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IDIOPATHIC RETROPERITONEAL FIBROSIS WITH LARGE VESSEL THROMBOSIS

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A 53-year-old female was hospitalized for evaluation of swelling in the bilateral lower extremities. A computed tomography (CT) scan of the abdomen revealed bilateral hydronephrosis and features consistent with retroperitoneal fibrosis. Transfemoral venography and magnetic resonance angiography (MRA) showed thrombosis of both the left common iliac vein and inferior vena cava, and filling of numerous collateral veins in the retroperitoneal area. A diagnosis of idiopathic retroperitoneal fibrosis with central venous thrombosis was made. Ureteral stenting, medication with corticosteroids and subsequent warfarin were started, resulting in marked improvement of renal function and the lower extremities. Diagnosis and follow-up of deep venous thrombosis can be aided by MRA. Administration of steroids with anticoagulation was considered to be successful in the case presenting with deep venous thrombosis caused by retroperitoneal fibrosis.

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Key words: Retroperitoneal fibrosis, Thrombosis, MR angiography, Anticoagulation

INTRODUCTION

Retroperitoneal fibrosis is a disease characterized by development of a plaque of fibrous tissue in the retroperitoneum and related tissue planes, with associated obstructive effects depending on the size and extent of the lesion¹⁾ Knowledge of the condition has increased due to better understanding of its pathophysiology, mainly as an obstructive uropathy²⁾ Although vascular occlusion has been reported to occur from retroperitoneal fibrosis³⁾, the extensive vascular involvement that can occur in the venous system has been relatively little emphasized.

Here we report a case of both iliac and caval venous thrombosis due to idiopathic retroperitoneal fibrosis (IRF), which was confirmed by conventional radiographic modalities such as MRA. We describe the presentation, diagnostic considerations, and therapy of a patient with deep venous thrombosis caused by IRF.

CASE REPORT

A 53-year-old female was admitted to the internal medicine department because of oliguria and swelling in the bilateral lower extremities for a few days. She had had a medical checkup a month before the onset, but the investigation had been negative apart from poor renal function.

Family and past histories were noncontributory. Examinations of the chest and abdomen were negative. Admission laboratory tests included a blood urea nitrogen (BUN) level of 68.3 mg/dl and a serum creatinine level of 12.2 mg/dl. The levels of C-reactive protein (CRP) and lactate dehydrogenase (LDH) were 1.1 mg/dl and 675 IU/l, respectively. The serum sodium, potassium, chloride, and

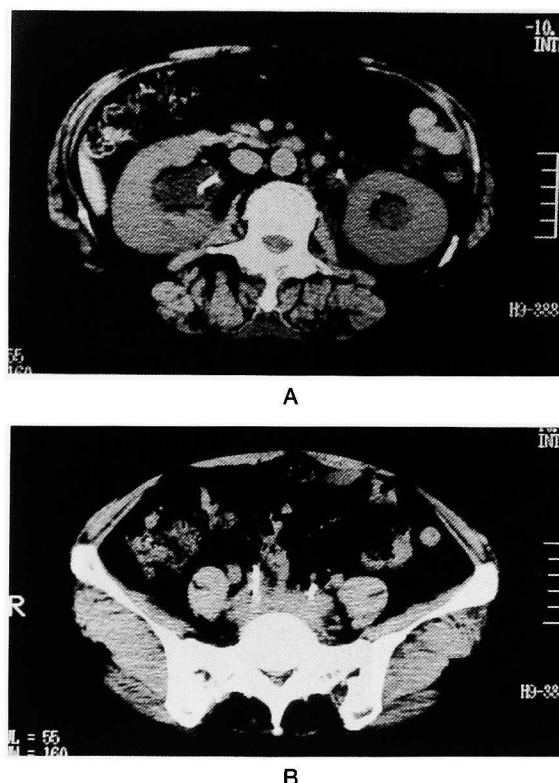


Fig. 1. Abdominal CT scans (two days after bilateral double-J catheters were inserted) showing bilateral hydronephrosis (A) and soft-tissue infiltration around the ureters in the sacral region (B), consistent with retroperitoneal fibrosis.

bicarbonate levels were normal. Renal ultrasound revealed bilateral hydronephrosis with normal-sized kidneys. A non-contrast CT scan demonstrated an extensive soft tissue mass with encasement of the common iliac arteries at the S2 level that extended

into the pelvis (Fig. 1B).

The immediated management of the patient was bilateral ureteral stent placement that lowered the serum creatinine level to 2.7 mg/dl. At this time, the patient suffered from anuria and subsequent CT scan revealed bilateral hydronephrosis (Fig. 1A),

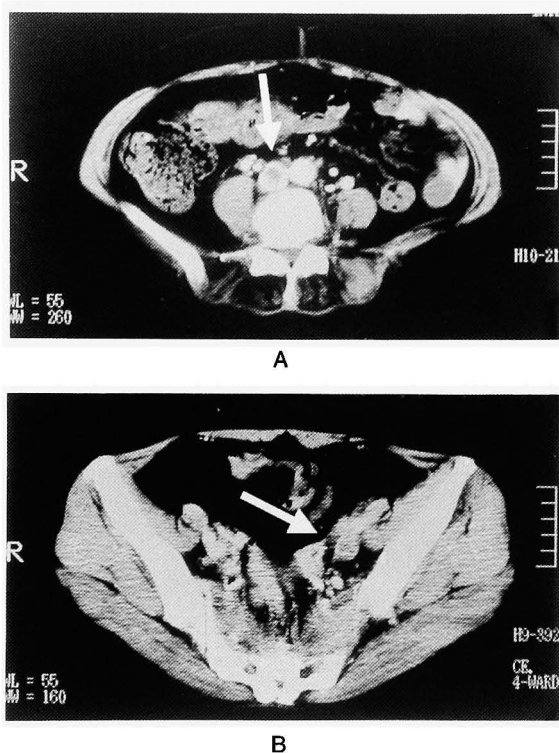


Fig. 2. CT scans of the abdomen. A low-attenuation thrombus is present in the inferior vena cava (A) and in the left common iliac vein (B).

necessitating a left nephrostomy, which brought the serum creatinine down to 1.0 mg/dl over the next week. Antegrade pyelography showed a left ureteral stricture at the level of L4/5.

The left leg edema still existed after the urine drainage. Contrast CT scanning of the abdomen suggested ilio caval venous thrombosis (Fig. 2A, B), which was confirmed by a magnetic resonance angiogram (MRA) and bilateral ascending venogram (Fig. 3A, B). A pulmonary scintigram was negative for pulmonary infarction. Therefore, the diagnosis of ilio caval thrombosis secondary to IRF was made.

On the basis of the clinical setting, CT, and angiographic findings, the patient was started on prednisolone and subsequent warfarin at doses of 1 mg/kg per day and 3 mg/day, respectively. In view of the patient's clinical condition, an inferior vena cava (IVC) filter was not placed.

After one month of medication, an abdominal CT scan showed mild resolution of the inflammatory mass and disappearance of the thrombus. MRA revealed recanalization of the left common iliac vein (Fig. 4). Antegrade pyelography showed smooth urine passage through the left ureter. At this time, the left nephrostomy was removed.

The steroids were tapered off gradually, resulting in suspension with a dose of 5 mg/day one year later. Warfarin was regulated according to the value of prothrombin time. Six months later, the ureteral stents were removed, and additional excretory urography showed good urine passage. CT scan revealed significant improvement of the retroperitoneal mass (Fig. 5). She remains well and asymptomatic at 4 years of follow-up, with stable

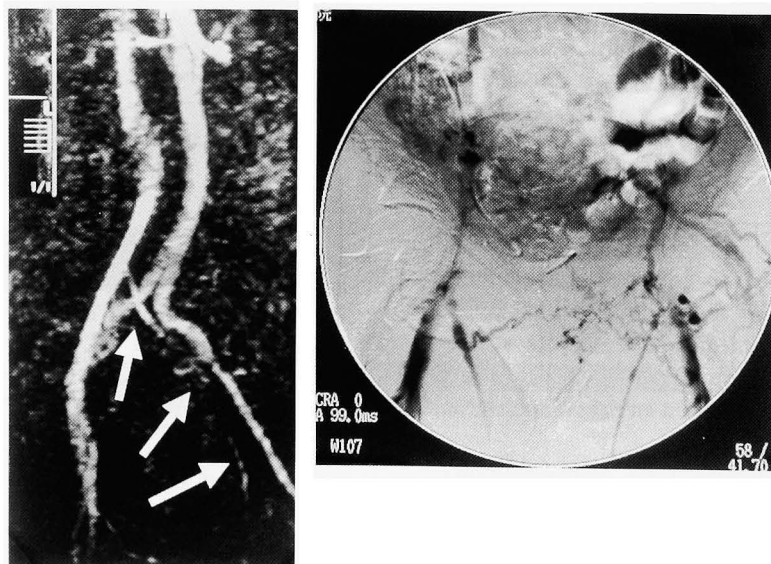


Fig. 3. A: Maximum intensity projection of subtracted MRA depicting deep vein thrombosis of the left iliac and femoral veins (arrowheads) as a signal void due to the absence of blood flow. B: Transfemoral venogram. Note filling of many collaterals in the retroperitoneal space.

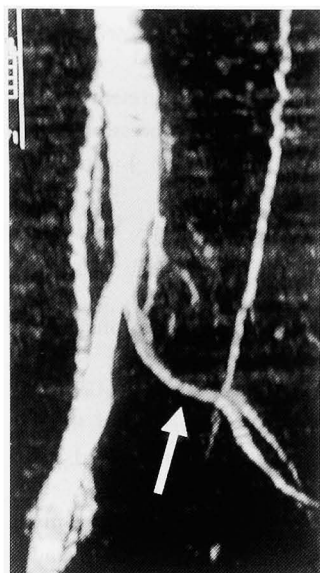


Fig. 4. Maximum intensity projection of subtracted MR venography after one month of medication showing recovery of vascularity of the left iliac and femoral veins (arrowhead). Mild venous stenosis remains without thrombus.

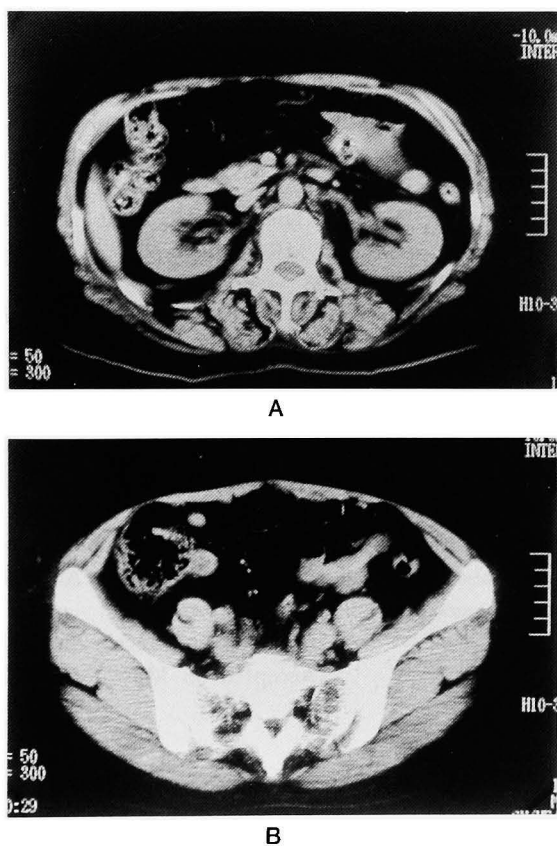


Fig. 5. CT scans obtained six months after the treatment demonstrates disappearance of hydronephrosis (A) and marked decrease in size of mass anterior to sacrum (B).

renal function and without edema.

DISCUSSION

Retroperitoneal fibrosis was first described by Albarran⁴⁾ in 1905 and established as a clinical entity by Ormond⁵⁾ in 1948. IRF represents a nonspecific, inflammatory reaction that involves various retroperitoneal structures, including the ureter and associated vascular structures^{1,2)}. The pathogenesis of IRF is unclear. This process is a chronic inflammatory response to a number of possible inciting factors, including infections, tumors, and atherosclerosis, and systemic processes such as vasculitis, lupus, and other autoallergic reactions⁶⁾. The reaction to advanced inflammatory atherosclerosis with IRF has been studied by a number of investigators⁷⁾.

Urinary findings have most frequently been reported as oliguria and microscopic hematuria⁸⁾. Other less common clinical presentations of IRF include lower-extremity edema and even thrombophlebitis from fibrotic impingement of the inferior vena cava, and claudication from arterial compromise⁹⁾. Our patient had some typical features of IRF, including oliguria, and central venous thrombosis.

The diagnosis of IRF is primarily made by performing imaging studies, as the history and laboratory findings are somewhat nonspecific in this uncommon disease. Although excretory urography and retrograde urography were the primary diagnostic modalities for this condition, CT has now supplanted other imaging modalities as the radiographic examination of choice for patients with suspected IRF¹⁰⁾. IRF on magnetic resonance imaging (MRI) is reported to be a variably shaped low intensity mass similar to the adjacent psoas muscle on T1- and T2-weighted images¹¹⁾. Although the most common clinical picture is obstruction of both ureters, the fibrosis sometimes envelops the aorta and iliac arteries, leading to claudication and gangrene, or it may involve and obstruct the inferior vena cava and iliac veins^{1,8-10)}, as seen in our case. The application of MRI and MRA to the IVC is obvious¹²⁾. Although some authors proposed that MR imaging provided more comprehensive information than catheter venography on central venous anatomy and disorders¹³⁾, a few authors reported the usefulness of the application to IRF according to the degree of central venous occlusion¹⁴⁾. We obtained an excellent correlation between the findings of venous obstruction and occlusion due to IRF by contrast venography and MRA. This suggested that MRA is an accurate, graphic, and noninvasive technique for evaluation of central venous thrombosis caused by IRF.

Corticosteroids are most useful in the active

inflammatory phase of the disease and may be used either as an adjunct to surgical therapy or alone in patients who are at poor surgical risk^{1,2,6,7,9,11}). The management of our patient with deep vein thrombosis due to IRF was mainly aimed at prevention of early and late complications of venous thrombosis—that is, prevention of pulmonary embolism and restoration of blood flow through a thrombosed vein with preservation of venous valve function. Anticoagulant therapy is the mainstay for acute venous thromboembolism¹⁵). Compared with pulmonary embolisms, deep vein thrombosis can be difficult to diagnose, and alone it only very rarely causes death¹⁶). Warfarin therapy is highly effective and is preferred in most patients with venous thrombosis^{15,16}). In the absence of pulmonary embolism and recurrent venous thrombosis, we administered warfarin to our patient, resulting in recanalization of the common iliac vein and the disappearance of the venous thrombus. Although thrombolytic therapy may benefit selected patients with acute massive venous thrombosis, it is considered that patients with established venous thrombosis, such as in our case, require long-term anticoagulant therapy to prevent recurrent disease.

CONCLUSION

In summary, IRF is an uncommon disorder that often presents in a subtle manner. The case described was possibly an unusual presentation of central venous thrombosis combined with IRF. MRA provides a non-invasive modality for evaluation of central venous occlusion caused by IRF. In addition, retroperitoneal fibrosis should be considered as a possible cause of deep venous thrombosis or chronic swelling of the lower extremities, and can be treated by administration of steroids combined with an oral anticoagulant.

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和文抄録

下大静脈および腸骨静脈血栓を伴った後腹膜線維症の1例

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田沼 康，横尾 彰文

53歳，女性。両下肢の腫脹と乏尿を主訴に当院受診，CT 上両側水腎症と両側腸骨動脈を取り囲む辺縁不整な軟部組織陰影を認めた。腎後性腎不全に対し両側ダブルJカテーテル留置，造影CTにて下大静脈および左腸骨静脈内に血栓を認め，また下大静脈造影およびMRアンジオグラフィー（MRA）にて左腸骨静脈閉塞を認めた。中心静脈血栓を伴う特発性後腹膜線維症と診断し，経口よりプレドニゾンおよびワー

ファリン投与開始した。投与1カ月後のCTおよびMRAで血栓の消失を認め，水腎症は改善，再発を認めていない。中心静脈血栓を伴う特発性後腹膜線維症に対してMRAは経時的評価に有用な画像検査であり，ステロイドおよび抗凝固薬投与は保存的治療の第一選択と考えられた。

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