Rupture of Renal Artery in a Patient with Behçet's Disease

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The vascular involvement is seen in 8% to 24% of patients with Behçet's syndrome. Arterial lesions are less frequently observed and the involvement of renal artery is very rare. Indeed, there is only one case report of ruptured renal aneurysm due to Behçet's disease. We report a case of renal artery rupture, which was likely caused by Behçet's disease. Even though we cannot completely rule out the other causes of renal artery rupture with this patient, the rupture was likely due to vascular involvement of Behçet's disease. Dermatologists always should consider the possibility of major vessel involvement when they treat patients with Behçet's disease, even if the incidence is very low and the patient may not complain of any systemic symptoms.

Introduction

Behçet's disease was first defined by Hulusi Behçet's[1] in 1937 as a multisystemic disease with the triad of recurrent aphthous stomatitis, genital ulceration and relapsing iritis. Its etiology remains obscure. It is accepted that the pathologic process of Behçet's disease is an immunologic vasculitis[2].

Among vascular lesions, the venous system is the major affected site[2-5]. Arterial lesions, such as aneurysms or occlusions, are rarely reported, but sometimes adversely affects the course of the disease. Aneurysm formation is known to be life-threatening because of the risk of rupture. (Ann Dermatol 14(2) ; 98-101, 2002).

Key Words : Behçet's disease, Aneurysm, Renal artery

CASE REPORT

A 33-year-old male patient was admitted to our hospital because of a sudden onset of left flank pain and syncope. He had already been diagnosed one year earlier with Behçet's disease. He had suffered from painful erythematous nodules on both lower legs (Fig. 1) and a skin biospy of the erythema nodosum-like nodules revealed thickening of fibrous trabeculae with septal infiltration by heavy chronic inflammatory cells. Inflammatory cell infiltrates had also been present at the periphery of fat lobules. There had been no evidence of eye involvement. He denied the history of recurrent genital ulcer. The pathergy test was negative. Since then, he had been treated with oral prednisolone, however, frequent recurrence of symptoms have been noted.

On physical examination, his blood pressure was 70/60 mmHg and CVA tenderness was noted on the left flank. Laboratory studies revealed elevated levels of white blood cells(23,600/mm3) and decreased levels of the hematocrit (16.6 %). Abdominal sonography showed a massive retroperitoneal hematoma and fluid collection in the left lower quadrant. Abdominal CT revealed leakage of contrast dye from the upper outer portion of the left kidney in the arterial phase and hematoma in the perirenal space (Fig. 2). Emergency angiography was performed and it showed active extravasation of contrast media on the accessory renal artery and pseudoaneurysm formation (Fig. 3). The diagnosis of incomplete type of Beh et's syndrome was made by

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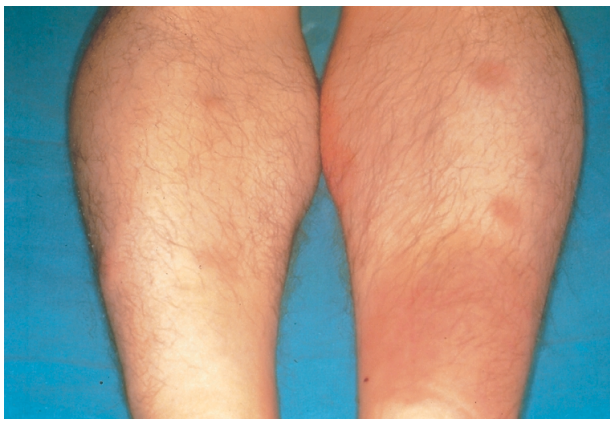

Fig. 1. Erythematous subcutaneous nodules and swelling on both lower legs.

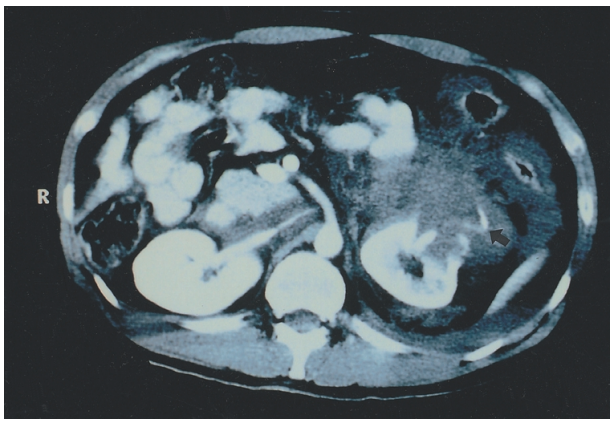

Fig. 2. CT scan shows leakage of contrast dye from the upper outer portion of the left kidney in the arterial phase and hematoma in the perirenal space (arrow).

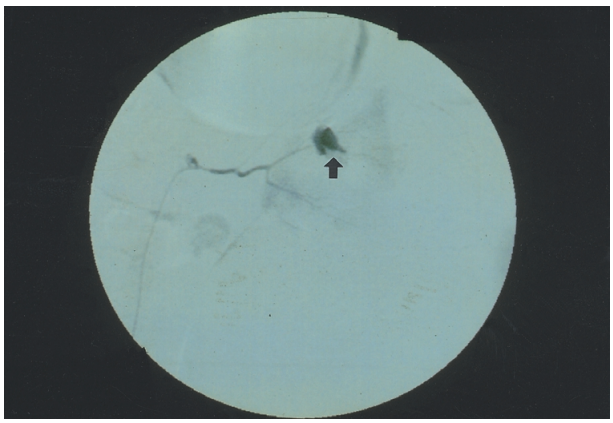

Fig. 3. Emergency angiography demonstrates direct extravasation of blood from left accessory renal artery (arrow).

Shimizu's classification on the basis of erythema nodosum-like nodules on both lower legs, recurrent oral ulcers, arthralgia and vascular involvement[4].

Instead of surgical treatment, arterial embolic occlusion was successfully performed, and his symptoms improved. The patient has been followed for over 20 months and remains in good health and displays no evidence of recurrence.

DISCUSSION

Behçet's disease is a multisystemic inflammatory disease, featuring mucocutaneous, ocular, articular, vascular, intestinal, pulmonary and neurologic involvement[1,6]. It has a worldwide distribution, and is especially prevalent in Japan and eastern Mediterranean countries[6]. Indeed, there are no specific and reliable laboratory features, so the diagnosis of Behçet's disease is based on clinical criteria.

The incidence of vascular involvement in Beh çet's disease is 8-24 % and it is more frequently observed in women than in men, the ratio being 5:1[3,4,7]. Vascular involvement is manifested most often as a venous disease, notably superficial thrombophlebitis, in as many as one third of patients[5].

Arterial involvement occurs in 6-13 % of patients with Behçet's disease and it usually develops late in the course of Behçet's disease[8-10]. The arterial lesion in our patient developed 1 year after the diagnosis of Behçet's disease. Comparing with the studies by Le Thi Huong[10], in which the mean duration between the first sign of Behçet's disease and arterial lesions was 7.3 ± 8 years, the arterial lesion in our patient developed relatively early.

In our conutry, Sung et al[11] reported a case of angio-Behçet's syndrome, arterial occlusion type in a 31 year-old male. Recently, Kim et al[12] reported two cases of deep vein thrombosis associated with Beh et's disease.

Clinical signs of arterial involvement in patients with Behçet's disease consist of occlusive lesions, and formation of true aneurysm and pseudoaneurysms[2,10-18]. The pathogenesis of the arterial ane urysms is thought to be obliterative endarteritis of the vasa vasorum with resulting dilatation and aneurysm formation, or perforation and pseudoaneurysm formation[13,15]. Deposits of C3, C4 and immunoglobulins(IgA, IgG and IgM) have been

detected in the arterial wall and immunological reaction has been suggested to be responsible for the destruction of the media and the formation of the aneurysm [4,14,15].

Aneurysmal dilatation and stenosis or occlusion of the aorta and its branches cause serious complications, such as renovascular hypertension, aseptic bone necrosis, aortic arch syndrome, pulseless disease, syncopal attacks, strokes, myocardial infarcts, valvular regurgitation, and pulmonary hemorrhages[2]. Sixty percent of the arterial aneurysms in Behçet's disease are complicated by rupture, and most vascular-related deaths are caused by aneurysmal rupture[15-17].

The most common site of aneurysm formation in patients with Behçet's disease is the abdominal aorta, followed by the femoral artery, thoracic aorta, common iliac artery, and brachial artery. The involvement of the renal artery is very rare, and indeed there was only one reported case of the involvement of the renal artery in a patient with Behçet's disease. In 1996, Sueyoshi et al[18] reported the case of a 42 year-old Japanese man with Behçet's disease who developed multiple aneurysms and massive retroperitoneal hemorrhage due to rupture of a right renal artery aneurysm. The patient died of pneumonia despite successful embolization.

For the treatment of aneurysms in Behçet's disease, operative replacement of the affected segment can be tried, but it is not recommended because of the high incidence of surgical complications, such as development of a false aneurysm at the site of anastomosis and infections[16,19]. Medical treatment like corticosteroid therapy and immunosuppression sometimes have been used with limited success[10]. Our patient showed marked improvement of symptoms after arterial embolic occlusion and supportive care without surgical intervention.

Dermatologists always should consider the possibility of major vessel involvement when they treat patients with Behçet's disease, even if the incidence is very low and the patient may not complain of any systemic symptoms.

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