

Case report

A young boy with affected heart and mind: arterial involvement in a patient with undiagnosed Behçet's disease

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ABSTRACT

Behçet's disease is a vasculitis that manifests with oral and genital ulcerations, skin lesions, uveitis, vascular, central nervous system and gastrointestinal involvement. While most patients develop mucocutaneous and genital ulcers along with eye disease, other patients may also present with arthritis, frank vasculitis, thrombophlebitis and central nervous system disease. This is the first case in the literature, where the diagnosis of Behçet's disease was made secondary to concomitant involvement of coronary and carotid arteries.

Case report

A 17-year-old male with a history of anteroseptal myocardial infarction (MI) and ischaemic stroke presented to our hospital for further investigation. In the physical examination, right-sided hemiplegia was detected. He had no family history for premature atherosclerosis. Electrocardiography (ECG) demonstrated sinus rhythm with pathological anteroseptal Q-waves. Routine laboratory tests; including complete blood count, liver enzymes, kidney function tests and lipid profile were normal. He was screened for hypercoagulability and no abnormalities were detected. The transthoracic echocardiogram revealed hypokinesis in the mid anterior and mid anterior septal regions. Patent foramen ovale was not detected in the transesophageal echocardiogram. In the cranial computed tomographic angiogram, there was a major dissection in the left internal carotid artery (ICA) (Fig. 1) and the cranial magnetic resonance imaging (MRI) revealed chronic haemorrhagic necrosis in the left middle cerebral artery territory secondary to occlusion of cavernous segment of the left ICA (Fig. 2). A coronary angiography was performed and an aneurysm of

17 mm was detected in the left main coronary artery (LMCA) (Fig. 3). The patient was thoroughly evaluated for the differential diagnosis of arterial aneurysms; including vasculitis (such as Kawasaki disease, Behçet's disease, polyarteritis nodosa, Churg-Strauss syndrome, Takayasu arteritis, fibromuscular dysplasia); connective tissue diseases (such as Ehlers-Danlos syndrome, Marfan syndrome) and other causes (such as polycystic renal disease, type 1 neurofibromatosis, cocaine abuse, bacterial and mycotic infections, exposure to herbicides). The patient was consulted with the Genetics department, however findings were not found to be compatible with any specific syndrome. The patient was also consulted with the Rheumatology department. Laboratory tests, including erythrocyte sedimentation rate, C-reactive protein and autoimmune parameters [(anti-nuclear antibody, anti-ds DNA, rheumatoid factor, anti-neutrophil cytoplasmic antibodies (C- and P), anti-cardiolipin Ig M and Ig G, anti-phospholipid Ig M and Ig G)] did not prove substantial results, except for positive human leukocyte antigen-B51 (HLA-B51). Among viral serological markers, the patient was found to be positive for anti-HBs and negative for anti-HCV and anti-HAV Ig M and G. Meanwhile, the patient was evaluated by Neurology and Cardiovascular Surgery departments; medical treatment with acetylsalicylic acid and warfarin was initiated.

Seropositivity for HLA-B51 accompanied with vascular involvement changed the direction of the examination to Behçet's disease. He underwent detailed examination for common findings and manifestations of the disease. After cessation of cigarette smoking, minor oral ulcers that were <10 mm in diameter occurred and the patient

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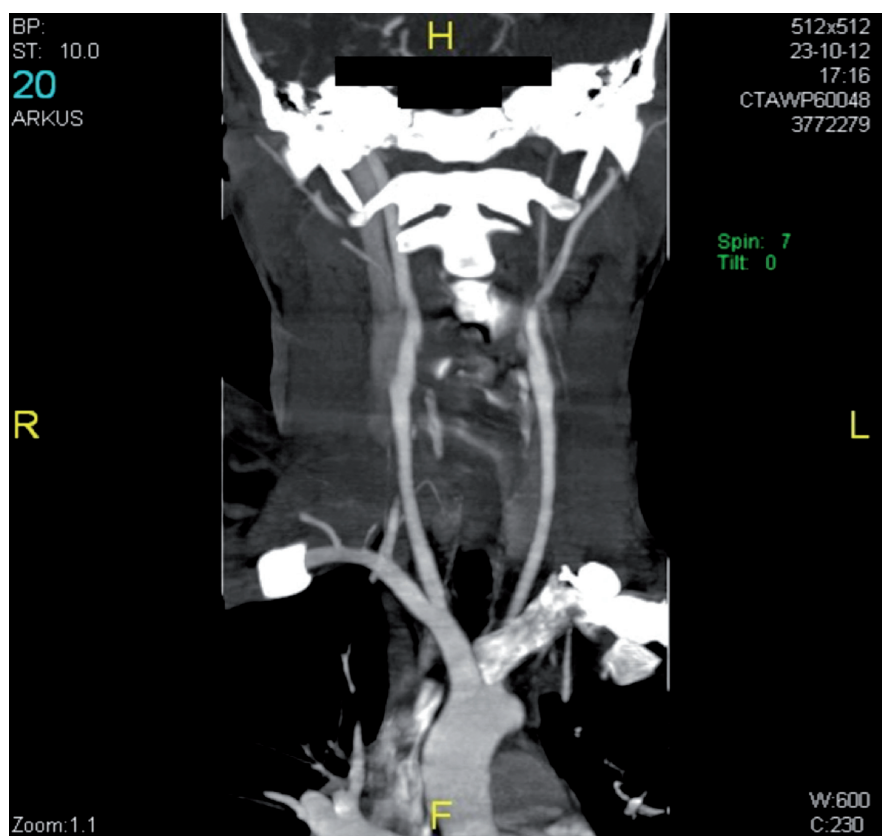


Fig. 1. Cranial computed tomographic angiogram revealing dissection in the left internal carotid artery (ICA). At 1 cm distal of its origin, ICA is patent with preocclusive submillimetric diameter. In the distal cavernous segment, it is totally occluded. Lumens of supraclinoid ICA, left A1 branches and middle cerebral artery (MCA) are significantly narrowed. Wide cystic encephalomalacic chronic infarct in the left MCA territory is present.

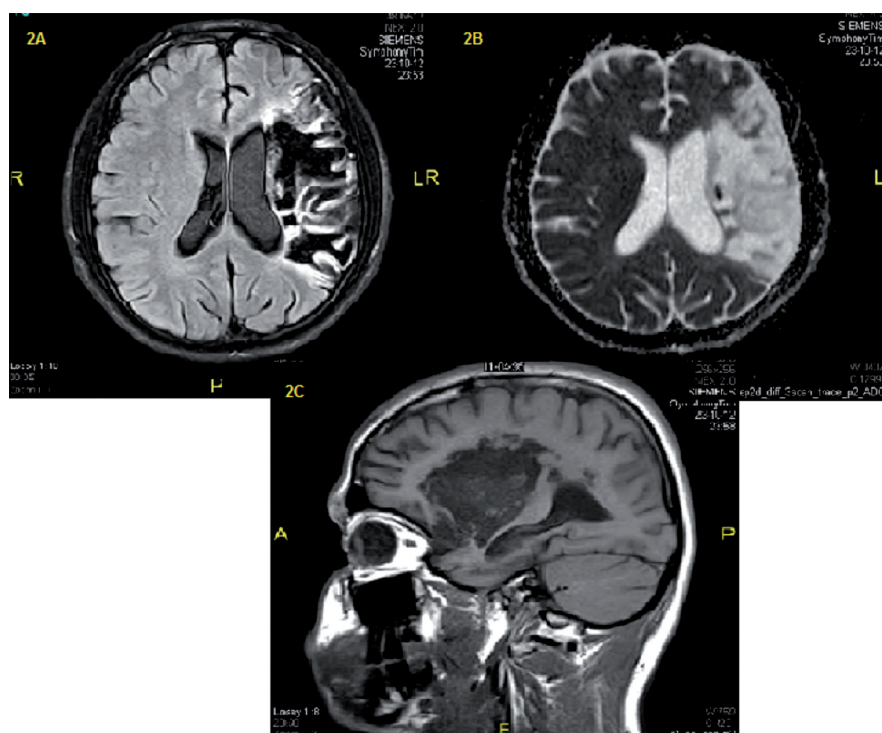


Fig. 2. Magnetic resonance imaging revealing chronic haemorrhagic infarct in the left MCA territory secondary to occlusion of cavernous segment of the left ICA.

mentioned that he had similar painful recurrent oral ulcers prior to smoking (approximately 5 times/ a year), which usually resolved spontaneously within few weeks. Ulcers were localised in the inner lips and buccal mucosa. It was learnt that he had no history of genital ulcers, besides his negative physical examination. He had papulopustular lesions indistinguishable from acne vulgaris, particularly on the trunk. On the anterior tibial surface, erythema nodosum-like lesions were observed. Skin pathergy test was performed on the hairless site of the flexor aspect of the forearm. A positive pathergy reaction, which is defined as formation of a papule >2mm in size developing 24-48 hours following oblique insertion of a 20-25 gauge needle 5 mm into the skin, was obtained (4/6 skin prick tests were positive). Patient was consulted with the Ophthalmology department and no ocular disease was detected. He had no gastrointestinal system-associated symptoms or musculoskeletal findings in his clinical examination except for his disability secondary to right-sided hemiplegia.

The patient was diagnosed with Behçet's disease according to the International Study Group for Behçet's disease criteria (1). He received systemic anticoagulation in addition to corticosteroids (prednisolone) and azathioprine, colchicine for the mucocutaneous lesions (2).

Vascular involvement usually affects the veins more commonly than the arteries in Behçet's disease. The arterial system involvement rate is 3–12% (3). The most common pathological lesions are aneurysms (4-6), followed by occlusion and stenoses of the aorta. The most common sites for aneurysm development are the abdominal aorta, pulmonary, femoral, popliteal and carotid arteries (4-6). The main pathologic event is a lymphocytic and neutrophilic necrotising vasculitis leading to saccular aneurysms and rupture (7). Peripheral arterial pseudoaneurysm was detected in a patient 20 years after disease onset (8). Tuzun *et al.* have reported one case of carotid artery aneurysm in a cohort of 24 patients diagnosed with Behçet's disease during

a follow-up period of 19 years. In this cohort, none of the patients developed coronary artery aneurysms (9). Presentation with aneurysms before its sine qua non lesions was reported in a patient diagnosed with Behçet's disease following the detection of a false aneurysm of the aorta (10).

Coronary arterial involvement is extremely rare and usually occurs following involvement of other systems in Behçet's disease (11). True and false pseudoaneurysms of the coronary arteries were reported in a patient 9 years after the disease onset (12). Our case is unique to be the first case in the literature, where the diagnosis of Behçet's disease was made secondary to concomitant involvement of coronary and carotid arteries. This case is significant for showing Behçet's disease may manifest in adolescents with arterial involvement before well-known, essential systemic manifestations occur.

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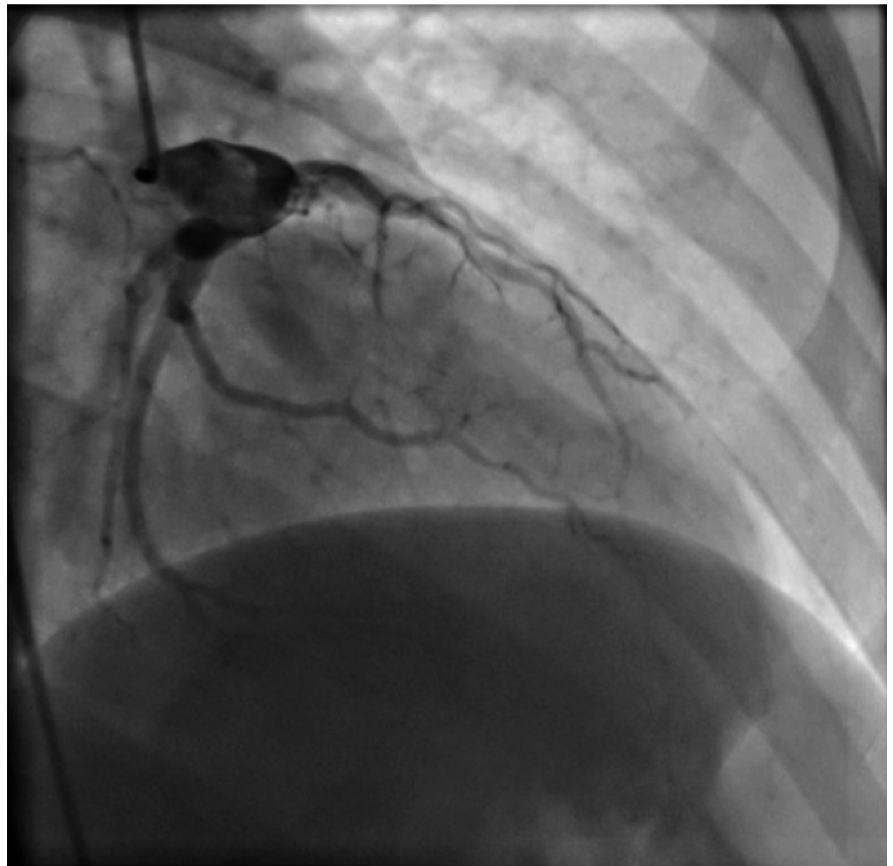


Fig. 3. Coronary angiogram showing a 17 mm aneurysm in the left main coronary artery.