CASE REPORT

Abdominal Aortic Aneurysm with Lumbar Vertebral Erosion in Behçet’s Disease. A Case Report and Review of the Literature


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Introduction

Although Behçet’s disease (BD) has been known for 2000 years, it was named after the author who described it properly in 1937. Recurrent oral or genital aphthosis and uveitis are the main characteristics, but involvement of other organ systems frequently occurs: i.e. the joints, the gastrointestinal tract, the central nervous system and also the vascular system.

A 57-year-old man with known BD was found to have an abdominal aortic aneurysm together with a huge posterior pseudoaneurysm eroding the lumbar vertebrae. Although erosion of the lumbar vertebrae due to an abdominal aortic aneurysm has been reported, the association with Behçet’s disease is unique. Diagnostic and therapeutic aspects of this unusual association are discussed, and a review of the literature presented.

Case Report

A 57-year-old Caucasian man with known BD was admitted to a general hospital because of anorexia, vomiting and dysphagia for solid food. His medical history included concomitant blindness and dysarthrism, an impaired oral glucose tolerance test (treated with 80 mg gliclazide daily) and mild coronary artery disease. The diagnosis of BD in this patient was based upon episodes of oral aphthosis, involvement of the eyes, the joints and the cardiovascular system. He had not smoked for approximately 20 years.

Physical examination did not demonstrate oral or genital ulceration.

Haematological evaluation revealed a slightly elevated white blood cell count of $8.3 \times 10^3$/ml (normal values: 4.0 – 7.0 $\times 10^3$/ml), a sedimentation rate of 18 mm for the first hour (normal values: 2 – 8 mm) and 40 mm for the second hour (normal values: 5 – 15 mm); and the C-reactive protein was 1.79 IU/ml (normal values: 0.00 – 0.60 IU/ml).

Biochemical analysis showed a haemoglobin A1 level of 8.7 % and an alkaline phosphatase level of 267 IU/l (normal values: 60 – 180 IU/l).

Routine abdominal X-ray showed extreme deformation of the lumbar vertebral bodies L2 and L3 with contralateral fusion of the vertebrae. On abdominal ultrasound, an aortic aneurysm with a diameter of 94 mm was suspected. Computerised tomography of the abdomen and lumbar spine confirmed an infrarenal aortic aneurysm with a diameter of 60 mm. A mass with a diameter of 117 mm and significant erosion of the vertebral bodies L2 and L3 was present posteriorly (Fig. 1). Magnetic resonance imaging confirmed a huge posterior pseudoaneurysm destroying the vertebrae and expanding into the epidural space (Fig. 2). Aortography demonstrated a fusiform dilatation of the aorta with an irregular wall.

Because of these findings, the patient was referred to our hospital and combined vascular and orthopaedic reconstruction was performed in one stage.
Inflammation with fibrotic reaction around the femoral arteries was present. After standard median laparotomy a 60 mm large aortic aneurysm extending from the renal arteries to the bifurcation was found. Here too, an inflammatory reaction with calcification was present. Because a vascular clamp could not be placed safely below the renal arteries, the aorta was clamped just above the right and a left polar renal artery after dividing the left renal vein. After opening the aneurysm, a communication with the pseudoaneurysm was observed through a tear in the posterior aortic wall. The lumbar arteries and inferior mesenteric artery were found to be occluded. A 14 / 7 mm albumin coated Dacron prosthesis was sutured end to end to the infrarenal transected aorta. The total suprarenal clamping time was 25 min and forced diuresis with mannitol was initiated. The common iliac arteries were ligated. End-to-side anastomoses to both femoral arteries were performed. Under the same anaesthetic, the orthopaedic surgeons stabilised the lumbar spine from L1 to L4. After mobilisation, the left hemicolon, the pancreas, the spleen and the left kidney were turned to the right side. A left anterolateral fixation with a Kaneda system from L1 to L4 was performed while the eroded vertebral bodies were filled with a femoral allograft and bony chips (Fig. 3).

Pathological examination of the resected aortic wall showed a non-specific inflammatory infiltrate compatible with Behçet's vasculitis. Perivascular infiltrates of lymphocytes and plasma cells were present. Atheroma was also present. Samples from the pseudoaneurysm were sent for microbiological examination, but cultures were sterile.

His postoperative recovery was only complicated by mild renal insufficiency which did not require dialysis and he was discharged after 5 weeks. Unfortunately, he died 6 months later due to a myocardial infarction. An autopsy was not performed.

Discussion and Literature Review

Behçet's disease is a multisystem disorder with a peculiar geographic distribution. It is most frequently reported in the Mediterranean region and in Japan. The prevalence rate in Japan is estimated at 10 / 100 000,34 in Northern Turkey at 190 / 100 0003,5 and in western countries at 5 / 100 000.3,6 It is most often diagnosed in the second or third decade and men are more often affected than women, with a ratio of 2.3 / 1.7 In Japan and the Mediterranean countries,7 there is a proven association with HLA B5, but not in western countries.3,5 The diagnosis of BD is based upon five major criteria: recurrent oral ulcerations in combination with genital ulceration, eye lesions, skin lesions or
BD can also affect the joints, the gastrointestinal tract, the central nervous system and the cardiovascular system. Because vascular involvement is a major cause of death in Behçet's patients, some authors regard vasculitis as a sixth major diagnostic criterion. In our patient blindness had occurred due to eye lesions and joint and cardiovascular involvement were clearly present.

In a literature review by Koç et al., 7.7–60% of the patients with BD, showed signs of vascular involvement (Table 1); 25% of these lesions affected exclusively the venous system, whereas only 7% affected only the arterial system. The majority of the patients with vascular involvement (68%) presented with both venous and arterial lesions. The vascular lesions in BD include arterial and venous occlusions, aneurysm and pseudoaneurysm formation, or combinations of these conditions. The most frequently reported vascular complication is thrombosis of the large veins of the limbs and superior or inferior caval vein syndrome. Recurrent subcutaneous thrombophlebitis is an important risk factor for deep venous thrombosis or caval syndrome. Early detection and aggressive therapy of peripheral vein thrombosis are crucial in preventing propagation towards deep venous thrombosis. Arterial involvement is less common and consists mostly of aneurysm and pseudoaneurysm formation or arterial occlusion. Aneurysms of large arteries like the abdominal aorta and the pulmonary artery are most frequently reported although nearly every major artery has been found to be involved. This aneurysm formation can present as a very rapidly progressive disease. Lumbar erosion due to a large aortic aneurysm or pseudoaneurysm is very unusual, and has been reported only once in combination with BD. In patients with BD, pseudoaneurysms are related to arterial punctures for aortography or to rupture of the inflamed arterial wall.

The aetiology of BD is still unknown. In 1937 a viral infection was considered to be the cause, but nowadays autoantibodies to human oral mucosa and immune complex formation are thought to be involved. Deposition of these immune complexes in the arterial wall result in an activation of the complement cascade with inflammatory reaction. Therefore, vasculo-Behçet can be considered as an immune induced vasculitis. The characteristic pathological changes of Behçet’s aortitis and of vasculo-Behçet in general have been well described in many studies. In the active stage there is perivascular inflammatory infiltration in the media and adventitia (consisting of neutrophils, eosinophils, lymphocytes and plasma cells), proliferation and obliteration of the vasa vasorum, and proliferation of fibroblasts with loss of elastic and muscle fibres in the media. Aortic aneurysm formation can be attributed to this intense inflammation of the media which weakens the arterial wall, sometimes complicated by perforation resulting in a pseudoaneurysm. All these histological characteristics were found in the present case. In the later stage of this vasculitis, fibrous rearrangement of the intima and adventitia occur leading to stenosis and arterial occlusion. These lesions however are usually encountered in the smaller arteries of the extremities. The pathological changes in thrombophlebitis are similar with intimal fibrous thickening, luminal obstruction and vessel wall inflammation.

This inflammation can also be observed in blood analysis with leukocytosis and an increased sedimentation rate. These two factors were also observed in our present case.

Although the treatment of vasculo-Behçet is controversial, reconstruction with prosthetic graft is usually unavoidable for an aneurysm or pseudoaneur-
Aortic Aneurysm with Behçet's Disease

Table 1. Vascular involvement in patients with Behçet's disease.

<table>
<thead>
<tr>
<th>Vascular involvement</th>
<th>Venous</th>
<th>Arterial</th>
<th>Both</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kuzu</td>
<td>1°</td>
<td>1200</td>
<td>192</td>
</tr>
<tr>
<td>Koç</td>
<td>1°</td>
<td>137</td>
<td>38</td>
</tr>
<tr>
<td>Kabbaj</td>
<td>1°</td>
<td>125</td>
<td>40</td>
</tr>
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<td>Benamour</td>
<td>1°</td>
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<td>72</td>
</tr>
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<td>Literature review</td>
<td>ns</td>
<td>728</td>
<td>7.7 - 60%</td>
</tr>
<tr>
<td>Present review</td>
<td>1070</td>
<td>490</td>
<td>46%</td>
</tr>
</tbody>
</table>

ns = not specified.

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References


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