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Extensive primary retroperitoneal fibrosis (Ormond's disease) with common bile duct and ureteral obstruction: A rare case report

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ABSTRACT

INTRODUCTION: Idiopathic retroperitoneal fibrosis (Ormond's disease) may involve the perirenal tissue, mesentery and biliary system in extremely uncommon situations in addition to classical compression of retroperitoneal structures particularly the ureters.
PRESENTATION OF CASE: We report the case of a 60 year's old man with clinical manifestation of obstructive jaundice, due to the common bile duct narrowing caused by a primary retroperitoneal fibrosis. Magnetic resonance cholangiopancreatography (MRCP) showed the presence of intrahepatic bile duct obstruction, suggesting the diagnosis of a hilar cholangiocarcinoma. Endoscopic retrograde cholangiopancreatography (ERCP) showed strictures in the proximal common bile duct. A biliary endoprosthesis was inserted at ERCP. Histological investigations as well as CT-scan were compatible with retroperitoneal fibrosis.
DISCUSSION: The main clinical presentation of the Ormond's disease is compression of the ureters, and less commonly blood vessels and nerves. Our patient presented with obstructive jaundice, due to the common bile duct (CBD) compression, which was unusual. The first description of the extrahepatic biliary obstruction secondary to retroperitoneal fibrosis was made in 1964 and since then only 13 cases have been reported in the medical literature.
CONCLUSION: Retroperitoneal fibrosis can cause compression of the CBD and therefore mimic a cholangiocarcinoma. Patients can be successfully managed with long-term CBD stent placement.
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1. Introduction

Retroperitoneal fibrosis (RPF) is a rare inflammatory fibrotic process causing compression of retroperitoneal structures particularly the ureters [1]. Idiopathic RPF (Ormond's disease) may involve the perirenal tissue, mesentery and biliary system in extremely uncommon situations [2]. In this case report, we describe an uncommon presentation of retroperitoneal fibrosis presenting with a malignant-appearing common bile duct stricture.

2. Presentation of case

A 60 year's old man presented with a one month history of jaundice, weight loss and right upper quadrant pain. Clinical examination found no stigmata of chronic liver disease. Laboratory tests revealed a C-reactive protein (CRP) at 50 mg/dL, blood urea nitrogen at 0,6 g/L and creatinine level at 23 mg/L. Total bilirubin was 137 umol/L (normal 2–20 umol/L) with direct bilirubin at 120 umol/L (normal 0–3.4 umol/L). Abdominal ultrasonography showed splenomegaly and intrahepatic duct dilatation but no evidence of stone or mass in the head of the pancreas; there was also bilateral hydronephrosis.

Magnetic resonance cholangiopancreatography (MRCP) revealed a suspected lesion in the hepatic hilum responsible for a significant dilatation of the intrahepatic bile ducts. A presumptive diagnosis of cholangiocarcinoma was made. Endoscopic retrograde cholangiopancreatography (ERCP) showed strictures in the proximal common bile duct. A sphincterotomy with brushings and biopsies were performed, and a 10 F stent was placed to drain the biliary tree. The histology revealed an acute inflammatory and infiltrative process without any evidence of malignancy.

Abdomino-pelvic CT-scan revealed a soft tissue surrounding the aorta with extension to the renal and hepatic hilum. Laterally, RPF spreads to involve the ureters and right kidney causing bilateral hydronephrosis (Fig. 1). A biopsy of the retroperitoneal mass under computed tomography control was performed. Histologic examination showed a non-specific fibrotic and inflammatory tissue with no malignancy signs, compatible with idiopathic retroperitoneal fibrosis (Ormond's disease). The patient underwent successful bilateral ureteral stenting (Fig. 2) with good renal function evolution. Treatment with corticosteroids and immunosuppressive therapy was initiated.

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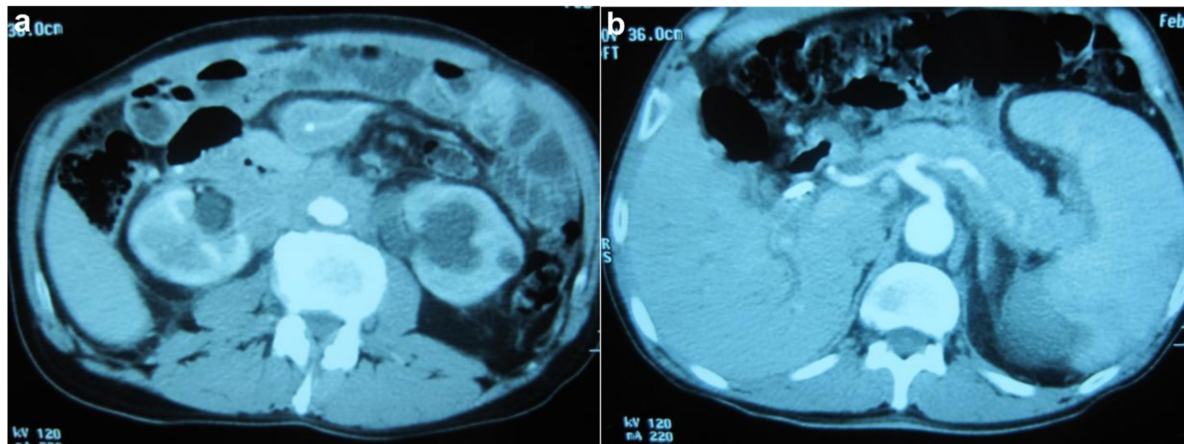

Fig. 1. (a and b): Abdomino-pelvic CT scan with bilateral hydronephrosis and soft tissue surrounding the abdominal aorta, extended to the hepatic and renal hilum, including the bifurcation of iliac common arteries.

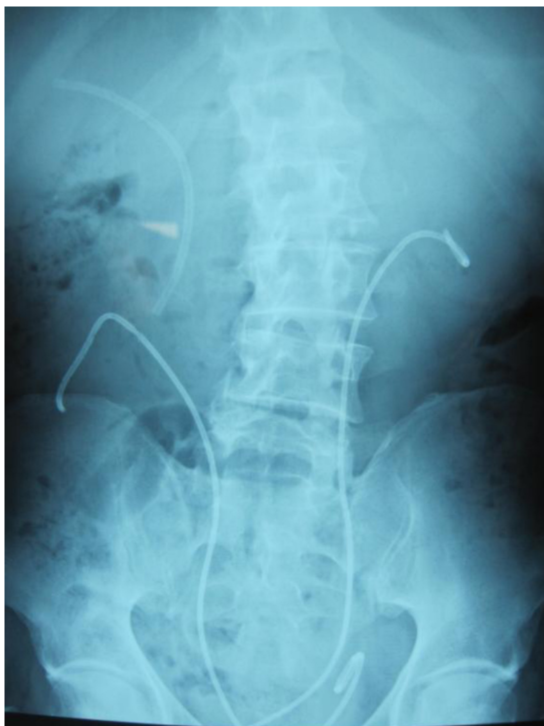

Fig. 2. Abdominal radiograph showing correct positioning of the ureteral and biliary stents.

3. Discussion

Retroperitoneal fibrosis, also called Ormond's disease, does not have a definitive cause [3]. The main mechanism is supposed to be an autoimmune disorder due to the presence of antibodies against ceroid, a complex of oxidized lipid and protein [4,5]. On the other hand, malignant tumors and proliferations can occur in 8% of cases [6,7]. The main clinical presentation of the Ormond's disease is often the ureters's compression, as well as blood vessels and nerves. Our patient appeared with clinical manifestation of obstructive jaundice, due to the common bile duct narrowing, which was an unusual presentation [4,8]. The first description of the extrahepatic biliary obstruction secondary to RPF was made in 1964 [9]. Since then 13 cases have been reported in the medical literature. Eleven were operated and two cases were managed with a single plastic stent [9,10]. Treatment of RPF focuses on relieving any compression from body structures. For ureteric involvement, ureteric stenting or ureterolysis are options. Obtaining deep tissue biopsies can be done to confirm the diagnosis and to exclude the malignant type. Corticosteroids (and other immunosuppressants) can be used to prevent disease progression [8].

4. Conclusion

In conclusion, this atypical case illustrates that RPF can affect the CBD and mimic a cholangiocarcinoma. Such patients can successfully be managed using long-term tumor stents.

Conflict of interest

Authors have no conflict of interest to declare.

Author contribution

Dr. Fahd Khalil: study concept and writing the paper.
Dr. Ouslim Hicham: data collection.
Dr. Tarik Mhanna: data collection.
Dr. Ali Barki: data interpretation and correction.

Guarantor

Nothing to declare.

References

[1] M.S. Cappell, Obstructive jaundice due to retroperitoneal fibrosis involving the head of the pancreas, J. Clin. Gastroenterol. 18 (1994) 53–56.
[2] J. Vega, H. Goecke, H. Tapia, et al., Idiopathic retroperitoneal fibrosis treatment with colchicines and steroids: a case series, Am. J. Kidney Dis. 53 (2009) 628–637.
[3] D. Richard, M.D. Swartz, Idiopathic retroperitoneal fibrosis: a review of the pathogenesis and approaches to treatment, Am. J. Kidney Dis. 54 (September 3) (2009) 546–553.
[4] E. Chris, M.D. Lascarides, J. Edmund, M.D. Bini, E. Newman, Intrinsic common bile duct stricture: an unusual presentation of retroperitoneal fibrosis, Gastrointestinal Endoscopy 50 (1) (1999).
[5] M.-F. Zhao, Y. Tian, K.-J. Guo, Z.-G. Ma, H.-H. Liao, Common bile duct obstruction due to fibrous pseudotumor of pancreas associated with retroperitoneal fibrosis, World J. Gastroenterol. 10 (20) (2004) 3078–3079.
[6] M. Torella, S. Luca, D. Santo, A. Della Corte, S. Esposito, Extensive retroperitoneal fibrosis with duodenal and ureteral obstruction, Texas Heart Inst. J. 30 (2003) 311–313.

[7] S. Tamura, Y. Yokoyama, K. Nakajo, T. Morita, A rare case of idiopathic retroperitoneal fibrosis involving obstruction of the mesenteric arteries, duodenum, common bile duct, and inferior vena cava, Int. Med. 42 (September 9) (2003).

[8] M. Quante, B. Appenrodt, S. Randerath, M. Wolff, H.P. Fischer, T. Sauerbruch, Atypical Ormond's disease associated with bile duct stricture mimicking cholangiocarcinoma, Scand. J. Gastroenterol. 44 (January 1) (2009) 116–120.

[9] B. Matthew Smith, D. Grove, Obstructive jaundice secondary to retroperitoneal fibrosis: long term nonsurgical management resulting in regression of obstruction, AJG 95 (9) (2000).

[10] V. Pugliese, M. Conio, G. Nicolo, S. Saccomanno, B. Gatteschi, Endoscopic retrograde forceps biopsy and brush cytology of biliary strictures: a prospective study, Gastrointestinal Endoscopy 42 (1995) 520–526.

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