FAMILIAL RETROPERITONEAL FIBROSIS: TWO UNUSUAL CASES

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SUMMARY: Retroperitoneal fibrosis is an inflammatory process which infrequently affects any part of the alimentary tract from the oesophagus to rectosigmoid. A number of etiological factors is proposed, but a familial tendency has not been described before. The familial link between our two cases, one with both colonic and bilateral ureter involvement and the other with unilateral ureter involvement, prompted us to report them.

Key Words: Retroperitoneal Fibrosis, Osmond’s Disease, Gastrointestinal Tract, Heredity.

INTRODUCTION

Retroperitoneal fibrosis (RPF) is an inflammatory process which usually presents in the fifth or sixth decades with non-specific symptoms and retroperitoneal involvement of the ureters and vascular structures. The urinary system tract problems are frequently seen in RPF. The alimentary tract is rarely involved in RPF and consequently this is infrequently considered in the differential diagnosis of bowel diseases. RPF may affect any part of the alimentary tract from the oesophagus to rectosigmoid (6, 11).

We present 2 sisters with retroperitoneal fibrosis, one of which has gastrointestinal tract involvement.

CASE REPORT

A 40-year-old lady presented with an abdominal pain in the right lumbar region. She had experienced lethargy and anorexia for a month and diarrhea last week. Clinical examination revealed abdominal tenderness, defense, direct rebound and hypactive bowel sounds in oscillation. The chest and plain abdominal radiographs were normal, with no evidence of bowel obstruction. Ultrasonography (USG) revealed right hydronephrosis, an intraperitoneal mass at the lumbar region, increased bowel wall thickness and hepatomegaly. An intravenous urogram revealed right hydronephrosis and dilatation of pelvis renalis. Abdominal tomography revealed that there was multiple synchieus in the retroperitoneal region (Fig. 1). Colonoscopy revealed, multiple external compressions to the colon which were confirmed as lymphadenopathy. Based on sonography findings, laparoscopy and biopsy were considered. A laparoscopy was performed and no mass lesion was detected. Also, multiple synchieus were seen in this laparoscopy and biopsy was performed. Histology showed dense connective tissue with focal inflammatory change consistent with retroperitoneal fibrosis. No neoplastic or carcinoid cells were seen. We have learned that the sister of our patient had also been ad-
mitted to urology department with a temporary abdominal pain and urologic problems. Her intravenous urogram had revealed bilateral hydronephrosis. As she had not accepted the procedure we could not perform a laparoscopy (Fig. 2).

Steroid therapy was commenced, initially prednisolone 40 mg daily for 2 months, then 10 mg daily for a further 12 months before gradually terminating treatment. The abdominal tomography was repeated after the treatment (Fig. 3). All symptoms disappeared after steroid therapy in a few months.

DISCUSSION

RPF, first described in 1948, represents an incompletely understood entity characterised by a proliferation of retroperitoneal fibrous tissue with varying amounts of chronic inflammatory change (1,4). The etiology of retroperitoneal fibrosis (Ormond's disease) has not been clarified. Most cases are considered to be idiopathic and may represent an autoimmune reaction to components within atheromatus plaques (9, 12). It is also recognised to occur following retroperitoneal haemorrhage or trauma, regional anitis, pancreatitis, and radiation. It has been found in association with drugs and mainly ergot derivatives such as methysergide (2, 3). Despite all of these etiologic factors genetic tendency has not been shown in literature. Our two cases had no history of trauma, regional anitis, pancreatitis and exposition to radiation. Both of them had no history of using methysergide.

Retroperitoneal involvement often causes ureteric obstruction, perianotic fibrosis and venous obstruction. Bowel involvement in RPF is uncommon. Bowel involvement may present with pseudoobstruction caused by fibrotic mesenteric plaques (18). These bowel involvements in RPF is affecting the small bowel. Strictures of the large bowel in RPF have been reported in only 11 cases (2, 4, 5, 7, 8, 10, 11, 15). By adding our one case which has stricture of large bowel the number of cases in the literature rise to 12.

Symptoms of presentation resembled ileus. Colonoscopic and barium examinations of the involved large bowel have shown normal mucosa with narrowing and angulation. Additional imaging in cases of large bowel involvement has invariably demonstrated either unilateral or bilateral ureteric
obstruction.

Steroid treatment is known to be the best choice in RPF with ureter involvement. The beneficial effect of steroids in RPF is thought to be due to their anti-inflammatory action and inhibition of fibrotic tissue maturation. In our patient after steroid therapy symptoms were diminished considerably but not completely. Steroids are believed to be less effective in cases of mature fibrosis (12, 13). On the other hand, the disappearance of retroperitoneal fibrous tissue demonstrated by CT, on low-dose postoperative steroid therapy, was reported (17). The use of steroid in the adjuvant treatment of RPF was first reported in 1958 (16) but there is only one report documenting its use in large bowel involvement.

RPF has been known to affect a number of widely separate organ systems but can also affect only one organ. Literature survey reveals that RPF can be responsible for only unilateral ureterolysis (1). Because it was not allowed we could not support the pathologic diagnosis with biopsy. For the differential diagnosis of unilateral ureterolysis with noninvasive methods we looked for malignancy and tuberculosis to no avail. Low doses of steroid therapy (20 mg/day) was given to our patient and at the third month of therapy the decrease in her complaints supported our diagnosis of retroperitoneal fibrosis.

The familial factor between our two cases: one with colonic symptoms and the other having unilateral ureter involvement urged us to write this report on retroperitoneal fibrosis.

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