or nasal discharge (46).

At nonenhanced CT, nasal pleomorphic adenoma appears as a well-defined lobulated soft-tissue mass that is isoattenuating to muscle and often occupies the entire nasal cavity. It can cause displacement and remodeling of the nasal septum and occasionally bone destruction. Intralesional punctate calcifications also may be present, and contrast enhancement is diffuse and heterogeneous (Fig 21a) (46,47).

Nasal pleomorphic adenomas have heterogeneous low or intermediate signal intensity, compared with muscle, on T1-weighted MR images and intermediate to high signal intensity on T2-weighted MR images (46,47). A peripheral hypointense capsule may be seen on T2-weighted MR images. Contrast enhancement is usually heterogeneous, with occasional internal nonenhancing areas, which may represent cystic changes (46,47) (Fig 21b).

Treatment consists of surgical excision. Recurrence and malignant transformation are rare.

**Osseous Lesions**

Several benign neoplasms and neoplasm-like lesions can arise from the osseous structures adjacent to the nasal cavity. These lesions can appear as nasal masses, depending on their site of origin and growth pattern. Such lesions
include giant cell tumor, ABC, fibrous dysplasia, and ossifying fibroma.

**Giant Cell Tumor and Central Giant Cell Granuloma**

Giant cell tumor, a relatively common (20% of cases) benign bone tumor, most often arises from the epiphysis of long bones. However, approximately 2% of all giant cell tumors occur in the head and neck; these are known as central giant cell granulomas, and they predominantly affect the mandible. Less frequently, central giant cell granulomas involve the maxilla, with extension into the nasal cavity (48).

Jaffe first described giant cell tumor in 1953, calling it *giant cell reparative granuloma*. This term was abandoned, given that a giant cell tumor is considered to be a benign neoplasm rather than a reactive mass. Some authors suggest that giant cell tumor of the long bones and central giant cell granuloma are different entities. However, others consider both of these lesions to be part of a spectrum of the same disease based on their histologic characteristics. Giant cell tumors are composed of the fibroblastic stroma of spindle-shaped cells, with multinucleated giant cells throughout the stroma, and thin-walled capillaries. There are also areas of hemorrhage and hemosiderin deposition. Spindle cells rather than giant cells are considered the proliferating part of the tumor. Spindle cells are osteoblast-like cells that express proteins that promote osteoid and osteoclast formation (48). The cause of central giant cell granulomas is unclear, but they appear to occur in association with syndromes such as neurofibromatosis type 1, cherubism, and Noonan syndrome. Central giant cell granulomas manifest in individuals at any age but more frequently in those younger than 30 years, with a slight female predominance.

At imaging, central giant cell granuloma appears as a well-circumscribed soft-tissue mass, with bone remodeling and erosion. The mass is isoattenuating at nonenhanced CT and enhances diffusely at contrast-enhanced CT (Fig 22). On T2-weighted MR images, central giant cell granuloma can have a markedly hypointense signal in the presence of hemorrhage and/or hemosiderin, but it may be mildly hyperintense to muscle, with small cystic components occasionally seen. Contrast enhancement is diffuse and homogeneous (49).

Treatment consists mainly of surgical curettage, but other treatments such as the use of corticosteroid injections, calcitonin, and interferon-α have been suggested (48). The recurrence rate is higher with the aggressive subtypes of central giant cell granuloma.
Aneurysmal Bone Cyst

ABC is a benign osteolytic lesion that can affect any bone, but it most often arises in the metaphyses of long bones. These lesions in the jaws are rare, accounting for only 1%–2% of all ABCs, and affect the mandible more frequently than they affect the maxilla. The mandibular ramus is the most common site for craniofacial involvement. The World Health Organization considers ABCs to be benign cystic bone lesions that are composed of blood-filled spaces separated by connective tissue septa that contain fibroblasts, osteoclast-type giant cells, and reactive woven bone (50). Primary ABCs arise de novo, while the secondary form arises from other benign or malignant bone tumors. However, secondary ABCs in the jaws have been reported to arise only from benign tumors such as fibrous dysplasia, osteoblastoma, central giant cell granuloma, and cementifying fibroma. These cysts can manifest in individuals at any age (median age, 13 years) and have no sex predilection.

The clinical presentation varies according to the size, location, and destructive behavior of the cyst. An ABC in the maxilla can extend to the nasal cavity and orbit and cause nasal obstruction, epistaxis, or other symptoms such as diplopia, epiphora, strabismus, and exophthalmos. Jaw ABCs can cause tooth displacement and loosening (51).

On plain radiographs, ABCs appear as radio-lucent expansile lesions, which may be unilocular but are most often multiloculated with a soap-bubble appearance. CT and MR images show cortical thinning, internal thin septa, blood-filled spaces with fluid-fluid levels, tumor extension, and adjacent structure involvement (Fig 23a).

Bone changes are best assessed with CT, while fluid-fluid levels are typically more apparent on MR images (Fig 23b). Contrast-enhanced images may show mild septal enhancement. Although the described treatment for ABCs varies widely in the literature, curettage and resection are the most common procedures.

Fibro-osseous Lesions

Fibro-osseous lesions of the craniofacial region are benign lesions that replace normal bone and share common histologic characteristics, which consist of a combination of spindle cells (fibrous component) and a matrix of woven bone (osseous component). The proportions of these components vary with each lesion. Craniofacial fibro-osseous lesions include fibrous dysplasia, ossifying fibroma, and osseous dysplasia. Herein, we review fibrous dysplasia and ossifying fibroma. The final diagnoses of these two lesions are heavily based on their radiologic characteristics, as they have distinct imaging findings. Histologically, they can be very similar (52).

Fibrous Dysplasia.—Fibrous dysplasia is a fibro-osseous medullary lesion; it affects a single bone (monostotic) in 70% of cases and more than one bone (polyostotic) in 30% of cases. Polyostotic fibrous dysplasia manifests in association with skin hyperpigmentation and endocrine abnormalities in McCune-Albright syndrome. Fibrous dysplasia in the craniofacial bones accounts for 35% of monostotic fibrous dysplasias, even if it involves several adjacent bones. The skull base and facial bones are common sites of involvement, and in this setting, they may encroach on the nasal cavity. The jaws are the most frequently
affected bones. Fibrous dysplasia is more common in the 1st and 2nd decades of life and has a slight female predilection (53,54).

The clinical manifestations of fibrous dysplasia include painless swelling, facial deformity, exophthalmos, and symptoms related to narrowing of skull base foramina. Fibrous dysplasias that undergo periods of increased activity can cause increased swelling and discomfort and may mimic osteomyelitis on bone scans. The growth of this lesion tends to decrease after puberty. The diagnosis is confirmed by the presence of the mutation of GNAS 1α, which enables the differentiation of fibrous dysplasia from other fibro-osseous lesions (53).

The imaging characteristics of fibrous dysplasias depend on the stage of development of the disease and the amount of bone matrix, and vary from lucent to sclerotic lesions. At plain radiography and CT, this lesion has the characteristic ground-glass appearance, which is secondary to the amount of woven bone present. Fibrous dysplasia manifests as an expansile lesion that has ill-defined margins and a thinned cortex; it replaces normal bone but rarely involves bone erosion (Fig 24). CT is key to rendering an accurate diagnosis, given that fibrous dysplasias can exhibit aggressive characteristics at MR imaging, mimicking malignant lesions. At MR imaging, the appearance of fibrous dysplasia is variable, but it typically has low signal intensity on T1-weighted images. The signal intensity is variable on T2-weighted MR images: it can be low, intermediate, or high. At contrast-enhanced imaging, there is internal heterogeneous enhancement (54), with the fibrous component typically enhancing more than the osseous component. As a benign lesion, fibrous dysplasia has increased diffusivity on apparent diffusion coefficient maps, which can help in differentiating it from a malignant lesion (55).

The treatment of patients with fibrous dysplasia depends on their symptoms and is mainly conservative. Curettage, decompressive surgery, and rarely, total resection are reserved for cases involving compression of critical structures such as the optic nerve, or cases of substantial craniofacial involvement resulting in functional or aesthetic impairment. Biopsy is often the only procedure required (53).

**Ossifying Fibroma.**—Ossifying fibroma is a rare expansile fibro-osseous tumor that can arise from any facial bone, but it more frequently occurs in the mandible. The sinonasal cavity may be involved by lesions extending from either the maxilla, which is affected in 10%–20% of cases, or the paranasal sinuses—most commonly the ethmoid sinus. Ossifying fibroma most often appears in the 3rd and 4th decades of life and has a slight female predominance. However, juvenile ossifying fibroma affects young patients aged 5–10 years. In contrast to fibrous dysplasia, ossifying fibroma is a well-marginated expansile and destructive bone lesion. The juvenile form has been shown to be more aggressive, causes bone erosion, and has a greater rate of recurrence.

Histologically, ossifying fibroma consists of a fibrous stroma of fibroblasts and collagenous fibers, mixed with mineralized components of woven and lamellar bone (56). It usually exhibits a thin mineralized rim. In contrast, juvenile ossifying fibroma usually has a thick mineralized rim and is characterized by internal mineralized ossicles of various shapes, similar to cementum, called psammomatoid bodies.

The most common clinical symptoms of ossifying fibroma include nasal obstruction, proptosis, facial pain, and deformity. However, this tumor can be asymptomatic and found incidentally (57).

CT is an excellent imaging modality to assess ossifying fibroma, allowing characterization, delineation, and extension of this lesion. Ossifying fibroma manifests as a well-marginated round or oval mass with a thin or thick sclerotic rim and internal lucent areas mixed with mineralized foci of various shapes (Fig 25). The sharp margins of ossifying fibroma can help differentiate it from a
malignant lesion such as RMS or chondrosarcoma (57). However, juvenile ossifying fibroma tends to cause more osseous destruction and erosion and can adhere to soft tissues. At MR imaging, ossifying fibroma has low to intermediate signal intensity on T1-weighted images and variable, often heterogeneous signal intensity on T2-weighted images. The mineralized portions appear hypointense on T2-weighted MR images. This appearance may mimic that of fibrous dysplasia; however, ossifying fibroma tends to have higher signal intensity at T2-weighted MR imaging and can have cystic spaces with fluid-fluid levels. There can be contrast enhancement of the outer shell and septa (57).

Treatment consists of complete resection, which may be difficult when there is dural or periorbital adherence of the tumor. An endoscopic approach is preferred and is considered an effective and safe treatment approach (58).

Malignant Neoplasms

Rhabdomyosarcoma

RMS accounts for 3%–4% of all pediatric malignancies and is the most common solid tumor in the pediatric population. It affects the head and neck in approximately 35%–40% of cases. RMS has a bimodal distribution in children, occurring in those aged 2–4 years and 12–16 years. Approximately 60% of cases are seen in children younger than 6 years (59).

The two histologic variants of pediatric RMS are the embryonal and alveolar subtypes. The embryonal subtype is more common and is associated with an intermediate prognosis, while the prognosis is poor for children who have the alveolar type. Furthermore, the presence of the PAX3–FOXO1 fusion gene in alveolar RMS results in a more aggressive clinical course. The majority of RMS cases are sporadic; however, associations with genetic conditions such as Li Fraumeni syndrome and neurofibromatosis type 1 have been reported (59).

The disease stage, prognosis, and treatment options are determined on the basis of the para- meningeal or nonparameningeal locations of the tumor. Parameningeal tumors can invade the meninges and may manifest in the nasopharynx and nasal cavities, paranasal sinuses, infratemporal and pterygopalatine fossae, and/or middle ear cavity. Nonparameningeal tumors may manifest at all other sites except the orbit.

The clinical presentation is based on the tumor’s location. Nasopharyngeal RMS may be asymptomatic for months, or it can manifest with signs and symptoms that are similar to those of common benign diseases and, consequently, result in a delayed diagnosis (60). These signs and symptoms include nasal or sinus obstruction with or without nasal discharge, snoring, obstructive sleep apnea, and headache. At the time of diagnosis, nasopharyngeal RMS can be large with intracranial extension and can spread to nearby structures by way of hematogenous or lymphatic routes. Distant metastatic disease has been detected in up to 23% of cases at initial diagnosis (61).

Tumor severity is classified by using a system created by the Intergroup Rhabdomyosarcoma Study Group, which determines treatment proto-
cols. These protocols mainly consist of a combination of chemotherapy and local control with surgery and/or radiation therapy. Owing to the invasive nature of nasopharyngeal masses, most often only biopsy rather than resection can be performed (60,62).

CT is helpful in identifying osseous erosion, which may be present with RMS. Contrast-enhanced images show an invasive soft-tissue mass with variable enhancement and occasionally central areas of necrosis. MR imaging enables the best delineation of this soft-tissue tumor and the best assessment of intracranial extension, perineural spread, or involvement of other nearby structures. Dural, intraorbital, and bone marrow involvement is well demonstrated with MR imaging. RMS is isointense on T1-weighted MR images and has variable signal intensity—usually with an iso- to hypointense signal—on T2-weighted MR images, with variable contrast enhancement (Fig 26a). At diffusion-weighted MR imaging, there is reduced diffusivity (ie, restricted diffusion) secondary to a high nuclear-cytoplasmic tumor cell ratio (Fig 26b, 26c). Metastatic lymph nodes also exhibit decreased diffusivity. At fluorine 18 fluorodeoxyglucose (FDG) positron emission tomography (PET), RMS and metastatic lymph nodes appear as hypermetabolic masses with increased FDG uptake (Fig 26d) (60).

Leukemia and Lymphoma
Extranodal non-Hodgkin lymphoma of the head and neck is rare and even more so in children. This malignancy is most often a natural killer cell/T-cell lymphoma (NKTL), with a high prevalence in Southeast Asia and a male predominance. It affects the sinonasal cavity and Waldeyer tonsillar ring, and the tonsils more frequently than the nasopharynx. The nasal cavity is the primary affected site in about 10% of cases of sinonasal tract lymphoma (63).

The pathogenesis of extranodal non-Hodgkin lymphoma is still unclear; however, immunosuppressed children are at an especially high risk when they are exposed to carcinogens or oncogenic infections such as Epstein-Barr virus. Natural killer cells that undergo malignant transformation express CD56 markers and can be distinguished from T-cell lymphoma with use of immunophenotypic and molecular genetic tests only. Sinonasal NKTL is an aggressive tumor that is associated with a poor survival rate (63).
The clinical presentation varies according to histologic type. A low-grade tumor may manifest as an obstructing mass, while a high-grade tumor manifests as a nonhealing ulcer, with cranial nerve deficits, facial swelling, epistaxis, and pain. In children, some of these nonspecific signs and symptoms usually are manifestations of benign processes such as sinusitis, polyps, nasal septal deviation, or hypertrophied turbinates, which can delay the diagnosis. Therefore, a high index of suspicion and endoscopic examination are necessary to identify ulcerative changes that raise suspicion and prompt biopsy. The prognosis also varies according to the histologic type and stage of the disease, with a 5-year overall survival rate of 52%. The recommended treatment consists of a combination of radiation therapy and chemotherapy (64).

On CT images, nasal lymphoma appears as a soft-tissue mass in the nasal cavity that may extend into the adjacent paranasal sinuses, osseous structures, and/or subcutaneous soft tissues. Contrast enhancement is mild to moderate, and heterogeneous. Nonsclerotic bone destruction or remodeling is common. MR imaging is important for evaluation of dural involvement if there is cribiform plate erosion. It facilitates superior evaluation of the extent of soft-tissue involvement. Lymphoma exhibits an isointense signal relative to muscle on T1-weighted MR images, an intermediate to mildly hyperintense signal on T2-weighted MR images, and mild to moderate heterogeneous contrast enhancement (Fig 27). Granulocytic sarcoma, or chloroma, rarely occurs in the nasal cavity and is usually associated with acute myeloid leukemia in the setting of widespread disease. However, it may precede acute myeloid leukemia.

Nasopharyngeal lymphoma usually manifests as a large exophytic homogeneous mass that occupies most of the nasopharyngeal airway, and unlike NPC, it rarely invades deep adjacent structures. At MR imaging, nasopharyngeal lymphoma has increased contrast enhancement and lower apparent diffusion coefficient values compared with NPC. However, both of these cancers manifest with cervical lymphadenopathy, and they demonstrate similar radiopharmaceutical uptake at FDG PET (Fig 27b) (65).

### Nasopharyngeal Carcinoma

NPCs are epithelial tumors that arise from the nasopharyngeal mucosa, and they account for less than 1% of all childhood malignant tumors. NPC is a rare entity in the United States and Europe, with an incidence of 0.5 to two cases per 100,000 persons annually. However, NPC is endemic in Southeast Asia, where the incidence is 10–30 cases per 100,000 persons annually (66). In the 2005 World Health Organization classification system, NPC is divided into three histopathologic types: type 1, keratinizing squamous cell carcinoma, which is seen in older adults; type 2, nonkeratinizing differentiated carcinoma; and type 3, nonkeratinizing undifferentiated carcinoma. The most common NPC type in children is undifferentiated carcinoma, which mainly affects persons between the ages of 10 and 19 years. Undifferentiated NPC differs from RMS, which occurs more often in persons of a younger age. The cause of NPC has been closely linked to Epstein-Barr virus infection and genetic and environmental factors, including carcinogens such as nitrosamines (67).

![Figure 27. Nasopharyngeal lymphoma in a 10-year-old boy with a history of Epstein-Barr virus–positive T-cell lymphoma.](image-url)
The clinical presentation consists of nonspecific signs and symptoms, such as an upper neck painless mass, nasal obstruction, rhinorrhea, and/or fever of unknown cause. NPC may also manifest as otitis media and hearing loss secondary to eustachian tube obstruction. Invasion to the skull base results in cranial neuropathies, with symptoms that include ptosis, vision loss, diplopia, dysphagia, trismus, and hoarseness. Cervical lymphadenopathy is often the initial manifestation. In order of frequency, distant metastases to the bones, liver, bone marrow, lungs, and mediastinum can occur (67,68).

At imaging, NPC appears as an asymmetric mass centered at the lateral nasopharyngeal recess (ie, Rosenmüller fossa), with aggressive features such as bone erosion, intracranial extension, and metastatic cervical lymphadenopathy. It can invade the parapharyngeal space and extend to the pterygopalatine fossa, frequently widening the petroclival fissure (68). NPC may be indistinguishable from lymphoma or RMS at conventional imaging alone. The CT characteristics of NPC are a heterogeneously enhancing soft-tissue mass and associated bone erosion or sclerosis at the skull base (Fig 28a). Cervical adenopathy also is common at manifestation. On T1- and T2-weighted MR images, this lesion typically has an isointense signal (Fig 28b), with variable—usually heterogeneous—contrast enhancement (Fig 28c). MR imaging enables the best evaluation of
perineural and intracranial spread. The high cellularity of this tumor leads to decreased diffusivity on diffusion-weighted MR images. FDG PET/CT is useful for detection of distant metastases and monitoring posttreatment tumor recurrence. However, compared with MR imaging, FDG PET/CT has low sensitivity for determining tumor extension and regional lymphadenopathy at diagnosis (Fig 28d) (69,70).

NPC is generally considered to be nonreseetable, and the surgical approach is mainly limited to biopsy. Currently, the preferred treatment consists of a combined regimen of radiation therapy and chemotherapy. It has been demonstrated that in children, neoadjuvant chemotherapy can be used to achieve tumor reduction, improve local tumor control, and treat systemic micrometastases—thus, lowering radiation therapy doses (71). In addition to chemotherapy, interferon-β is used, as it appears to have antitumor effects in patients with Epstein-Barr virus–positive NPC (71,72).

**Metastatic Neuroblastoma**

Neuroblastoma is known for its widespread metastatic potential in children. It most often manifests as skeletal metastases; however, hepatic metastases are most common in infants. In the head and neck, neuroblastoma metastasizes to the skull base, calvarium, and orbit, but it rarely involves the facial bones and paranasal sinuses. Metastases are rare in the mandible and maxilla, and there have been only a few reported cases in the paranasal sinuses—in the maxillary and sphenoid sinuses specifically—with potential extension to the nasal cavity (73).

Orbital and skull base metastases can manifest with proptosis, Horner syndrome, and periocular soft-tissue hematomas (ie, black or raccoon eyes) that are caused by hemorrhage in osseous and/or soft-tissue lesions but can be mistaken for a sign of nonaccidental trauma. The clinical symptoms of facial metastatic disease include systemic illness, bone pain, an abnormal dentition eruption pattern, and teeth displacement (73).

Metastatic neuroblastoma appears as an expansile osteolytic lesion in association with a “hair-on-end” periosteal reaction, and an associated enhancing soft-tissue mass that is well demonstrated at CT. MR images can be used to assess marrow involvement and the perineural spread of tumor and may show an enhancing soft-tissue mass with corresponding restricted diffusion. Iodine $^{123}$I metaiodobenzylguanidine (MIBG) scans show radiopharmaceutical uptake in metastatic neuroblastoma lesions. The differential diagnosis includes lymphoma, Langhans cell histiocytosis, RMS, osteosarcoma, and malignant odontogenic tumors.

**Esthesioneuroblastoma**

Esthesioneuroblastoma is a tumor of neural crest origin that arises from olfactory mucosa at the cribriform plate in the roof of the nasal cavity—hence, the term *olfactory neuroblastoma*. It is very rare in children and has a bimodal distribution, with peaks during the 2nd and 6th decades of life. The clinical presentation includes unilateral nasal obstruction and epistaxis. Cervical node metastatic disease can be found by the time the patient presents. The 5-year overall survival rate is 75%–77%.

At imaging, esthesioneuroblastoma appears as a polypoid or dumbbell-shaped mass extending from the upper nasal cavity to the anterior cranial fossa. CT images best depict osseous destruction and a soft-tissue mass that enhances homogeneously, with bone remodeling and/or destruction in the region of the cribriform plate. Occasionally, speckled internal calcifications or areas of necrosis are present. MR imaging is a key diagnostic tool for evaluating intracranial tumors, which appear as low- to intermediate-signal-intensity masses on T1-weighted images and as intermediate- to high-signal-intensity masses on T2-weighted images, with avid homogeneous enhancement. Cysts, as well as hemorrhage and necrotic areas, may be present within the intracranial portion of the mass; this can result in areas of variable signal intensity at T1- and T2-weighted MR imaging.

Treatment consists of multimodal regimens involving a combination of surgery, chemotherapy, and radiation therapy. Although treatment mainly consists of extensive craniofacial resection and radiation therapy, chemotherapy seems to improve resectability in children and young adolescents (74).

**Conclusion**

A variety of masses may involve the nose, nasal cavity, and nasopharynx in pediatric patients. These lesions may be secondary to congenital and developmental anomalies, inflammatory and infectious processes, neoplasms, or trauma. Lesions may arise from the structures of the nose, adjacent structures, or the nasal-nasopharyngeal mucosa. Depending on the lesion, a child may present with a variety of symptoms, including but not limited to visible palpable masses involving the nasal ala or nasal cavity, airway obstruction, epistaxis, and rhinorrhea. Because the symptoms frequently are not specific to any one disorder, imaging can be quite helpful in determining the site of origin and the involved adjacent structures and guiding the clinician’s presurgical planning for those conditions for which surgical treatment is required. In most patients, the imaging characteristics can be used
to substantially narrow the differential diagnosis, and occasionally they are characteristic, enabling the clinician to suggest a specific diagnosis.

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