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CASE REPORT

Vasculo-Behçet's disease with non-traumatic subcapsular hematoma of the kidney and aneurysmal dilatations of the celiac and superior mesenteric arteries

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Abstract We report a patient with vasculo-Behçet's disease treated successfully with a high dose of prednisolone. In 2002, the patient was diagnosed with vasculo-Behçet's disease. He was admitted to our hospital because of sudden-onset right lower back pain in June 2006. Upon admission, abdominal angiography revealed aneurysmal dilatations of the celiac and superior mesenteric arteries. He was treated promptly with high-dose prednisolone, after which the aneurysms displayed no further enlargement. As we believe this case to be quite rare, we report this case with a literature review in support of this characterization.

Keywords Arterial aneurysm · Behçet's disease · Spontaneous hematoma · Subcapsular hematoma

Introduction

Behçet's disease, a systemic inflammatory disease of unknown origin, is characterized by recurrent oral aphthous ulcers, skin disorders, external genital ulcerations, and eye diseases. Vasculo-Behçet's disease is a special sub-type of Behçet's disease that exhibits large, medium, or small arterial and/or venous abnormalities, with clinical symptoms varying according to the type of disease, i.e., venous occlusion, arterial occlusion, or aneurysmal change, as well as variation with the affected site.

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We report a patient with vasculo-Behçet's disease presenting with multiple vascular lesions. The patient developed a non-traumatic subcapsular hematoma of the right kidney and aneurysmal dilatations of the celiac and superior mesenteric arteries, which were alleviated by treatment with high-dose prednisolone (PSL). We also review the relevant clinical documents.

Case report

A 24-year-old man was admitted to our hospital because of sudden-onset right lower back pain in June 2006. In November 1999, he had experienced pain and swelling of the left lower limb and been diagnosed as having deep venous thrombosis in the left anterior tibial artery. He received warfarin treatment and the symptoms subsided. In April 2000, subcutaneous bleeding in the patient's right thigh was observed and the warfarin was discontinued. In March 2002, he underwent a thorough checkup for thrombosis. Neither genital ulcer nor eye disease was evident, but recurrent oral aphthous ulcers and polyarthritis were present and the HLA-B51 type was positive. Based on these findings, a diagnosis of incomplete-Behçet's disease (vasculo-Behçet's disease) was made. In June, however, thrombosis of the left femoral vein and of the right popliteal artery were detected, and the warfarin 2 mg/day was reinitiated.

In January 2004, the swelling and the pain around both the left and right ankles worsened and was diagnosed as an exacerbation of the Behçet's disease. Treatment with prednisolone (PSL) was initiated. The dosage of PSL was increased up to 20 mg/day and then tapered to 12 mg/day by the 20th of May, 2006. Warfarin treatment maintained the prothrombin time-INR at 1.5–1.8 throughout this

period. The patient was admitted to our hospital in June 2006 because of a sudden onset of severe right lower back pain.

Upon admission, physical findings revealed a temperature of 36.8°C, blood pressure of 140/72 mmHg and a pulse rate that was 80 beats/min and regular. There were no visible signs of hemorrhage on the skin, no marked abdominal pain and no gross hematuria noted. Neither recurrent oral aphthous ulcer nor polyarthritis was observed.

Laboratory studies revealed the white blood cell count was 10,680/μl, hemoglobin 15.4 g/dl, and erythrocyte sedimentation rate (ESR) 13 mm/h. The serum concentration of lactic dehydrogenase (LDH) was 600 IU/l, creatinin was 1.12 mg/dl and C-reactive protein (CRP) 2.57 mg/dl. As for the blood coagulation test, the prothrombin time-INR was 1.59, but the activated partial thromboplastin time, fibrin degradation product, and D-dimer were within normal limits. Urinalysis identified proteinuria and microscopic hematuria. Renal ultrasound (U/S) and abdominal computed tomography (CT) showed a subcapsular hematoma of the right kidney (Fig. 1a, b). Vascular U/S also showed occlusions of the left anterior tibial artery and the right popliteal artery (Fig. 2).

Anticoagulant therapy was discontinued in accordance with the diagnosis of non-traumatic subcapsular hematoma of the right kidney. During the course of treatment in the hospital, serial U/S and CT revealed that the right renal hematoma was shrinking with the conservative treatment (Fig. 1c).

On the 32nd day of hospitalization, abdominal angiography was performed. Renal artery imaging showed the superior branch of the right renal artery was irregular and narrowed (Fig. 3a). Part of the apical segment of the kidney did not appear on the angiography because it was fed from the apical segment artery arising from the aorta. Abdominal artery imaging revealed aneurysmal dilatations of celiac and superior mesenteric arteries (Fig. 3b, c). No abnormal finding was observed in the common hepatic artery or the bifurcation of aorta. Taken together with the finding of non-traumatic renal hematoma, the diagnosis was made of an exacerbation of vasculo-Behçet's disease.

As shown in the chart of Fig. 4, although the elevated CRP had been decreasing, the treatment with high dose PSL (60 mg/day) was initiated. The levels of CRP, ESR, LDH and creatinine were gradually reduced to the normal range and the aneurysmal dilatation of the superior mesenteric artery did not progress. The dosage of PSL was tapered in small steps and after 3 months was reduced to 40 mg/day. Subsequently there has been no episode of any new vascular lesion.

Discussion

Lie et al. [1] investigated 728 cases of vasculo-Behçet's disease and reported that 49 patients had arterial abnormalities, 181 venous abnormalities and 498 both arterial and venous abnormalities. In our patient, CT revealed a

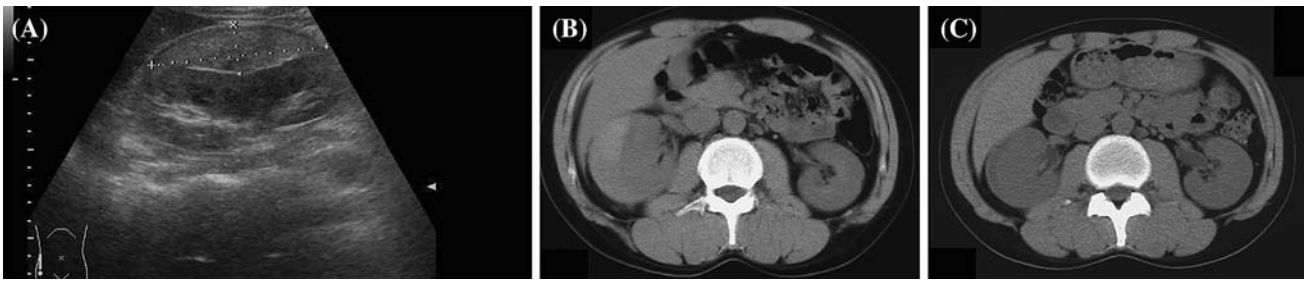

Fig. 1 U/S of the right kidney showing a subcapsular hematoma (**a**); CT section of the kidney showing the subcapsular hematoma (**b**); CT obtained 2 months after admission shows a decreased hematoma size (**c**)

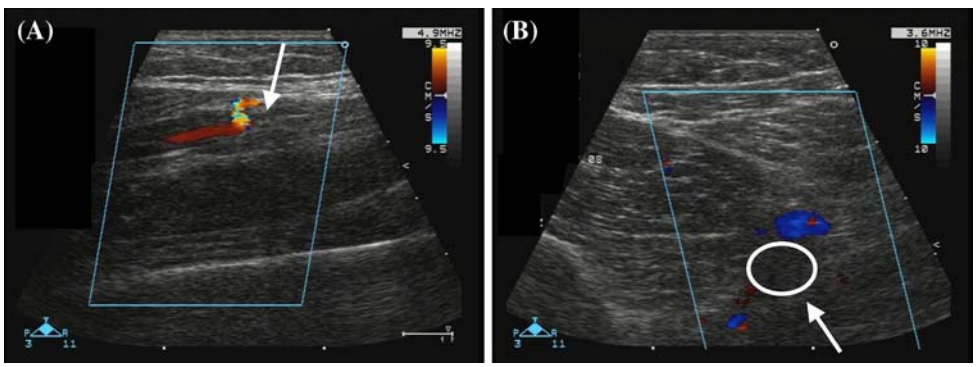

Fig. 2 U/S showing occlusions of the left anterior tibial artery (**a**, *arrow*) and the right popliteal artery (**b**, *arrow*)

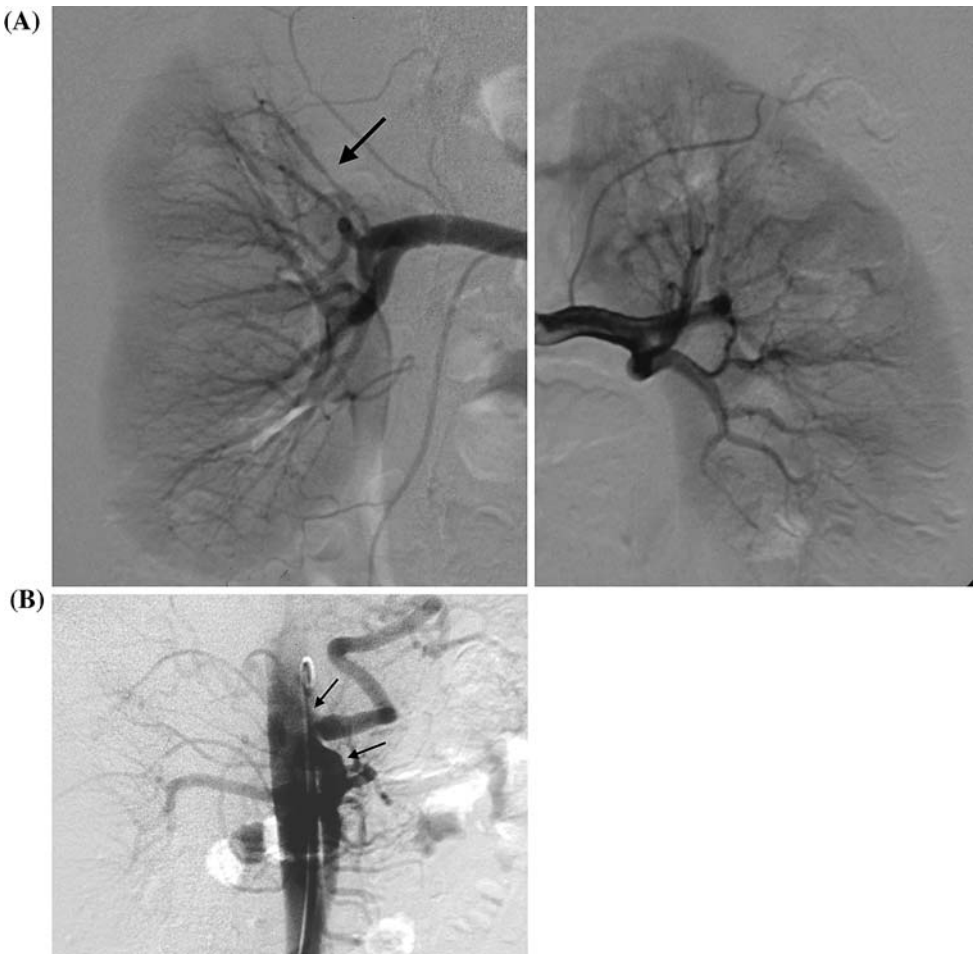

Fig. 3 Abdominal angiography showing the superior branch of the right renal artery was irregular and narrowing (**a**, *arrow*) and also the aneurysmal dilatations of the celiac and superior mesenteric arteries on the 32nd day of hospitalization. *Arrows* indicate the main lesions in the arteries (**b**)

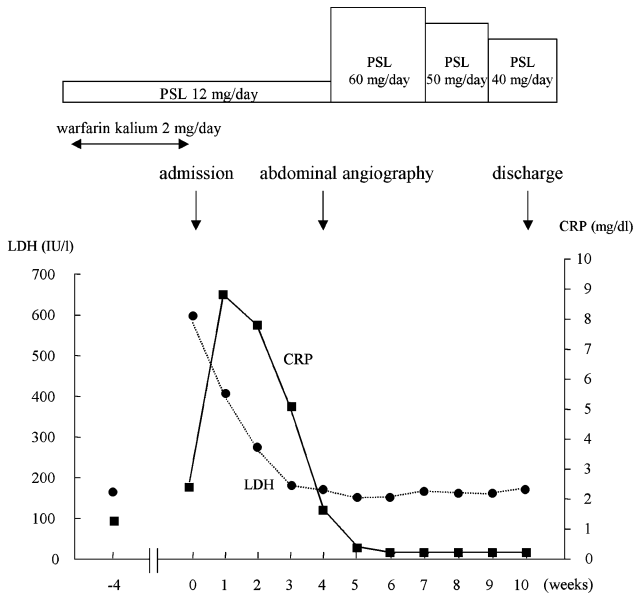

Fig. 4 The levels of CRP and LDH and the PSL dose during hospitalization

subcapsular hematoma of the right kidney and abdominal angiography uncovered aneurysmal dilatations of the celiac and superior mesenteric arteries. U/S also showed an occlusion of the left anterior tibial artery, the right popliteal artery, and the bilateral superficial femoral veins. These are all characteristic findings of vasculo-Behçet's disease.

In the paper mentioned above, it was also reported that aneurysms make up approximately 60% of the arterial abnormalities associated with vasculo-Behçet's disease and that arterial occlusions approximately 30%, and there are also some cases presenting with multiple aneurysms in the paper. The aorta is the most susceptible site to aneurysm, followed by the pulmonary artery, femoral artery, subclavian artery, cervical artery and popliteal artery. Murakami et al. [2] reported that they investigated 97 cases of vasculo-Behçet's disease and found no patient who developed abnormalities in the small renal, celiac or superior mesenteric arteries. To the best of our knowledge, at least six cases of vasculo-Behçet's disease, including our case, have been reported with the complication of renal artery abnormalities [3–7]. Moreover, including our case, only five cases of vasculo-Behçet's disease have been

reported to be complicated with abnormalities in the celiac and superior mesenteric arteries [4, 8–10]. Therefore, we assume this to be a very rare case.

As vasculo-Behçet's disease associated with critical organ involvement is thought to have a poor prognosis, corticosteroids in combination with an immunosuppressant are recommended [11, 12]. There is a report showing that the combination works effectively when it is initiated at an early stage [13]. In our patient, PSL therapy was initiated because there was a risk of a digestive system disorder, which is one of the complications of aneurysmal dilatations in the celiac and superior mesenteric arteries.

Upon admission, our patient had a sudden-onset right lower back pain and was diagnosed with non-traumatic subcapsular hematoma of the right kidney. Considering the various abnormal vascular findings observed in this patient, we concluded that the exacerbation of the present disease in addition to anticoagulation by warfarin might have induced this rare condition. Zhang et al. [14] reported that in 165 cases of non-traumatic renal subcapsular bleeding, 101 patients had developed it from a tumor and 20 from vasculitis, including 12 cases of polyarteritis nodosa. There was no Behçet's disease in their report. These patients had the same symptoms as our patient (lower back pain, abdominal pain, and hematuria) and some of them suffered shock. A literature search found that there have been no reports of Behçet's disease with non-traumatic subcapsular hematoma of the kidney.

Although a considerable number of patients with Behçet's disease have developed venous disease and received anticoagulant therapy, few have complained of non-traumatic lower back pain. However, if a patient does manifest this symptom, a subcapsular hematoma of the kidney should be considered as a possible cause. A patient with vasculo-Behçet's disease may present with multiple arterial abnormalities with diverse clinical features, so early identification and treatment are critical for proper management and a good prognosis.

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