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Oriental cholangiohepatitis masquerading as cholangiocarcinoma: A rare presentation that surgeons need to know

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ABSTRACT

INTRODUCTION: The detection of an abnormal hepatic mass with ductal dilatation is highly concerning for malignancy. However, if such patients happen to be immigrants from endemic parts of Asia or South America, further investigations are necessary to rule out oriental cholangiohepatitis, a rare recurrent disease of the hepatobiliary system that can masquerade as cholangiocarcinoma.
PRESENTATION OF CASE: We report a case of a patient of South Asian origin who presented to us with acute cholangitis and moderately dilated left hepatic ducts. The findings were highly suspicious for advanced hepatic malignancy; however the laboratory and pathological investigations remained normal. We suspected an unlikely etiology and proceeded with conservative hepatic resection. The histology revealed cholangiohepatitis without any evidence of malignancy.
DISCUSSION: Cholangiohepatitis is a complex hepatobiliary disease that commonly manifests as recurrent cholangitis or overt biliary sepsis and can rarely present as an abnormal hepatic mass. It results from the development of intrahepatic or extrahepatic strictures that causes stone formation and biliary dilation in the absence of gallbladder disease. Although it is endemic in many parts of the world, it is rare in the western world, and therefore it can present as a significant diagnostic enigma.
CONCLUSION: Cholangiohepatitis is a rare clinical entity that requires a multi-disciplinary team approach. Surgery plays a dominant role in the management of such patients and therefore surgeons need to be aware of this disease.

1. Introduction

Oriental cholangiohepatitis (OCH), also known as recurrent pyogenic cholangitis or Hong Kong disease, is an unusual complex hepatobiliary disease that is rarely seen in the western countries. It is endemic in certain parts of Asia and South America, and with increasing number of immigrants arriving from these areas; many such cases are referred to surgeons for advanced surgical management.[1,2]

OCH is characterized by development of intrahepatic and extrahepatic strictures, pigment stones and biliary dilation in the absence of gallbladder disease.[2] The patients present with spectrum of findings ranging from mild recurrent abdominal pain to complicated biliary sepsis and hepatic masses.[1,3] These patients are treated successfully using a multidisciplinary approach.

2. Presentation of case

A 74 year old male of South Asian origin was admitted with abdominal pain, nausea and vomiting for few weeks. His medical history was significant for gout, chronic renal insufficiency, hyperlipidemia and cerebrovascular accident. An initial ultrasound was unremarkable, but the CT scan revealed a suspicious area with moderate dilatation of the left hepatic ducts (Fig. 1a). Upper endoscopy showed a bulge at the duodenal bulb, however, the blood tests including CA 19-9 levels (2.3 U/ml) and biopsies came back negative for any abnormal cells/malignancy. The patient underwent a transhepatic cholangiogram with brush biopsy and placement of a biliary drainage catheter. The cholangiogram revealed an obstruction at the junction of the right and left biliary ducts, without evidence of stones (Fig. 1b), while the cytology revealed a normal ductal epithelium with mild atypia. The procedure was complicated with development of fever and leukocytosis for which he was treated with intravenous fluids and antibiotics for presumed cholangitis. At this stage he was referred for evaluation to our center.

As the etiology of biliary obstruction in the presence of a suspicious mass remained unclear, it was decided to proceed with conservative surgery. The patient successfully underwent left

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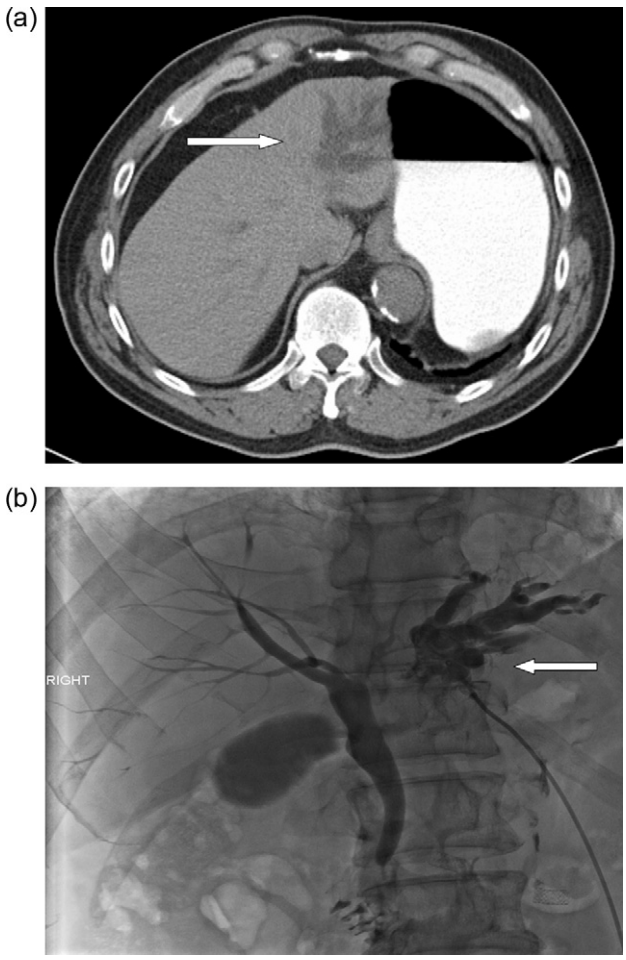

Fig. 1. (a) Imaging–CT scan showing dilated ducts. (b) Imaging–cholangiogram showing dilated ducts.

hepatic lobectomy and cholecystectomy. An intraoperative hepatic ultrasound did not detect any mass, while the intraoperative frozen section of the bile duct margin was negative for malignancy. Serial sectioning of the lobectomy specimen showed marked fibrosis around the larger bile ducts. Numerous, hard and crumbly, yellow and black calculi, and sludge, was present within the bile ducts (Fig. 2). The bile ducts and surrounding hepatic parenchyma were sampled for microscopic evaluation. Microscopically, there was marked periductal fibrosis, with an associated infiltrate of acute and chronic inflammatory cells in the ducts. Although there was mild duct epithelial atypia, which was secondary to the calculi, there was no evidence of malignancy, either in the ducts or in the surrounding hepatic parenchyma. These features are diagnostic of Oriental cholangiohepatitis.

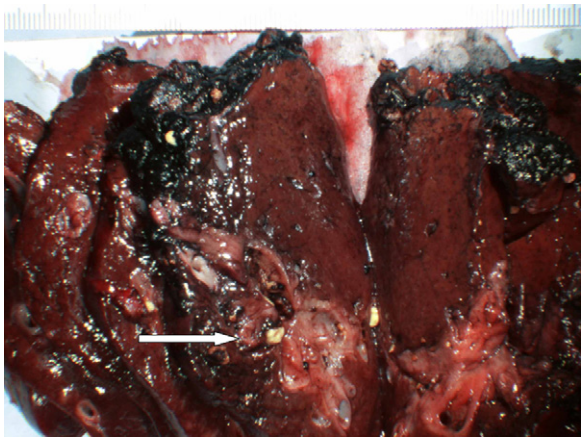

Fig. 2. Pathology–cut section showing intra-hepatic stones, ductal dilatation and focal strictures.

The patient had an uneventful postoperative recovery. Follow-up MRCP six months later showed no evidence of right lobe involvement. He remains asymptomatic one year after resection.

3. Discussion

Oriental cholangiohepatitis was initially described in 1930's by Digby[4] and has since been known by various names including recurrent pyogenic cholangitis (RPC), Hong Kong disease, and Oriental infestational cholangitis.[1] It is characterized by the development of intrahepatic and extrahepatic strictures, pigment stones and biliary dilation in the absence of gallbladder disease.[2] Due to bile stasis and stone formation proximal to the site of strictures, patients often develop recurrent episodes of cholangitis with symptoms ranging from mild abdominal pain to pancreatitis and overt biliary sepsis.[1,3]

The disease is reported to be typically seen in people who live in or have immigrated from certain endemic parts of Asia and South America.[1,3,5] However, there have been recent reports of patients who are neither from endemic areas nor have visited these areas.[6,7] There is no specific gender predisposition and the peak incidence is seen in the third and fourth decades of life, albeit in endemic areas it can also be seen in infants and in patients above the age of 60 years.[5,8,9]

The etiology is unknown, although the observation that this disease occurs in certain endemic areas where biliary parasites such as liver flukes and Ascaris are common suggests that infestation may play a role in pathogenesis. The three major liver flukes infecting humans (*Clonorchis sinensis*, *Opisthorchis* species, and *Fasciola hepatica*) and Ascaris have been shown to cause epithelial damage and biliary obstruction, possibly initiating the inflammatory response that eventually leads to overt disease.[9] However, despite the epidemiologic association, evidence supporting the role of these infections in the pathogenesis of OCH is inconclusive. While these infections can be demonstrated in 20–45% of patients with OCH, they are not uniformly present.[8–11] Further, OCH is relatively common in some Asian countries in which these parasites are not endemic.[12]

The characteristic clinical presentation is the development of recurrent cholangitis, secondary to bile stasis and stone impaction proximal to the ductal strictures. Stones are formed de novo within the intrahepatic bile ducts, and are primarily pigmented stones.[5,8] The exact mechanism of stone formation is not well defined.[1] In addition, the bile ducts are markedly abnormal, characterized by extrahepatic and intrahepatic ductal dilatation with areas of focal stricturing. The ductal walls are fibrotic with an intense inflammatory cell infiltration.[2,3,5,8,10] The bile is often purulent and contains debris of small stones, sludge, bile pigment, desquamated epithelial cells and bacteria.[10] Common bacteria cultured from this bile include E. coli, Klebsiella, Pseudomonas, and Proteus species and, less frequently, anaerobes, although growth of multiple organisms is common.[8,11]

In a study by Sperling and colleagues, the most common presentation was cholangitis (44%), followed by abdominal pain without cholangitis (32%), and pancreatitis (17%).[13] Patients may also present with abscess formation at distant sites, such as the lungs and the brain. Rarely, patients are referred with hemobilia, portal vein thrombosis and suspected hepatic masses.[10]

The finding of a hepatic mass is concerning because these patients have a predisposition for developing cholangiocarcinoma. Cholangiocarcinoma is known to frequently arise in diseased and atrophic liver. In a study of 427 patients, Jan[14] found cholangiocarcinoma in 3 percent of patients with OCH while in another report, Chen and colleagues[15] noted cholangiocarcinoma in 55 of the 1105 patients studied over a three-year period. Further, it has been proposed that cholangiocarcinoma should be suspected in patients who have worsening jaundice or weight loss with an unexplained increase in liver enzymes.[15]

For reasons that are not quite well understood, the left hepatic duct and its branches, especially the left lateral segmental duct, are commonly involved in the early part of the disease. However, in the later stages, both right and the left hepatic lobes along with the extrahepatic biliary tract tend to get involved.[2,16,17] A proposed explanation is that the left hepatic ducts come off at a more acute angle compared with the right hepatic ducts, thus predisposing to stasis and stricture formation. Some other findings include a hypertrophied papilla of the sphincter of Oddi due to repeated passage of stones and sludge, a scarred and shrunken liver with multiple adhesions or abscesses, dilated ducts and abnormal masses that may confused for abnormal hepatic lesions.[9,10]

The diagnosis is established by combination of imaging studies and pathology. Ultrasound can detect ductal dilation, stones and abscesses in 85–90% of patients.[9] More detailed information can be obtained using a CT scan, which can delineate dilated central intrahepatic ducts, tapering peripheral ducts, duct wall enhancement, abscesses, bilomas, and stones.[9,18] CT scans also help in differentiating localized diseased segments and atrophied parenchyma from the rest of the normal parenchyma. Other helpful modalities include cholangiography and magnetic resonance cholangiopancreatography. Additionaly, all patients need to have their stool tested for ova and parasites.[9]

Once the patient is confirmed to have OCH, the further management requires a multidisciplinary team approach, with endoscopic, surgical and radiologic intervention as necessary.[1,2] Patients presenting with cholangitis are treated appropriately with fluid resuscitation, antibiotics and biliary drainage.[2,19] Acutely ill patients in whom ERCP or radiologic intervention fails to achieve desirable results generally require surgical intervention that may entail cholecystectomy, common bile duct exploration or resection. If an isolated area of the liver is affected, resection can be curative.[1,2]

Hepatic resection is advised for patients with focal diseased hepatobiliary segments, atrophied lobes or suspicious masses. Hepatic lobectomy and bilateral partial hepatectomy are some of the common surgical techniques described for management of these patients. Although, there have been no large controlled trials comparing hepatic resection with other modalities, there are retrospective reports of patients followed in major centers across Asia over many years. These reports have suggested better quality of life, lower rates of secondary biliary cirrhosis, cholangiocarcinoma, and mortality in patients treated with surgical resection.[1,2,20]

4. Conclusion

In conclusion, Oriental cholangiohepatitis is a complex disease of the hepato-biliary system that requires a multi-disciplinary team approach. Surgery plays a dominant role in the management of these patients and surgeons need to be aware of this rare entity.

Conflict of interest statement

The authors have no conflict of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

References

1. Al-Sukhni W, Gallinger S, Pratzer A, et al. Recurrent pyogenic cholangitis with hepatolithiasis—the role of surgical therapy in North America. *J Gastrointest Surg* 2008;**12**(3):496–503.
2. Harris HW, Kumwenda ZL, Sheen-Chen SM, Shah A, Schecter WP. Recurrent pyogenic cholangitis. *Am J Surg* 1998;**176**(1):34–7.
3. Ong GB. A study of recurrent pyogenic cholangitis. *Arch Surg* 1962;**84**:199–225.
4. Digby K. Common duct stones of liver origin. *Br J Surg* 1930;**17**:578.
5. Yellin AE, Donovan AJ. Biliary lithiasis and helminthiasis. *Am J Surg* 1981;**142**(1):128–36.
6. Wilson MK, Stephen MS, Mathur M, Sheldon D, Storey D. Recurrent pyogenic cholangitis or "oriental cholangiohepatitis' in occidentals: case reports of four patients. *Aust N Z J Surg* 1996;**66**(9):649–52.
7. Bergman S, Di Carlo A, Chaudhury P, Blum C, Nahal A, Metrakos P. Recurrent pyogenic cholangitis in a white woman. *Can J Surg* 2007;**50**(5):E9–10.
8. Carmona RH, Crass RA, Lim Jr RC, Trunkey DD. Oriental cholangitis. *Am J Surg* 1984;**148**:117.
9. Lim JH. Oriental cholangiohepatitis: pathologic, clinical, and radiologic features. *AJR Am J Roentgenol* 1991;**157**:1.
10. Chou ST, Chan CW. Recurrent pyogenic cholangitis: a necropsy study. *Pathology* 1980;**12**:415.
11. Seel DJ, Park YK. Oriental infestational cholangitis. *Am J Surg* 1983;**146**:366.
12. Nakayama F, Koga A. Hepatolithiasis: present status. *World J Surg* 1984;**8**:9.
13. Sperling RM, Koch J, Sandhu JS, Cello JP. Recurrent pyogenic cholangitis in Asian immigrants to the United States: natural history and role of therapeutic ERCP. *Dig Dis Sci* 1997;**42**:865.
14. Jan YY, Chen MF, Wang CS, et al. Surgical treatment of hepatolithiasis: long-term results. *Surgery* 1996;**120**:509.
15. Chen MF, Jan YY, Wang CS, et al. A reappraisal of cholangiocarcinoma in patient with hepatolithiasis. *Cancer* 1993;**71**:2461.
16. Gott PE, Tieva MH, Barcia PJ, Laberge JM. Biliary access procedure in the management of oriental cholangiohepatitis. *Am Surg* 1996;**62**(11):930–4.
17. Lim JH, Ko YT, Lee DH, Hong KS. Oriental cholangiohepatitis: sonographic findings in 48 cases. *AJR Am J Roentgenol* 1990;**155**(3):511–4.
18. Chan FL, Man SW, Leong LL, Fan ST. Evaluation of recurrent pyogenic cholangitis with CT: analysis of 50 patients. *Radiology* 1989;**170**:165.
19. Fan ST, Choi TK, Wong J. Recurrent pyogenic cholangitis: current management. *World J Surg* 1991;**15**:248.
20. Cheung MT, Kwok PC. Liver resection for intrahepatic stones. *Arch Surg* 2005;**140**(10):993–7.