INTRODUCTION

Described for the first time by John Kelso Ormond in 1948, idiopathic retroperitoneal fibrosis usually involves Gerota’s fascia and compresses the ureters leading to the apparition of progressive lumbar pain and urinary dysfunction inducing various degrees of renal failure. In certain cases, other organ might be affected inducing a different clinical aspect, in the absence of urinary tract involvement signs.

ABSTRACT

Idiopathic retroperitoneal fibrosis, also known under the name of Ormond’s disease usually compresses the retroperitoneal structures, especially the ureters. In rare cases other structures such as the biliary system can be involved. We report the case of a 49-year-old patient diagnosed with Ormond’s disease two years before, who developed jaundice and acute cholecystitis due to the progression of the retroperitoneal fibrosis.

Keywords: idiopathic retroperitoneal fibrosis, common bile duct obstruction, acute cholecystitis.

CASE REPORT AND MINIREVIEW

A RARE CASE OF ACUTE CHOLECYSTITIS AND COMMON BILE DUCT OBSTRUCTION DUE TO ORMOND’S DISEASE – A CASE REPORT AND LITERATURE MINIREVIEW

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RÉSUMÉ

Un cas rare de chélocystite aigue et d’obstruction des voies biliaires du à la maladie d’Ormond- rapport de cas et revue de la littérature

La fibrose rétropéritonéale idiopathique, également connue sous le nom de maladie d’Ormond, comprime généralement les structures rétropéritonéales, en particulier les uréters. Dans de rares cas, d’autres structures telles que le système biliaire peuvent être impliquées. Nous rapportons le cas d’un patient de 49 ans diagnostiqué avec la maladie d’Ormond deux ans auparavant, qui a développé une jaunisse et une cholécytite aigüë en raison de la progression de la fibrose rétropéritonéale.

Mots-clés: fibrose rétropéritonéale idiopathique, l’obstruction des voies biliaires, la cholécytite aigüë
For example, retroperitoneal fibrosis might affect the common biliary duct, inducing the appearance of jaundice or unexplained elevated serum hepatic enzymes in association with significant biliary duct enlargement. In the meantime, the presence of jaundice and cholestasis in the absence of upper abdominal pain is usually significantly suggestive for malignancy. Most often performing preoperative studies such as magnetic resonance imaging or endoscopic retrograde cholangiopancreatoscopy usually provides a good orientation of the diagnosis. However, in certain cases this fact is not possible, imposing the necessity of surgery.

Extensive primary retroperitoneal fibrosis, also known as Ormond’s disease is most often characterized by the involvement of perirenal tissues although mesenteric and biliary system involvement has been reported so far in extremely rare situations.

Its pathogenesis is not fully understood, various hypothesis such as immunologic mechanisms being suspected so far.

**CASE REPORT**

A 47-year-old patient with medical past history of diabetes presented for diffuse abdominal pain, weight loss and nausea and was diagnosed after performing a computed tomography with a large retroperitoneal tumor measuring 54x47x84 mm developed in the mesenteric root, rounding the abdominal aorta and inferior vena cava. The patient was submitted to surgery, at that moment a biopsy being performed. The histopathological studies revealed the presence of an idiopathic diffuse retroperitoneal fibrosis (Ormond’s disease) with fibro-inflammatory elements. The immunohistochemical studies revealed the diffuse positivity of actin, cluster designation (CD) 68, CD 34 (at the level of the blood vessels) and the negativity of ALK1, AE1-AE3 while Ki67 proved to be positive in up to 5% of the tumoral cells. Postoperatively the patient was submitted to adjuvant chemotherapy with six series of ifosfamide, dacarbazine and epirubicin. Two years later the patient presented for upper abdominal pain, jaundice and nausea. The computed tomography revealed the presence of a large retroperitoneal mass measuring 146/65/73 mm invading the abdominal aorta and compressing the common bile duct, inducing an intrahepatic biliary ducts dilatation; in the meantime the gallbladder presented diffuse thickened walls with local signs of acute lithiasic cholecystitis. The patient was referred to surgery; intraoperatively a typical aspect of acute lithiasic pycholecystitis and common bile duct lithiasis was found so cholecystectomy and Kehr drainage were performed. The postoperative course was uneventful the patient being discharged in the fifth postoperative day.

**DISCUSSION**

The definitive cause of Ormond’s disease is not well established so far, although autoimmune and immunological disorders have been incriminated. Most authors sustain the hypothesis of the presence of antibodies against ceroid, although other causes such as medication (hydralazine, bromocriptine, methysergide, beta-blockers), malignant processes (carcinoids, lymphomas, sarcomas, breast, colorectal or bladder carcinomas), radiation therapy, surgery for aortic aneurysms or infections (tuberculosis) have been cited so far.

Most of the cases are diagnosed due to the presence of direct and indirect signs of ureteral compression, although other structures such as nerves, blood vessels or bile ducts might be affected. According to Ormond’s theory, the course of the disease includes the onset of the process followed by fibrosis of the retroperitoneal tissues and contracture of the fibrous mass inducing compression of the affected structures. More recent studies sustain the influence of immunoglobulin G4 (IgG4) producing plasma cells which induce tumor swelling of certain viscera and different degrees of fibrosis. As a consequence, the tumefaction of the involved organ might appear which leads to its destruction.

Biliary obstruction due to Ormond’s disease has been reported for the first time in 1964; from that moment, fourteen cases have been reported so far. Eleven cases were submitted to surgery while the other three cases have been managed conservatively. However, biopsy should be always performed in order to differentiate the benign subtype from the malignant one. Malignant proliferation has been reported in up to 8% of cases.

The goals of therapeutic strategy in Ormond’s disease consist in relieving the obstruction, stop the progression of the disease and prevent the recurrence.

The main therapeutic options consist in stent placement or other surgical procedures in order to relieve the compressed structures followed by medical treatment based on corticosteroids and immunosuppressive agents in order to prevent the evolution of the disease. In cases presenting secondary retroperitoneal fibrosis, the treatment is addressed to the cause which has led to this pathology while in cases with idiopathic fibrosis prednisolone has been reported as the first intention therapy. In case of inadequate response, azathioprine, methotrexate, rituximab or mycophenolate mofetil may be added while other authors sustain the benefits of tamoxifen’s.
**Figure 1.** The aspect of retrograde dissection of the gallbladder

**Figure 2.** The aspect after cholecystectomy – retroperitoneal fibrosis and common bile duct dilatation

**Figure 3.** Releasing lithiasic material from the common bile duct

**Figure 4.** Placing a Kehr’s tube
administration as first line therapy. The effect of tamoxifen in Ormond’s disease is related to the inhibition of fibroblast proliferation and angiogenesis; however, this action proved to be efficient especially in non-malignant conditions. In von Bommel’s study published in 2006 which included 19 cases with non-malignant Ormond’s disease administration of tamoxifen (20 mg twice daily) significantly improved the outcomes.

CONCLUSION

Ormond’s disease is a very rare entity inducing retroperitoneal fibrosis which usually affects the ureters. However, in certain cases other structures such as the common bile duct might be affected. In these cases severe modifications such as common bile duct stenosis, lithiasis and acute cholecystitis might be observed. In order to provide a good outcome of these cases, biliary duct decompression followed by systemic therapy should be taken into consideration.

Acknowledgement: This work was supported by a grant from the Romanian National Authority for Scientific Research and Innovation, CNCS – UEFISCDI, project number PN-II-PI-2014-4-2533.

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