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Letter to the Editor

Stroke revealing Neuro-Behçet's disease with parenchymal and extensive vascular involvement

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Dear Editor

1. Background

The frequency of neurologic manifestations in Behçet's disease (BD) ranges between 5% and 15% [1]. Central nervous system manifestations are represented by parenchymal involvement, which includes brainstem involvement, hemispheric manifestations, spinal cord lesions, and meningo-encephalitic presentations and non parenchymal involvement, including dural sinus thrombosis, arterial occlusion, and/or aneurysms. The association of parenchymal and vascular manifestations is extremely rare [2]. Among the neuro-vasculo-Behçet's disease patients, venous thrombosis is well-known while arterial involvement has rarely been reported and seldom concerned cerebral arteries. We describe here a stroke case revealing neuro-Behçet's disease (NBD) with parenchymal involvement and extensive vascular lesions interesting several intracranial and systemic arteries. To the best of our knowledge, this is the first reported case of Behçet's disease manifesting as concomitant involvement of systemic and cerebral arteries.

2. Case report

A 45-year-old man with hypertension first diagnosed 8 years ago and a 40 pack-year smoking history presented with sudden right-sided weakness and mutism. On admission, he was afebrile and conscious with no verbal output and a right hemiplegia. He had an asymmetric brachial blood pressure (240/120 mmHg in the right arm and 160/90 mmHg in the left arm).

Magnetic resonance imaging (MRI) revealed an acute ischemic lesion in the territory of the left anterior cerebral artery, sequelae of left capsulo-lenticular hematoma, pontine lacunes and subcortical microbleeds. It also showed unilateral meso-diencephalic and bilateral middle cerebellar peduncle lesions (Fig. 1). MR angiography showed an occlusive stenosis at the right middle cerebral artery (MCA) M2 segment and at the left vertebral artery V4 segment (Fig. 2). Thoraco-abdominal angioscan revealed infiltrated walls with aortic isthmus stenosis, multiple aneurysmal dilatations in descending aorta, both deep circonflex iliac arteries and left common femoral artery as well as occlusive stenosis of left subclavian artery, celiac trunk, superior mesenteric artery and both renal arteries (Fig. 3). Electrocardiogram and transthoracic echography showed left ventricular hypertrophy. Complete blood counts, biochemical screening, erythrocyte segmentation rate and C-reactive protein level were all normal and antinuclear antibodies, rheumatoid factor, and antiphospholipid antibodies were negative.

Two days after admission, the patient recovered normal speech and reported a history of recurrent oral ulcerations. Therefore, the diagnosis of Behçet's disease was suspected. Skin examination showed pseudo-folliculitis lesions on the trunk. The pathergic test was negative and ophthalmologic examination was normal. HLA typing showed HLA A30-B17 haplotype.

Lumbar puncture revealed hyperproteinorachia at 0,69 g/l with normal cytology.

The diagnosis of BD was made referring to « International consensus recommendation (ICR) diagnostic criteria of NBD ».

The patient received aspirin and intravenous methylprednisolone 1000 mg/day for 5 days. He was discharged from hospital with the antiplatelet associated with oral steroid and pulse cyclophosphamide as maintenance treatment. He had complete recovery within 2 months.

3. Discussion

Behçet's disease has high prevalence rates in the Mediterranean, Middle Eastern, and Far Eastern regions. It affects mainly young people with a more severe course among men [3].

BD is unique among other vasculitis as it has the propensity to affect the venous rather than the arterial side. It is also unique in that it has significant thrombotic tendency associated with vascular inflammation, which cannot be explained with specific thrombophilic factors [4]. Arterial involvement affects 4 to 17% of patients according to the series [4]. It is probably underestimated since in a Japanese autopsy series, arterial involvement was found in 34% of 170 patients [5]. It manifests mostly in the form of aneurysms (45–70%) and rarely as thrombotic occlusions [6] or arterial stenosis (13%) [7]. All three types of arterial disease were present in our patient which was unusual.

Vessels of different calibres can be involved in BD. Pulmonary arteries, which resemble venous structures, are often involved, while the aorta and peripheral arteries are less commonly affected [6]. Peripheral

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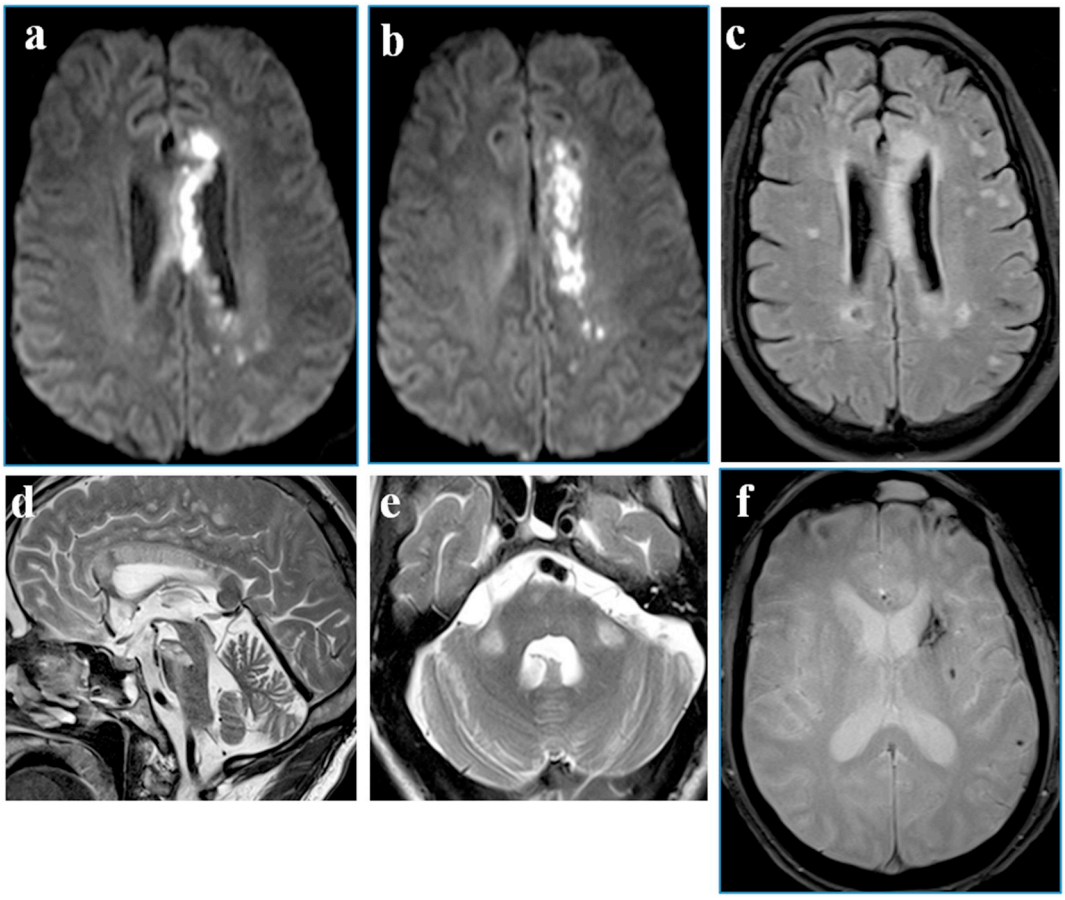

Fig. 1. Cerebral MRI showed an acute ischemic lesion in the territory of the left anterior cerebral artery on diffusion images (a, b). FLAIR (c) and T2 (d, e) images showed subcortical nodular hyperintensities (c), chronic pontic ischemia (d) and bilateral middle cerebellar peduncle hyperintensities (e). Sequelae of left capsulo-lenticular hematoma and subcortical microbleeds on T2* image (f).

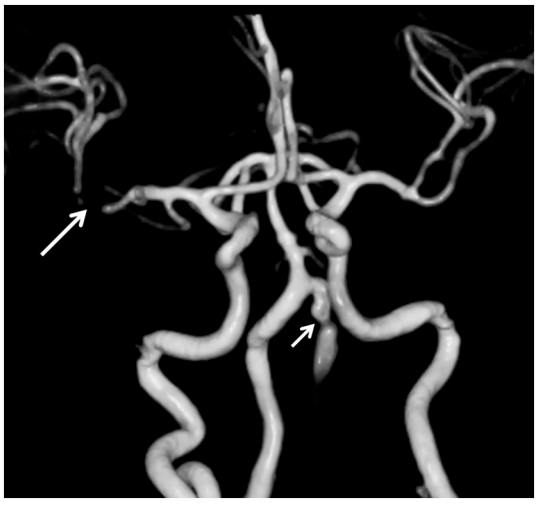

Fig. 2. MR angiography of Willis circle: Occlusive stenosis at the right MCA M2 segment (long arrow) and at the left vertebral artery V4 segment (short arrow).

arterial involvement is found in less than 5%. Commonly affected arterial sites are the infra-renal abdominal aorta (11%) and iliac, femoral, popliteal, and carotid arteries. Thoracic aorta is diseased in only 5% of cases. Digestive, coronary and cerebral arteries are much less affected [8]. In our patient there was an association of different rare localisations of artery involvement, represented by thoracic aorta, digestive and cerebral arteries (the right MCA and the left vertebral artery). Cerebral vasculitis and brain infarction represent particular and unusual manifestations during the course of BD, reported in 1–5% of major series in the literature (1999; Al-Araji et al., 2003; Al-Fahad et al., 1999; Barros et al., 2007; Benamour et al., 2006; Borhani-Haghighi et al., 2006; Houman et al., 2007; Joseph et al., 2007; Monaco et al., 2006; Turker et al.) [9].

Intra- or extra-cranial aneurysm or aneurysmal dilatation represent well-known entities, but intra-cranial cerebral artery stenosis or occlusion, as observed in our patient, are very rare. The occlusions of

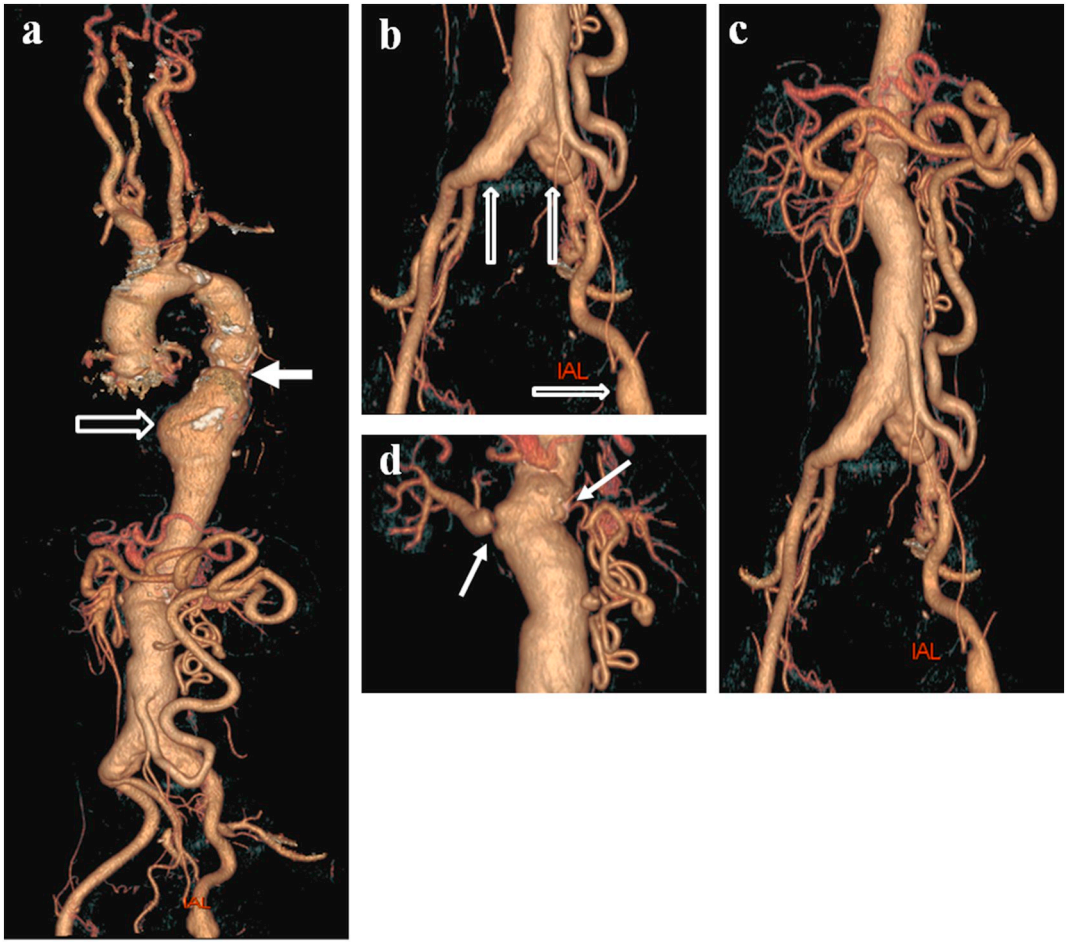

Fig. 3. Thoraco-abdominal angioscan: (a): aortic isthmus stenosis (thick arrow) and aneurysmal dilatation in descending aorta (empty arrow), (b): aneurysmal dilatations in both deep circonflex iliac arteries and left common femoral artery (empty arrows). (c): occlusive stenosis of celiac trunk and superior mesenteric artery. (d): occlusive of both renal arteries (arrows).

proximal segments of medium sized intra-cranial vessels are the most common especially of the MCA.

Our patient had diffuse aortitis which is rarely observed in patients with BD unlike other large vessel vasculitis (i.e. giant cell arteritis and Takayasu's arteritis) and aortic aneurism is most often unique [10]. In accordance with previous BD reports [6], the homogenous and concentric arterial wall thickness, commonly observed in Takayasu arteritis, was absent in our case.

Premature atherosclerosis does not seem to be a part of vessel inflammation in BD [3]. However, cigarette smoking is a well known cofactor in BD vascular lesions.

Besides vascular BD manifestations, our patient presented mesodiencephalic and pontine lesions typical of parenchymal BD. This association has seldom been reported in the litterature.

Some authors had considered that a low-grade chronic meningoencephalitis is the principal neuropathological process in NBD rather than a true vasculitis. However, in our case, the simultaneous involvement of multiple intra and extra cranial arteries and the absence of meningitis seem to be better explained by the fact that BD is a multiorgan vasculitis possibly affecting the central nervous system.

In conclusion, this case illustrates that Behçet's disease should be considered in cases of concomitant cerebral and systemic artery lesions particularly in young patients from the endemic areas.

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Conflicts of interest

The authors declare that they have no links of interest.

Authorship

HN, BSS, NF, BAI and HF took care of the patient and drafted the report. NS and MM realized figures.

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