Uncommon Intraluminal Tumors of the Gallbladder and Biliary Tract: Spectrum of Imaging Appearances


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Jeffrey S. Klein, MD Hi. I’m Jeff Klein, Editor of RadioGraphics and today I am pleased to have with us Dr. Argha Chatterjee from the Department of Radiology in Northwestern University Feinberg School of Medicine currently an attending at the Tata Medical Center in Calcutta, India who is first author of one of our featured papers in the March 2019 issue of RadioGraphics. Dr. Chatterjee's article is entitled “Uncommon Intraluminal Tumors of the Gallbladder and Biliary Tract: Spectrum of Imaging Appearances”. Dr. Chatterjee welcome to our podcast.

Argha Chatterjee, MD Thank you very much Dr. Klein. It’s a pleasure to be with you today presenting our paper on uncommon intraluminal biliary tumors. I'd also like to take this opportunity to thank my mentor at Northwestern, Dr. Frank Miller, who conceived the idea of this article and is the corresponding author, for his guidance and support.

JSK Terrific. Well thank you. So Dr. Chatterjee I'd like to have you tell the audience how your group, Dr. Miller and colleagues, came to accumulate such a broad spectrum of gallbladder and biliary and neoplasms which are highlighted in your article. Can you describe the endoscopic hepatobiliary and GI pathologist services at Northwestern and how you and your group interfaced with the clinicians in these particular cases?

AC Sure. At Northwestern we have a large, high-volume practice. We currently do over 11,000 abdominal and pelvic MRs and are a major liver transplant center. We see a lot of hepatobiliary patients. We have three intervention gastroenterologists who do ERCP and endoscopic ultrasound. We work closely with them and have a weekly tumor board conference. We work closely with Dr. Guang-Yu who is our expert hepatobiliary pathologist, and his research fellow Dr. Andrew Bandy who is also an author, to obtain pathology correlation. After having seen hepatobiliary IPNs presented by the doctor, the late Richard <Baron> in the University of Chicago course, Dr. Miller had an interest in finding more cases and become more aware of them. As we became more aware of this entity, we realize that we can suggest it more frequently. So gradually in our experience, our diagnosis were no longer retrospective. In fact we started suggesting them prospectively during the process of this research. A nice quote that he often tells me and I often heard him saying is, “You see what you know”.

JSK That’s very true. So beginning with intraductal neoplasms of the bile ducts, your paper begins by briefly describing biliary intraepithelial neoplasms and then discusses intraductal papillary neoplasms of the bile duct or IPN-Bs which are analogous to the intraductal papillary neoplasm, mucinous neoplasm that we see in the pancreas. Table 1 in your paper shows the clinical and pathological characteristics of the four histologic subtypes of IPN-Bs and then you review the multi-modality imaging findings and show several cases. Can we review Figure 3 which shows an oncocytic type subtype of intraepithelial neoplasms of the bile duct in the left hepatic duct as seen on MR imaging?

AC Yes, sure. This is one of the earliest cases that we saw. This was a 61-year-old gentleman who presented with obstructive jaundice. Image 3A is the axial T1-weighted fat saturated MR image that shows a lobulated mass lesion in the proximal left hepatic duct. Figures 3B and C respectively are arterial and portal venous phases of gadolinium enhanced T1-weighted fat saturated MR images. We can see mild enhancement of the lesion and that is persistent. There is no wash out or delayed hyper enhancement. However, the clinching image is the image 3D which is a coronal T2-weighted MR image that shows dilatation of not only the proximal left hepatic duct, but also the distal common bile duct. This is quite characteristic of IPNB as it produces mucin that causes dilatation of the distal bile duct as well in contrast to more common cholangiocarcinoma where only the proximal bile duct typically dilates due to obstruction to bile flow. Image 3E shows the high power photomicrograph of the lesion showing a papillary structure and cells with abundant eosinophilic cytoplasm which is characteristic of an oncocytic tumor.

JSK Terrific, thank you. In your paper you review the imaging mimics of intraductal papillary neoplasms of the bile duct including focal biliary dilatation as may be seen in Caroli disease, primary sclerotic cholangitis, and recurrent pyogenic cholangitis and a variety of intraductal filling defects. Let’s look at one case in particular which is Figure 10 which illustrates the biliary cast syndrome producing intraluminal filling defects in a liver transplant recipient.

AC Sure. This is a particularly interesting case of a 50-year-old lady who has undergone living donor liver transplant a year back. She has presented with features of cholangitis. Figures 10A is the axial fat saturated T2-weighted image that...
shows a hypointense filling defect in the segment seven bile duct with surrounding edema. Coronal T1-weighted image in Figure 10B shows that the filling defect is hyperintense in T1. This T1 hyperintense filling defect is what made us think about biliary cast. Biliary cast syndrome effects about 4 to 18 percent of liver transplant recipients and since these are mostly composed of bilirubin, they are often hyperintense on T1. And that was the suggestion that we can give because of this T1 hyperintensity.

**JSK** Terrific. So after you reviewed a management of intraductal papillary neoplasms of the bile duct, the paper discusses intraductal tubulopapillary neoplasms or ITPN which is a recently described neoplasm affecting women in their sixties that present with abdominal pain and jaundice. These lesions usually arise in the intrahepatic or perihilar bile ducts. Let’s review Figure 16 which illustrates a case of an intrahepatic ITPN.

**AC** Yes. Intraductal intraductal tubulopapillary neoplasm or ITPN is a relatively recently described entity and gradually we have started discovering more cases. This example case is a 36-year-old gentleman who presented with obstructive jaundice. Figure 16A shows that the axial T2-weighted MR image. We can see a fairly large T2 hyperintense mass within the hilar bile ducts. There is upstream biliary dilatation in both lobes of the liver. On T1-weighted contrast enhanced image shown in Figure 16B, the mass shows mild homogeneous enhancement. Since there is no diagnostic feature of ITPN as far as we know yet, the differential was given between cholangiocarcinoma and biliary IPMN. Pathology showed biliary ITPN. We must keep in mind that ITPN is often associated with invasive component or an invasive cancer and often there might be metastasis. So we must keep an eye out for them.

**JSK** Great, well thank you. So moving to the gallbladder, you describe the various intraluminal choleystic masses beginning with non-neoplastic gallbladder polyps, cholesterol polyps and adenomyomatosis of the gallbladder are both common conditions which are fairly characteristic imaging findings. Figure 17 shows the characteristic appearance of the adenomyomatosis on MR. Can we review the findings in this particular case?

**AC** Sure. This is a very important case that shows adenomyomatosis of gallbladder. It is a relatively common condition but often causes confusion with a neoplastic process as it presents with focal gallbladder wall thickening. Therefore, we must be aware of the imaging findings to avoid unnecessary biopsies or surgery. The pathology of adenomyomatosis comprised of hyperplasia of muscosa and muscularis of gallbladder and multiple diverticula within the gallbladder wall which are called Rokitansky-Aschoff sinuses. These sinuses, when identified in imaging as small cystic areas, clinches the diagnosis. Additionally, there are depositions of cholesterol crystals that can be identified by the classic comet-tail artifact in ultrasound. Figure 17 shows multimodality image of a 55-year-old lady. In the first image we see the ultrasound image of the fundus of the gallbladder. We see fundal wall thickening associated with the classic comet-tail or ring-down artifacts. Figure 17B shows axial contrast-enhanced CT scan of the right upper quadrant that again shows the focal wall thickening of the gallbladder. However, we can now see the small cystic areas within the gallbladder wall. In MRCP as shown in Figure 17C, the cystic areas are much better appreciated and seen as a garland or a necklace around the gallbladder lumen. This is often referred to as the pearl necklace sign and often is called the diagnostic sign of adenomyomatosis.

**JSK** Terrific. Well Dr. Chatterjee gallbladder adenomas intra-cholecyctic papillary and papillary tubuloneoplasms that are analogous to the histologically similar bile duct and pancreatic tumors and musonocystic neoplasms of the gallbladder are all fairly uncommon lesions. Primary gallbladder carcinoma is primarily adenocarcinoma with metastases most commonly seen from patients with malignant melanoma. You point out in your paper that gallbladder carcinoma usually presents as a large mass in the gallbladder fossa which replaces the gallbladder. Rare malignancies that can arise in the gallbladder include melanoma, primary of the gallbladder and lymphoma and you provide some examples of these conditions. The paper concludes with some mimics of gallbladder polyps such as tumefactive sludge. And Figure 27 shows a nice example on MR. Can we review this with you?

**AC** Gladly. Tumefactive sludge is an important mimicker of a gallbladder tumor and often is very hard to distinguish on routine abdominal ultrasound since these, unlike gallbladder calculus or regular sludge, do not show movement or change of position. Since these are cholesterol and bilirubin esters mostly suspended in mucus, they often show hyperintensity on T1-weighted images. Figure 27 shows the case of a 40-year-old lady who was thought to have gallbladder mass in her upper abdominal ultrasound. In the first image we see the axial TrueFISP image of gallbladder that shows and intraluminal mass. On T1-weighted MR in the next image we see that the lesion is T1 hyperintense. Figure 27C is the post-contrast image that shows no enhancement within the mass which suggests that the diagnosis of tumefactive sludge as a tumor of this size should have shown some enhancement.

**JSK** Terrific. Well Dr. Chatterjee I want to thank you for taking the time today to discuss with us your paper which addresses uncommon intraluminal tumors of the gallbladder and biliary tract which can be found in the current March 2019 issue of *RadioGraphics*. Dr. Chatterjee, thank you so much for your time today.

**AC** Thanks for having me. Thank you.