*Case Report*

# Idiopathic Retroperitoneal Fibrosis in a Patient Suspected of Impending Rupture of the Abdominal Aortic Aneurysm

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**Retroperitoneal fibrosis (RPF) is a relatively rare disease which shows a periaortic mass in the retroperitoneal area and predisposes to an obstructive uropathy. We report a case of idiopathic RPF occurring in a patient who was suspected of impending rupture of abdominal aortic aneurysm. A 60-year-old male, with a 2-week history of abdominal pain, was transferred for evaluation of the periaortic mass. Computed tomographic (CT) scan revealed radiological findings such as leakage of contrast media from the aortic lumen and expansion of the periaortic mass. The patient underwent laparotomy, which revealed retroperitoneal fibrotic plaques in the absence of aortic aneurysm. The pathological findings of the biopsy specimen were consistent with idiopathic RPF. The patient received ureteral stent placement and was treated with steroid therapy. When a similar case is encountered, our recommendations are as follows: (i) Both CT scan and magnetic resonance (MR) imaging should be performed to determine whether the retroperitoneal mass is due to idiopathic or secondary RPF. If idiopathic RPF is suspected, the patient should receive primary steroid therapy. (ii) Retroperitoneal periaortic mass indicates a need for the assessment of obstructive uropathy. Early placement of the ureteral stent is necessary for urinary decompression and preservation of the renal function in patients with obstructive uropathy. (Ann Thorac Cardiovasc Surg 2008; 14: 55–59)**

**Key words:** retroperitoneal fibrosis, abdominal aortic aneurysm, impending rupture, obstructive uropathy

## Introduction

Retroperitoneal fibrosis (RPF) was first described in 1905 as a chronic, non-purulent disease of undetermined etiology exhibiting progressive fibrosis in the retroperitoneal area.[1)] RPF was established as a clinical entity by Ormond in 1948, but had been considered a relatively rare disease until its radiological features became evident.[2)] Afterwards many cases of RPF have been reported mainly in the urology area and the reported incidence of RPF is about one in five million people.[3,4)] Recent studies also suggest that idiopathic RPF fits into the broader pathologic heading of chronic periaortitis, which any surgeon carrying out abdominal surgery may encounter.[4)] Although cases of RPF have been rarely reported in the area of cardiovascular surgery, inflammatory aortic aneurysms, which are thought to be a subtype of RPF, have been often reported in the literature. We report a case of idiopathic RPF occurring in a patient who was suspected of impending rupture of abdominal aortic aneurysm and underwent lap-

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arotomy for a definitive diagnosis.

## Case Report

A 60-year-old male had complained of abdominal pain since early October and visited another hospital for evaluation of the gastrointestinal tract. He had taken a couple of antiplatelet agents due to previous minor cerebral infarction, but had been in a good functional capacity without any motor neuron disturbances. With the exception of a colon polyp measuring 7 mm in diameter, all tests for gastrointestinal tract were reported to be normal. The physician treated him with non-steroidal anti-inflammatory drugs for his abdominal pain. Computed tomographic (CT) scan showed an abnormal periaortic mass measuring 34.4×35.0 mm in diameter, which also exhibited radiological findings such as leakage of contrast media from the internal lumen of the abdominal aorta (Fig. 1). The patient was then transferred to our hospital, with a 2-week history of abdominal pain, for evaluation of the periaortic mass surrounding the abdominal aorta on October 19, 2006.

On physical examination, he was a well-developed man with a height of 163 cm and body weight of 68.7 kg. There was no anemia, jaundice, or disturbance of consciousness. He was afebrile with a blood pressure of 146/80 and a regular pulse of 80 beats/min. Although there had been a history of minor cerebral infarction, his functional capacity was good condition without any sensory or motor neuron disturbances. The chest X-ray and an electrocardiogram (ECG) were normal. Table 1 shows laboratory data on admission. With an exception of elevated C-reactive protein, other laboratory tests were within normal limits.

Angiogram of the abdominal aorta showed irregular walls but no enlargement of the aortic lumen (Fig. 1). An angiogram of the celiac artery and the superior mesenteric artery was normal. There was no significant stenosis in the coronary artery. Abdominal CT taken on October 20 revealed a more expanding periaortic mass measuring 37.1×36.4 mm in diameter. The patient, who was suspected of an impending rupture of the abdominal aortic aneurysm, underwent surgery on October 26. On laparotomy, there was a thickened, fibrotic plaque along the infrarenal abdominal aorta (Fig. 2). After the abdominal aorta and bilateral common iliac arteries were taped, a part of the periaortic tissue was incised to find the absence of aortic aneurysm (Fig. 2). Resected specimens were submitted for pathological examination.

Postoperative courses were uneventful without deterioration of the renal function. Pathological examination of the thickened tissue revealed a proliferation of fibroblasts and collagen fiber running into many directions, with a diffuse cell infiltration of the lymphocytes in main, and histiocytes and neutrocytes (Fig. 3). These pathological findings were consistent with idiopathic RPF. Microscopic appearance of the fibrotic plaque also involved retroperitoneal nerve fibers, ganglions, and lymphnodes, and suggested that the fibrotic plaque would entrap the ureter leading to the obstructive uropathy. Indeed, postoperative intravenous pyelogram and venacavography showed left hydronephrosis probably due to the left ureter obstruction, and obstruction of the left common iliac vein (Fig. 4). There were many collaterals along the obstructed common iliac vein, thus the patient did not have lower extremity edema and did not receive surgery for venous reconstruction.

The patient was transferred to the urology department of other hospital and received the ureteral stent placement for urinary decompression. He has been treated with steroid therapy, and CT scan revealed a reduction of the retroperitoneal thickened mass surrounding the abdominal aorta.

## Discussion

RPF is a rare cause of obstructive uropathy which may also lead to venacaval or duodenal obstruction. Recent advances in radiology and research into the pathophysiology have improved our diagnostic ability and our understanding in this disease.[4] Idiopathic RPF has been reported to comprise about 60 to 70% of RPF, and secondary RPF due to drug use or neoplasms in the retroperitoneal area ranges from 30 to 40%.[4,5] Recent studies suggest that many cases of RPF are due to an autoallergic reaction to lipid material in the atheromatous aorta.[6,7] The patient in this case had not had retroperitoneal neoplasms nor taken drugs related to the development of secondary RPF, thus idiopathic RPF was the most reasonable diagnosis accounting for his symptoms and pathologic findings.

The clinical entity of RPF also includes an inflammatory aortic aneurysm (IAAA), reported by Walker in 1972, which comprises about 4–5% of abdominal aortic aneurysms.[8–10] IAAA has significant clinical features characterized by a specific radiological finding showing three different layers including a thickened wall lying outside the lucent rim of the intima and subintima, which is known

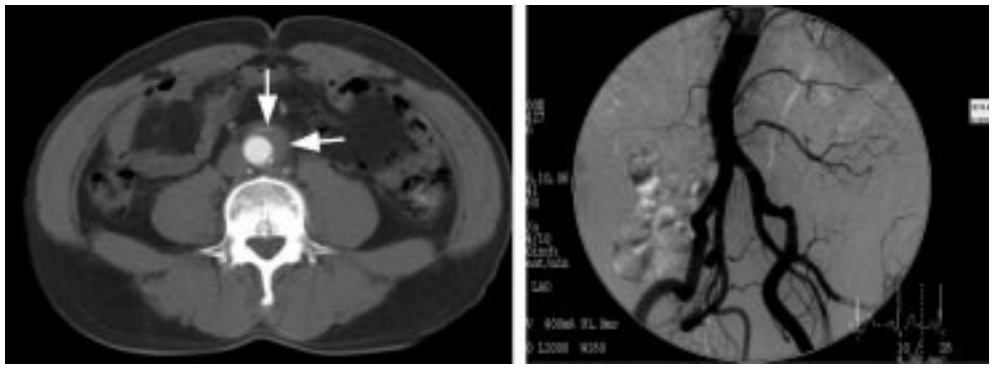

**Fig. 1.** Preoperative computed tomography (CT) scan (left) and angiogram (right) of the abdominal aorta.

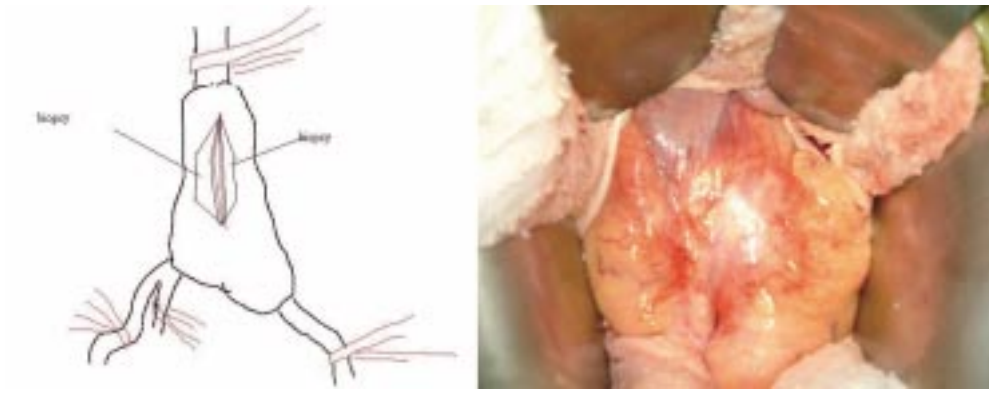

**Fig. 2.** Intraoperative findings: schema (left) and photograph (right).
There was a thickened, fibrotic plaque along the infrarenal abdominal aorta. After taping the abdominal aorta and bilateral common iliac arteries, a part of the periaortic tissue was incised to find the absence of pseudoaneurysm.

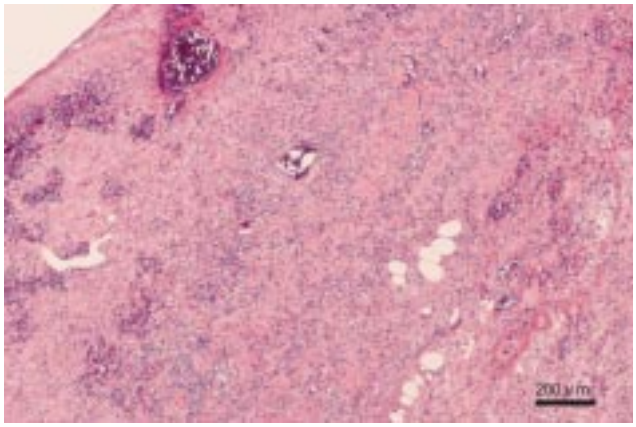

**Fig. 3.** A histological photograph by hematoxylin and eosin staining with a magnification of ×100.
There was a proliferation of fibroblasts and collagen fiber running in many directions, with a diffuse cell infiltration of the lymphocytes in main, and histiocytes and neutrocytes in the biopsy specimen.

as "mantle core sign."[11] The radiological findings of mantle core sign are usually limited to the periaortic tissue and rarely extend over the psoas line. A CT scan of the present case revealed leakage of the contrast media from the aortic lumen into the thickened periaortic tissue. However, operative findings revealed no connection between the aortic lumen and the lesion seemed like periaortic hematoma. There was no strong suspicion of IAAA preoperatively due to the absence of typical mantle core sign in radiological findings.

RPF is an uncommon disease that surgeons may encounter in patients complaining of abdominal pain who are suspected of pseudoaneurysm or impending rupture of the aortic aneurysm. In urology and radiology, however, the distinction between idiopathic RPF and secondary RPF due to malignant neoplasms has been focuse in some literature.[12,13] Magnetic resonance (MR) imaging has been reported to be beneficial in differentiating be-

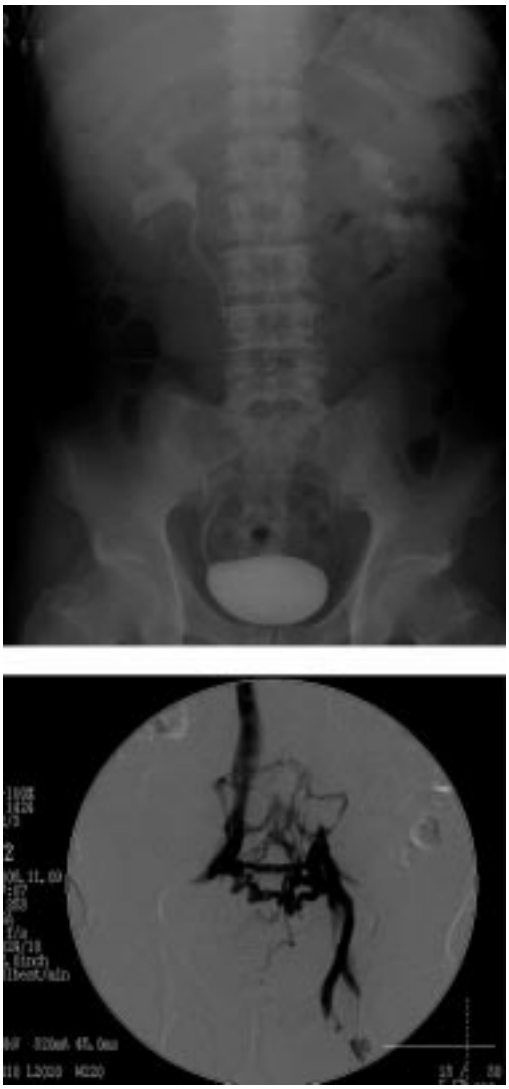

**Fig. 4.** The intravenous pyelogram (upper) and venacavography (lower).
The intravenous pyelogram and venacavography showed left hydronephrosis and obstruction of the left common iliac vein. Since many collaterals along the obstructed common iliac vein were present, the patient did not have lower extremity edema.

**Table 1. Laboratory data on admission**

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|---|---|---|---|---|---|
| WBC | 8,700 | (/μL) | CRP | 6.02 | (mg/dL) |
| RBC | 445×10$^4$ | (/μL) | BUN | 11.2 | (mg/dL) |
| Hb | 13.4 | (g/dL) | Cr | 0.9 | (mg/dL) |
| Plt | 26.8×10$^4$ | (/μL) | Na | 143 | (mEq/L) |
| TP | 7.6 | (g/dL) | K | 3.8 | (mEq/L) |
| GOT | 12 | (IU/L) | Cl | 103 | (mEq/L) |
| GPT | 10 | (IU/L) | Ca | 8.6 | (mg/dL) |
| LDH | 189 | (IU/L) | T-chol | 170 | (mg/dL) |
| T-Bil | 0.5 | (mg/dL) | TG | 62 | (mg/dL) |

WBC, white blood cells; RBC, red blood cells; Hb, hemoglobin; Plt, platelet counts; TP, total protein; GOT, glutamic oxaloacetic transaminase; GPT, glutamic pyruvic transaminase; LDH, lactic dehydrogenase; T-Bil, total bilirubin; CRP, C-reactive protein; BUN, blood uria nitrogen; Cr, creatinin; Na, sodium; K, potassium; Cl, chloride; Ca, calcium; T-chol, total cholesterol; TG, triglyceride.

tween malignant and nonmalignant RPF. Biopsy of the fibrotic tissue is required to make the definitive diagnosis, but Higgins and associates advocate a 1-to-2-week steroid therapy as a trial treatment for a patient suspected of RPF without performing a biopsy. If the patient shows a reduction in the size of the retroperitoneal mass, the patient can be diagnosed as having an idiopathic RPF. In this condition, they advocate that continuing treatment with steroid therapy is indicated without surgical biopsy.[14] The present case also shows a reduction in the size of thickened periaortic tissue after steroid therapy, which was started after definitive diagnosis by surgical biopsy. As a result, primary steroid therapy without surgical biopsy could have had the same outcome for the present case. However, it was difficult to perform only steroid therapy for a patient who was suspected of impending rupture of the abdominal aorta without a strong suspicion of IAAA or RPF. When similar cases are encountered, our recommendations based on the present case experience and review of the literature are as follows: (i) Both CT scan and MR imaging should be performed to determine whether the retroperitoneal mass is due to idiopathic or secondary RPF, by referring a signal intensity of T2 enhanced imaging. If idiopathic RPF is suspected, the patient should receive primary steroid therapy. (ii) Retroperitoneal periaortic mass indicates a need for the assessment of obstructive uropathy. Intravenous pyelogram should be performed early even in the absence of renal dysfunction. Early placement of the ureteral stent is necessary for urinary decompression and preservation of the renal function in patients with obstructive uropathy.

In summary, we describe a case of idiopathic RPF occurring in a patient who was suspected of impending rupture of the abdominal aortic aneurysm and underwent laparotomy. Usually a cardiovascular surgeon rarely encounters idiopathic RPF, which belongs to a family of inflammatory diseases presenting with adjacent inflammation in the periaortic tissue. Some considerations are given to the process of diagnosis and treatment of RPF to avoid clinical pitfalls.

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