Spontaneous dissection of the extracranial vertebral artery with spinal subarachnoid haemorrhage in a patient with Behçet’s disease

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Abstract. A 40-year-old man with known definite Behçet’s disease (BD) was admitted with confusional state which had started 4 days before admission with an acute headache and vomiting. Neurological examination revealed confusion, stiff neck, right facial weakness, left hemiparesis, dysarthria and truncal ataxia. CSF was haemorrhagic and xanthochromic. Cranial CT scans were negative, but MRI showed a right pontine hyperintense lesion on T2-weighted images. Bilateral carotid angiograms were normal. Right vertebral angiogram showed findings consistent with a dissection at the V2 segment of the artery. At the level of the fifth cervical vertebra, a radiculomedullary branch of the vertebral artery with an aneurysmal dilatation in its intradural portion was notable. This case shows that, in BD, aneurysm formation can also occur in a spinal artery and spontaneous vertebral artery dissection can be seen.

Key words: Spontaneous arterial dissections – Subarachnoid haemorrhage – Spinal aneurysm – Arterial dissection – Behçet’s disease

Behçet’s disease, first described by the Turkish dermatologist Hulusi Behçet in 1937, is a systemic disease of unknown aetiology. Involvement of various organ systems is due to a vasculitis affecting mainly veins, venules, and the capillaries. The less frequent arteritis, which often worsens the course of the disease, usually affects large arteries [1].

Case report

A 40-year-old right-handed man presented with acute, severe head and backache, vomiting, confusion, diplopia, dysarthria and dysphagia. The head and backache had come on 4 days prior to admission, diplopia, dysarthria and dysphagia the next day, and subsequently a confusional state with hallucinations developed. He had a 10-year history of recurrent oral and genital ulcers with occasional erythema nodosum-like lesions. Seven years previously he had thrombophlebitis in the right leg and an undefined episode of blurred vision. The same year the skin pathergy test was positive and Behçet’s disease was diagnosed. During the next 7 years the disease followed a silent course except for recurrent oral ulcers.

The patient had abdominal distention and tenderness due to chronic oesophagitis, gastroduodenitis, and proctitis. He had no active mucocutaneous or ocular signs other than evidence of old glaucoma. He was confused and drowsy with occasional visual hallucinations and had a stiff neck with signs of meningeal irritation. Right abducens and facial palsies together with dysarthria were noted. He also had severe truncal ataxia with right cerebellar signs and a slight left hemiparesis.

The sedimentation rate was 60 mm/h. Other than 20,000 white blood cells/mm³ his complete blood count, blood chemistry and coagulation studies were normal. CSF was grossly bloody with marked xanthochromia.

CT was normal. A week after admission, four-vessel angiography revealed no intracranial abnormality. There was irregular stenosis of the right vertebral artery (VA), from the fifth to the second cervical vertebral levels, at the lower end of which was an aneurysmal dilatation of the anterolateral wall. A vertical linear filling defect was seen between the arterial wall and the aneurysm. An anterior radiculomedullary artery arising from the right VA at C5–C6 had a 3 x 2 mm aneurysm approximately 3 mm before it reached the anterior spinal artery (Fig. 1a, b). A month later, the right VA was unchanged, but the radiculomedullary artery at C5–C6 was not seen (Fig. 1c).

Cranial MRI on the 25th day showed a hyperintense lesion on proton density and T2-weighted images on the right of the pons extending to the medulla oblongata (Fig. 2). Coronal T1-weighted images revealed a narrow, irregular right VA at C2–C5 with a narrow-linear hyperintensity along its lateral wall at C4–C5 (Fig. 3a). MRI a year later showed a completely normal right VA (Fig. 3b).

High-dose methyl-prednisolone therapy was commenced on the day of admission. The patient showed steady improvement and was discharged 1 month later with only slight truncal ataxia.

Discussion

During the course of Behçet’s disease, neurological involvement is variable. In a prospective study we found neurological involvement in 5.3% of cases [2].

Arteries are affected much less frequently than veins: 1–6%, as compared to about 70% [3–5]. The arteries most
commonly affected are the aorta and peripheral arteries. Arterial lesions usually consist of an aneurysm, pseudoaneurysm or obstruction associated with an aneurysm. Arteritis of vasa vasorum or arteritis of the arterial wall itself are thought to be responsible [6]. There is one report of arteritic occlusion of the branches of the middle cerebral artery [7].

Our case fulfilled the diagnostic criteria of the International Study Group for Behçet's disease [8], and presented with two frequent distinct constellations of neurological findings, signs of meningeal irritation and brain stem involvement. What makes it unique is the presence of two very uncommon vascular lesions, which we believe caused the clinical picture: meningeal irritation was due to subarachnoid haemorrhage (SAH) from a spinal artery aneurysm, and brain stem signs were caused by VA dissection.

SAH is extremely rare in Behçet's disease. We found only 1 case of spinal SAH, a patient who developed spinal block signs [9]. In our case four-vessel angiography revealed aneurysmal dilatation of a radiculomedullary branch of the right VA. Although on the anteroposterior views it appeared fusiform, the possibility of a saccular aneurysm could not be excluded. Matsumoto et al. [6] reported four pathologically verified saccular aneurysms due to vasculitis in Behçet's disease. In either case, we considered this aneurysmal dilatation to be the cause of the SAH, and that the aneurysm developed because of Behçet's disease vasculitis, although there is no pathological verification.

Isolated spinal artery aneurysms are rare [10-14]; a review of the literature in 1981 revealed six cases in all [15]. In the last 10 years, eight more have been reported, including a posterior spinal artery aneurysm due to vasculitis in a patient with systemic lupus erythematosus, one associated with fibromuscular dysplasia (FMD) and two in patients with coarctation of the aorta, one associated with pseudoxanthoma elasticum [16-18].

In our case brain stem signs developed on the day following the SAH. The angiographic findings, i.e., the irregular narrowing at the V7 segment of the right VA, together with the aneurysmal dilatation at the beginning of this segment, explain these signs. The radiological findings are compatible with arterial dissection [19]. Neither spinal artery aneurysm, nor this type of arterial involvement has been reported in Behçet's disease.