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Retroperitoneal Fibrosis with Duodenal Stenosis

Retroperitoneal fibrosis is a rare disease characterized by the formation of dense plaque of fibrous tissue covering the retroperitoneal structures. This disease is commonly presented as ureteral obstruction, but the involvement of duodenum is rare. We report a case of retroperitoneal fibrosis which was complicated with duodenal stenosis and was successfully treated with corticosteroids. A 58-yr-old man, who had history of aorto-iliac bypass graft due to arteriosclerosis obliterans with infrarenal aortic occlusion was admitted to the hospital with abdominal pain and a mass. Abdominal CT scan revealed the periaortic soft tissue mass encircling grafted aorta and stenosis of duodenal third portion. Retroperitoneal fibrosis with duodenal stenosis was diagnosed and prednisolone therapy was initiated. Follow-up CT scan showed that the patient responded to prednisolone therapy with eased pain, shrinking periaortic mass, and reduced duodenal stenosis.

Key Words : *Retroperitoneal Fibrosis; Duodenum; Duodenal Obstruction; Steroids; Prednisolone*

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INTRODUCTION

Retroperitoneal fibrosis is a rare disease characterized by the formation of dense plaque of fibrous tissue covering the retroperitoneal structures. The first description of retroperitoneal fibrosis was done by Albarran in 1905 in the French literature (1), followed by the first English description by Ormond in 1948 (2). Since these original descriptions, many cases have been reported. Retroperitoneal fibrosis is commonly presented as ureteral obstruction, but the involvement of gastrointestinal tract is rare. We report a case of retroperitoneal fibrosis complicated with duodenal stenosis and was successfully treated with corticosteroids.

CASE REPORT

A 58-yr-old man was admitted to Asan Medical Center with abdominal pain. Four years prior to admission, he had intermittent limb claudication and visited the hospital to evaluate the problem. At that time, physical examination revealed that his femoral pulses were not palpated, and arterial angiography showed complete occlusion of infrarenal abdominal aorta. Arteriosclerosis obliterans was diagnosed and aorto-bifemoral bypass graft was performed. The patient recovered uneventfully from surgery and was discharged. Three months before admission, he had abdominal pain that radiated to the back. The nature of pain was dull and aching and was aggravated after meal, but not accompanied with nausea or vomiting. During these 3 months, he lost 10 kg in weight. He had not taken any medication.

On physical examination, he appeared ill. The temperature was 36.3℃, the pulse was 80/min, the respirations were 24/min and the blood pressure was 140/92 mmHg. No rash or lymphadenopathy was found. The abdomen was soft, and scar from previous operation was noted at the midline. Just left lateral to the midline of abdomen, poorly localized hard mass with mild tenderness was palpated. The results of a rectal examination were normal, as was anal tone. There was no pedal edema, and the femoral pulses were normal.

The results of initial laboratory tests were as follows: leucocytes 13,500/μL, hemoglobin 12.3 g/dL, platelets 518,000/μL, erythrocyte sedimentation rate (ESR) 57 mm/hr, blood urea nitrogen 11 mg/dL, creatinine 1.1 mg/dL, total protein 7.4 g/dL, albumin 3.0 g/dL, aspartate aminotransferase 15 IU/L, alanine aminotransferase 7 IU/L, alkaline phosphatase 135 IU/L, total bilirubin 0.7 mg/dL, sodium 129 mEq/L, potassium 4.2 mEq/L, chloride 93 mEq/L, amylase 77 U/L, and lipase 35 U/L.

A plain abdominal radiograph showed moderate amount of stool in the right side of the colon and

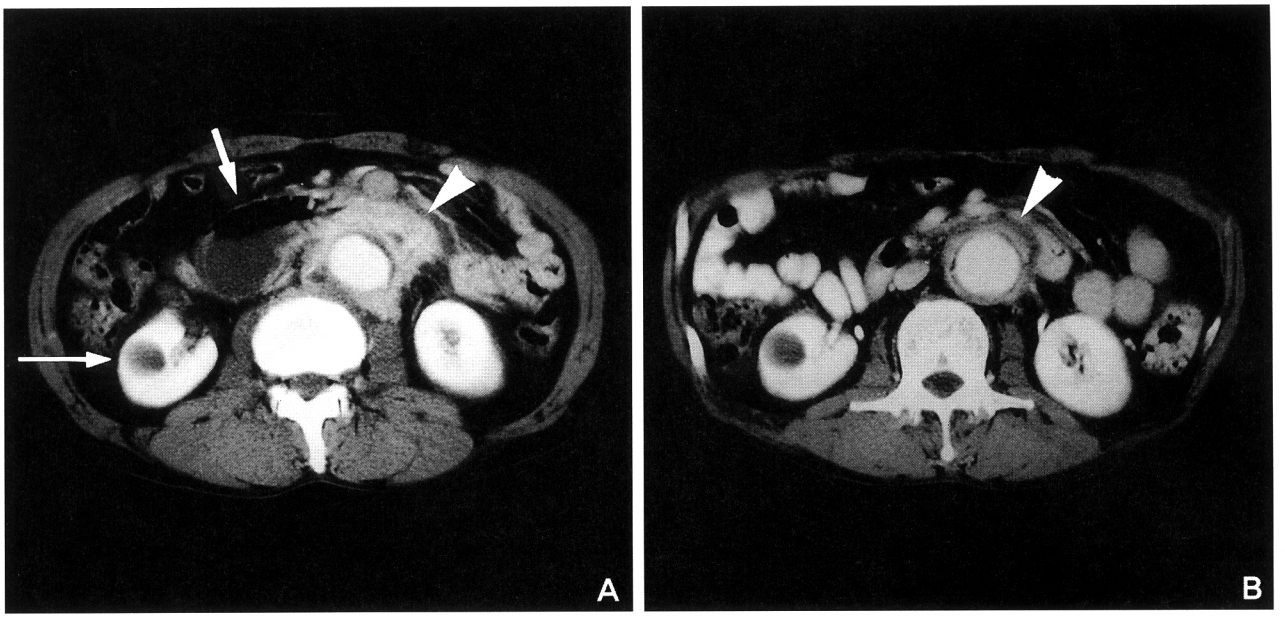

Fig. 1. Abdominal CT scan before and after treatment. **A:** Initial, pretreatment scan reveals a huge periaortic retroperitoneal mass (arrow head) and consequent duodenal stenosis (thick arrow) and incidental right renal cyst (thin arrow). **B:** After 3 weeks of prednisolone therapy, the mass has markedly decreased in size (arrow head) and duodenal stenosis was improved.

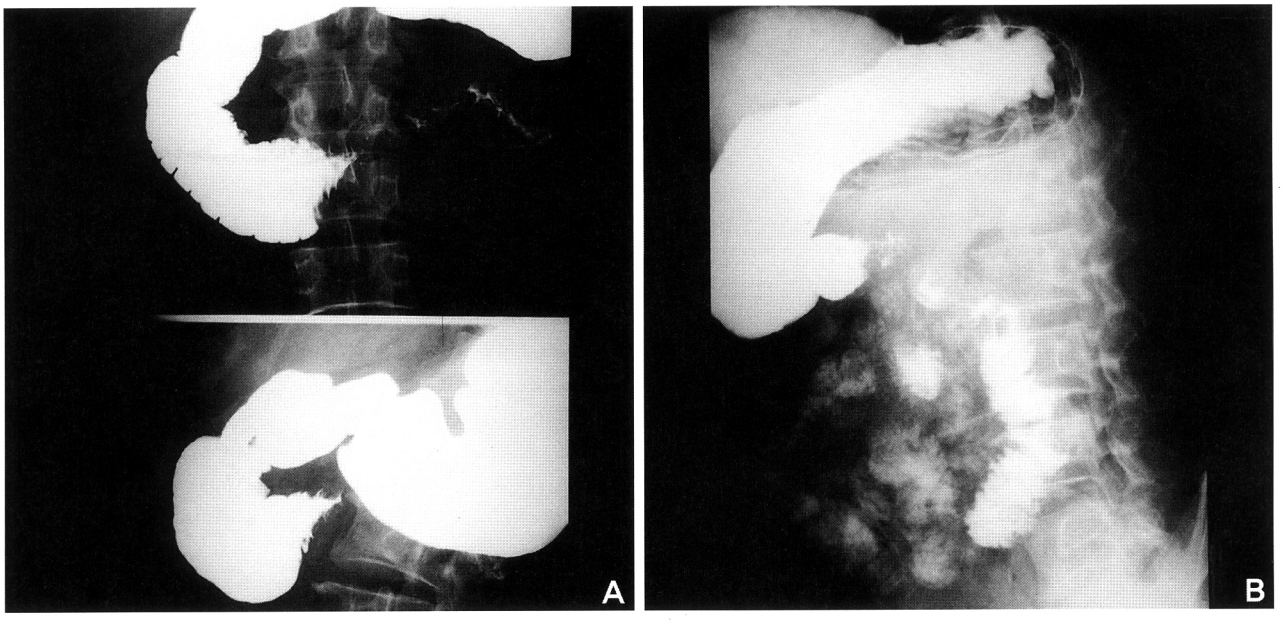

Fig. 2. Barium examination of upper gastrointestinal tract. **A:** Initial UGI series reveals stenosis of the duodenal third portion. **B:** Duodenal stenosis was improved after prednisolone therapy.

nonspecific gas pattern. Gastroduodenoscopic examination revealed chronic superficial gastritis but was otherwise normal. Abdominal CT scan demonstrated mass of soft tissue encircling the previously grafted aorta and stenosis of duodenal third portion (Fig. 1A). Barium examination of the upper gastrointestinal tract revealed the luminal narrowing of third and forth portion of duodenum and the dilatation of proximal duodenum, thus suggesting duodenal stenosis (Fig. 2A).

We diagnosed this case as duodenal stenosis due to retroperitoneal fibrosis, and prednisolone therapy was initiated at a dose of 1 mg/kg/day. The patient responded to prednisolone therapy with eased pain and decreased ESR to 9 mm/hr within 2 weeks. After 3 weeks of prednisolone therapy, follow-up CT scan and UGI series revealed the marked shrinkage of periaortic soft tissue mass

and the improvement of duodenal stenosis (Fig. 1B and 2B).

DISCUSSION

Retroperitoneal fibrosis is a rare disease characterized by the formation of dense plaque of fibrous tissue covering the retroperitoneal structures. The pathogenesis of retroperitoneal fibrosis remains unclear, although there is a high correlation with atherosclerotic disease of the aorta. Recent pathologic studies by Michinson and Parums et al. suggest that retroperitoneal fibrosis arises as an immune reaction to one or more of the components of atherosclerotic plaques (3-5).

Most cases of retroperitoneal fibrosis do not have identifiable causes or associated disease, but some are associated with drugs such as methysergide and ergot alkaloids and with malignancy, trauma, infection, radiation, and aneurysm of the abdominal aorta (5-7). The present case was considered as secondary retroperitoneal fibrosis because the patient developed retroperitoneal fibrosis after aortic bypass surgery. Surgical trauma was known as a cause of retroperitoneal fibrosis.

Grossly, retroperitoneal fibrosis appears as a dense, grayish white, fibrous mass surrounding the aorta. The fibrosis is generally localized to the retroperitoneum between the renal hilar and the sacral promontory and frequently extends laterally to entrap the ureters, resulting in variable degrees of hydronephrosis, which is the hallmark of the retroperitoneal fibrosis. It may also be extended into the thorax and cause mediastinal fibrosis. However, duodenal involvement is an extremely rare condition and has been reported in only 8 cases in the English literature (8, 9).

The pathology commonly reveals densely fibrotic areas of collagen with distinct areas of cellular inflammation (6, 10, 11). The inflammatory component consists predominantly of macrophages, lymphocytes, plasma cells, and occasional eosinophils, which are interspersed within a framework of fibroblast and collagen bundles. The areas of fibrosis are relatively avascular and acellular with scattered calcification admixed with the collagen bundles (6, 10, 11).

The symptoms of retroperitoneal fibrosis are vague and nonspecific. The most common symptoms are back, flank or abdominal pain, which is usually dull and poorly localized. Other common symptoms include weight loss, anorexia, nausea or vomiting, and malaise. Laboratory abnormalities may include azotemia, anemia, and an elevated ESR. Because of the nonspecific nature of the presenting symptoms, the diagnosis of retroperitoneal fibrosis is often delayed (6, 7, 11, 12).

In the past, laparotomy was necessary to make the diagnosis. However, there are inherent risks in taking deep biopsies from retroperitoneal tissues. Recently, CT scan has emerged as the modality of choice for the diagnosis and follow-up of patients with retroperitoneal fibrosis (12). The diagnosis is made by the demonstration of a periaortic soft tissue retroperitoneal mass by CT. Moreover, with improved CT scanning and MR imaging techniques, retroperitoneal neoplasms and retroperitoneal fibrosis can be differentiated in most cases (13-15). Therefore, an elevated ESR and CT findings have been considered sufficient even without histological proof because of the various risks associated with biopsy (8, 9, 16). This case was also diagnosed as retroperitoneal fibrosis without biopsy based on prior history of abdominal surgery, elevated ESR and CT finding.

Treatment of retroperitoneal fibrosis had been mainly surgical. However, Higgins et al. recommend non-operative management and there have been many cases of retroperitoneal fibrosis treated with corticosteroids alone without surgery (16). Non-operative management aims to arrest the chronic inflammation. The anti-inflammatory action of corticosteroids and their ability to inhibit fibrotic tissue maturation make them ideal for managing the chronic inflammation of retroperitoneal fibrosis. Although corticosteroids are effective in the active inflammatory stage of retroperitoneal fibrosis, they are less effective in the later acellular fibrotic stage of the disease. The dose of corticosteroids remains empirical. According to Higgins et al., the initial dose of prednisolone varied from 30 to 60 mg, depending on the seriousness of the patient's condition. Corticosteroids should be tapered gradually in accordance with the improvement of the patient's symptoms and with laboratory and radiologic findings.

Among the previously reported 8 cases of retroperitoneal fibrosis with duodenal involvement, only 3 cases were successfully treated with corticosteroids without surgery. The present case was also improved by corticosteroids therapy without surgical intervention.

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