Letters to the Editor

Intrarenal microaneurysms in a Behçet's disease patient simulating polyarteritis nodosa

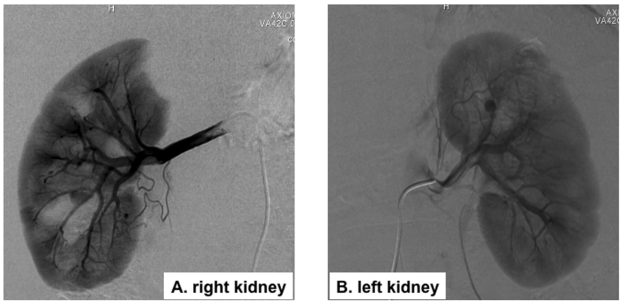

Fig. 1. Pretreatment right (A) and left (B) renal angiograms demonstrating multiple milimetric and one 7-8 mm aneurysm in the right side and one 5 mm aneurysm in the left side.

Sirs,

Renal vascular involvement is rare manifestation of Behçet's disease (BD). In a meta-analysis of 159 BD patients complicated with renal involvement, only 35 patients with renal vascular disease were reported (1). Involvement of the renal arteries with aneurysm formation is reported as isolated case reports in the English literature (2-6). Among those patients, only two demonstrated to have intrarenal microaneurysms (2, 3). We herein report a young man with BD complicated with intrarenal microaneurysms who had been mistakenly diagnosed as polyarteritis nodosa (PAN).

A 29-year-old male complained high fever, generalized fatigue and right flank pain. He had lost 10 kg of body weight within the last 3 months. Renal ultrasonography suggested the presence of bilateral renal hematoma or aneurysms. Digital subtraction angiography of the renal arterial system showed multiple intrarenal microaneurysms in both kidneys (Fig. 1). Those angiographic findings were thought to be consistent with PAN, and he was referred to our clinic. His medical history revealed recurrent oral and genital ulcers for 3 years, and his family history revealed BD in his cousin. In physical examination, blood pressure was 120/80 mmHg in both arms. Multiple large acneiform lesions over the trunk and typical scrotal ulcer scars were noted. Laboratory evaluation revealed that erythrocyte sedimentation rate (ESR) was 60 mm/hr and C reactive protein (CRP) level was 3.67 mg/dL (normal range 0-0.8). Complete blood count, urine analysis, renal and hepatic function tests were unremarkable. Twenty-four-hour urinary total protein excretion was 45 mg/day. Tests for antinuclear antibody, hepatitis B, hepatitis C, and antineutrophil cytoplasmic antigens revealed negative results. Ophthalmologic examination and echocardiographical evaluation revealed normal findings. The diagnosis of BD complicated with renal arterial involvement was made, and he was treated with 500 mg/day intravenous cyclophosphamide and 60 mg/day oral prednisolone. The pulse treatment with cyclophosphamide was repeated three times in one-week intervals, three times in 10-day intervals, and three times in 15-day intervals. Oral prednisolone was decreased 2.5 mg/day in every other day till 40 mg/day, and the dose was gradually switched to alternate day steroid regimen. In the follow-up visit 3 months later, the cumulative dose of cyclophosphamide that the patient received was 4.5 grams, and the dose of prednisolone was 40 mg/every other day. He was completely asymptomatic, ESR was 10 mm/hr and CRP was negative. Control renal angiography demonstrated disappearance of renal aneurysms.

The main pathological defect of BD is vasculitis. The pathophysiology of the arterial injury in BD is suggested to be vasculitis of the vasa vasorum, which eventually causes destruction and necrosis of elastic and muscle cells in the media. Those pathological events may lead to aneurysm formation (7, 8). Arterial aneurysms usually affect large arteries such as the aorta and femoral arteries, and involvement of visceral arteries is rare (8, 9).

Inflammation of the entire vessel wall resulting the formation of microaneurysms is a characteristic feature of PAN. Kidneys are among the most frequently involved internal organs, and the presence of multiple intraparenchymal microaneurysms on angiography was suggested as pathognomonic for this disease (10). However, although very rare, renal arterial involvement during the course of BD may show similar angiographic findings. To our knowledge, only 2 similar patients were reported in the English literature (2,3). Angiographic features of our patient were consistent with PAN. The patient also complained high fever, fatigue and weight loss, which were also suggestive for PAN. However, those symptoms were nonspecific, and can also be present during the clinical course of BD, especially with arterial involvement. Moreover, the history for recurrent orogenital ulcers, large acneiform lesions and scrotal ulcer scars on physical examination fulfilled the diagnostic criteria for BD. The diagnosis of BD is further supported by the family history of our patient. Furthermore, other typical features of PAN such as hypertension, mononeuritis multiplex, myalgia, arthralgia, cutaneuos findings and positivity for hepatitis B were absent.

Arterial aneurysm formation is a life-threatening manifestation of BD, and treatment requires high dose corticosteroids. Immunosuppressive agents are usually added to prevent progression and recurrence of the disease (7, 8). Likewise, we treated our patient with 60 mg/day oral steroids plus intravenous pulse cyclophosphamide initially, and gradually tapered the steroid dose. With this treatment, renal microaneurysms disappeared within 3 months.

In summary, we reported a BD patient with intrarenal microaneurysms who had been erroneously diagnosed as PAN. Therefore, BD should be considered in the differential diagnosis of renal microaneurysms in young adults, especially in countries where BD is endemic.

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