FAMILIAL RETROPERITONEAL FIBROSIS : TWO UNUSUAL CASES

Selahattin ÜNAL, M.D., Burçak KAYHAN, M.D., Zeynep AKI, M.D., Ahmet GÖRGÜL , M.D., Uğur KANDİLCİ, M.D.

Gazi University, Faculty of Medicine, Department of Internal Medicine, Ankara, Turkey
Gazi Medical Journal 7 : 23-25, 1996

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 a unilateral ureter involvement, prompted us to report them.*

Key Words : *Retroperitoneal Fibrosis, Ormond'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 lumbal region. She had experienced lethargy and anorexia for a month and diarrhea last week. Clinical examination revealed abdominal tenderness, defense, direct rebound and hypoactive bowel sounds in oscultation. The chest and plain abdominal radiographs were normal, with no evidence of bowel obstruction. Ultrasonography (USG) revealed right hydroureteronephrosis, an intraperitoneal mass at the lumbal region, increased bowel wall thickness and hepatomegaly. An intravenous urogram revealed right hydroureteronephrosis and dilatation of pelvis renalis. Abdominal tomography revealed that there was multiple synechias 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 synachiae 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 hydroureteronephrosis. 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 (14). The etiology of retroperitoneal fibrosis (Ormond's disease) has still not been clarified. Most cases are considered to be idiopathic and may represent an autoimmune reaction to components within atheromatous plaques (9, 12). It is also recognised to occur following retroperitoneal haemorrhage or trauma, regional anteritis, 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 enteritis, pancreatitis and exposition to radiation. Both of them had no history of using methysergide.

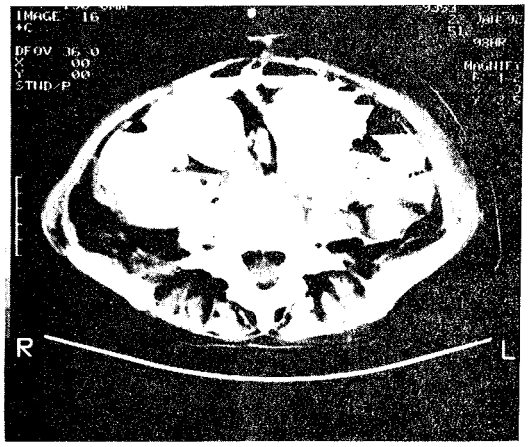

Fig - 1 : The tomographic view of the retroperitoneal multiple synechia.

Fig - 2 : The urographic view of the bilateral hydroureteronephrosis.

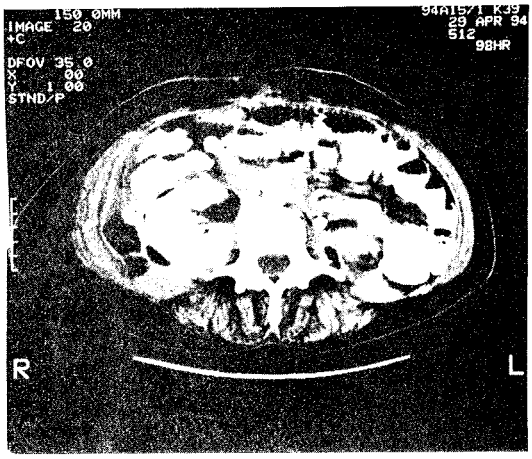

Fig - 3 : The tomographic view after the treatment.

Retroperitoneal involvement often causes ureteric obstruction, periaortic fibrosis and venous obstruction. Bowel involvement in RPF is uncommon. Bowel involvement may present with pseudo-obstruction 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 reflect 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 their use in large bowel involvement.

RPF has long 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 responsiblyle 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.

Correspondence to : Dr.Selahattin ÜNAL
Gazi Üniversitesi Tıp Fakültesi
İç Hastalıkları Anabilim Dalı
Beşevler
06510 ANKARA - TÜRKİYE
Phone : 312 - 214 10 00 / 5811

REFERENCES

1. Fujisava M, Higashi Y, Tonaka H, et al : Idiopathic retroperitoneal fibrosis : a case of unilateral lesion. Hinyokika-Kiyo 1993; 39 (5) : 451-454.
2. Graham JR : Methysergide for prevention of headache: experience in five hundred patients over three years. N Engl J Med 1964; 270 : 67-72.
3. Graham JR, Howard IS, Le Compte PR, Sadowsky NL : Fibrotic disorders associated with methysergide therapy for headache. N Engl J Med 1966; 274 : 359-368.
4. Gelford GJ, Cromwell DK : Methysergide, retroperitoneal fibrasis and rectosigmoid stricture. AJR 1968; 104 : 566-570.
5. Hissong SL, Freimans AK : Retroperitoneal fibrosis : extraperitoneal lesions. Report of three unusual areas of involvement. AJR 1969; 107 : 776-786.
6. Hughes PM, Desai S, Mosley JG : Large Bowel Strictures in Retroperitoneal Fibrosis : Case report. Clinical Radiology 1993; 47 : 282-284.
7. Hunick DH, Chatson GP, Megibow AJ, Bosnick MA, Ryoff M : Retroperitoneal fibrosis presenting as colonic dysfunction : CT diagnosis. J Comput. Ass. Tom. 1988; 12 : 159-161.
8. Leffoll LD, White J, Mann M : Retroperitoneal fibrosis-two unusual cases. Arch. Surg 1964; 89 : 1070-1076.
9. Martino FB, Nuech R, Gesser TC : Retroperitoneal fibrosis and chronic periaortitis : a new hypothesis. Eur-Urol 1993; 23 (3) : 371-374.
10. McCarthy JG, Porter MR, Veenema R : Retroperitoneal fibrosis and large bowel obstruction. Ann Surg 1972; 176 : 199-204.
11. Mitchell RJ : Alimentary complications of non-malignant retroperitoneal fibrosis. BJS 1971; 58 : 254-256.
12. Mitckinson MJ, Withycomba JFR, Jones RA : The response of idiopathic retroperitoneal fibrosis to corticosteroid. BJ Urol 1971; 43 : 44-49.
13. Ochsner MG, Bronnon W, Pond HS, Goodlet JS : Medical therapy in idiopathic retroperitoneal fibrosis. J Urology 1975; 114 : 703-704.
14. Ormond JK : Bilateral ureteral obstruction due to envelopment and compression of an inflammatory retroperitoneal process. J Urology 1948; 59 : 1072.
15. Pollette EM, Polfette EC, Harrington RW : Sclerosing lipogranulomatosis : its several abdominal syndromes. Arch. Surg 1967; 94 : 803-810.
16. Ross J, Tinchler LF : Renal failure due to peri-ureteric fibrosis. BJS 1958; 46 : 58-62.
17. Vincenzi V, Coston Biedo FD, Andrea P, Fovretti F, De-Marchi G, Bosovi A : Fibrosi retroperitonealle idiopatica : efficacia della terapi associate chirurgica corticosteroidea. Minerva-Med 1993 Jun; 84 (6) : 351-354.
18. Webb AJ, Dowson-Edwards P : Non-malignant retroperitoneal fibrosis. BJS 1967; 54 : 508-518.