Cardiology in the Young

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An unusual case of paediatric Behçet's disease with severe stenosis of the abdominal aorta and recurrent venous thrombi

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Brief Report

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Behçet's disease is a multisystemic inflammatory disease. Behçet's disease is usually observed in young adults; however, up to 26 % of cases are in childhood.[1] The vascular involvement is the initial sign of Behçet's disease in 27.5 % of adult patients and usually presented with recurrent superficial or deep vein thrombosis in the lower extremities.[2,3] Arterial lesions are the least frequent and develop between 5 and 18 % of adult cases.[2] Arterial stenosis is also uncommon complications with a prevalence rate of less than 5%.[3] Stenosis of the abdominal aorta due to Behçet's disease has not been reported in childhood before. Here, we reported a child presenting with unusual cardiovascular findings and discussed the challenge in diagnosis of vascular Behçet's disease in paediatric cases.

Case

A 15-year-old girl admitted with complaints of shortness of breath, palpitation, and headache. She had a history of intermittent arthralgia for 2 years. On admission, patient blood pressure of right arm and left leg was 219/126 and 110/80 mmHg, respectively. Renal Doppler ultrasonography showed severe stenosis in the abdominal aorta before giving truncus coeliacus and mesenteric artery branches. Aortic lumen showed 90% narrowing. Long segment (27 mm) stenosis in abdominal aorta with gradient of 70 mmHg was detected in subcostal examination at echocardiography. Aortic CT angiography showed diffuse increase in the wall thickness in the whole descending aorta. The lumen diameter of aorta abruptly decreased to 3 mm, especially at the level of the diaphragmatic hiatus (Fig 1). All these radiological findings were consistent with large vessel vasculitis. Ocular examination was normal; arterial and venous angiographic examinations of cranium and all extremities were normal. The pathergy test was negative. In laboratory tests; haemoglobin level was 11.7 g/dl, white blood cell count was 8.330 per mm3, and platelet count was 306.000 per mm3. C-reactive protein level (18.5 mg/L, upper limit: 5 mg/L) and erythrocyte sedimentation rate (24 mm per hour) were also high. Thrombose panel was normal, except mild decrease in Protein C, and high D-dimer (1.6 mg/L, upper limit 0.55 mg/L) and ferritin level (212 mg/L upper limit: 150 mg/L). Autoantibodies for collagen tissue disease and HLA B51 were negative. There was no laboratory evidence for bacterial, viral agents, and mycobacterium tuberculosis. Whole body PET scintigraphy showed an increased activity in abdominal aortic wall and bilateral femoral arteries. Then, intravenous immunosuppressive (cyclophosphamide and corticosteroid) treatment was started with the diagnosis of Behçet's disease or Takayasu arteritis type 4. On the third day of treatment, a swelling in antecubital region of the right arm of the patient developed. Superficial Doppler USG was displayed on a thrombus in the lumen of vena basilica. Then, anticoagulant therapy with enoxaparin was started. Although immunosuppressive treatment and anticoagulant therapy at prophylaxis dose, a thrombus was detected in abdominal aorta just above the stenotic segment by transthoracic echocardiographic examination at the second month of treatment. Thrombus disappeared with enoxaparin treatment. Heterozygous *p. Thr68Met (c.203C>T)* alteration detected in the *PSTPIP1* gene known as one of the rare genes associated with Behçet's disease and colchicine therapy was initiated with the diagnosis of Behçet's disease. Despite 1-year immunosuppressive therapy, no regression was detected in the diameter of the stenotic area of abdominal aorta and stent implantation was successfully performed in other cardiac centre. After stent implantation, it was observed that the patient's arterial blood pressure was normalised.

Discussion

Up to 80% of paediatric cases do not fulfil the international criteria of Behçet's disease.[4] Also, our case did not meet the criteria of the "Full International Study Group Criteria" as in many children. Similarly, it was found that she did not meet the "International Criteria for Behçet's Disease" and "New Pediatric Behçet's Disease Criteria". Acute phase reactants did not guide

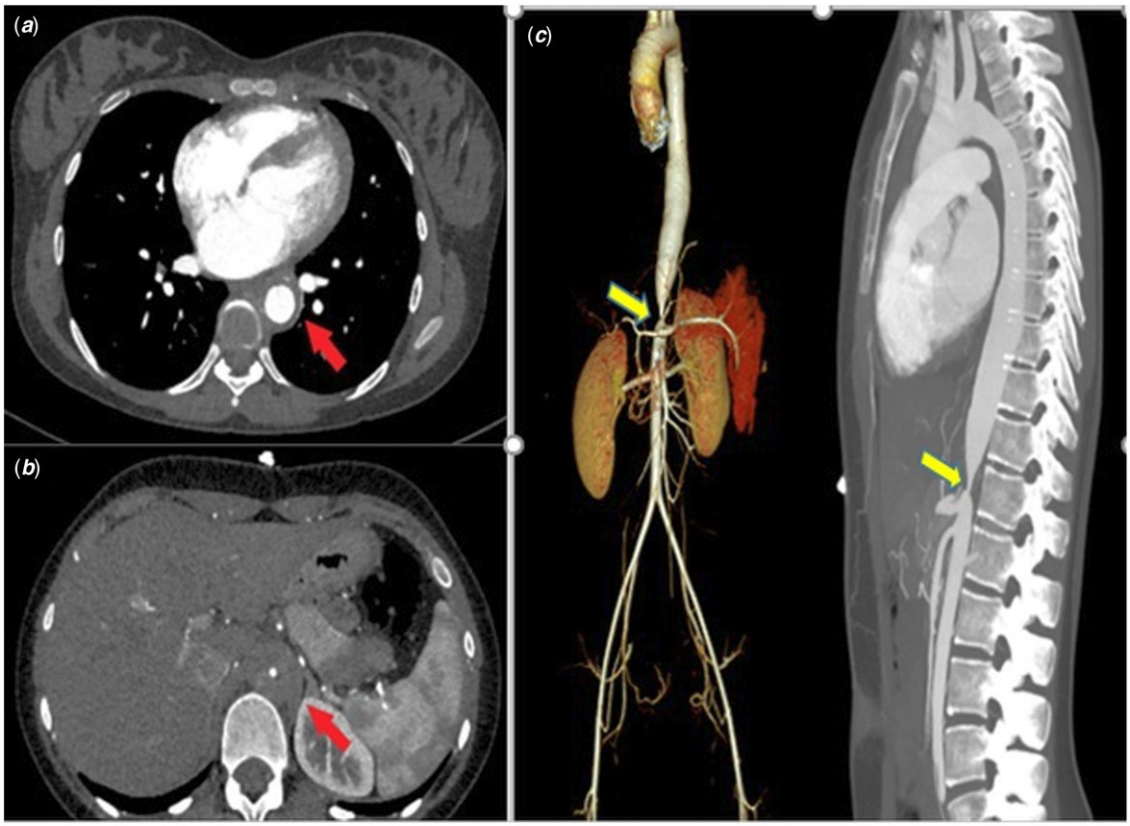

Figure 1. CT angiography shows *a*) that there was diffuse increase in the wall thickness (red arrow) in the descending aorta, the aortic wall was concentrically thickened. *b*) Aortic wall thickening progressively increased and the diameter of the thoracic aorta decreased abruptly (red arrow) at the level of the diaphragmatic hiatus up to coeliac trunk in axial slices. *c*) There is abrupt decrease (yellow arrow) in aorta up to coeliac trunk in 3D surface shaded image and MIP image.

to show activation of the vasculitis. However, PET scintigraphy was suggestive for active vasculitis. CT angiography showed the inflammation of the whole abdominal aortic wall and these findings indicated large vessel vasculitis. In genetic evaluation, heterozygous p. Thr68Met (c.203C>T) change was detected in the PSTPIP1 gene, which was previously shown in paediatric-onset Behçet's disease.[5] We experienced a diagnostic dilemma; was it large vessel vasculitis? or Behçet's disease? Because of extensive vascular involvement, coexistence of stenosis and thrombosis, and genetic features, it was accepted as Behçet's disease. Although "pathergy test, HLA B5 and HLA B51" are negative, there are children diagnosed with Behçet's disease with vascular findings in the literature.[6]

Arterial lesions in Behçet's disease are seen in the form of stenosis, occlusions, aneurysms, or aortitis.[7] *Kuzu* et al[8] reported arterial aneurysm in 1.6% of 1200 patients. They found arterial stenosis in only three patients. Abdominal aortic stenosis has not been described previously in paediatric-onset Behçet's disease. In 2018, a 20-year-old female with paediatric-onset Behçet's disease was reported with bilateral subclavian arterial stenosis.[9] Nakamura et al[10] reported a 18-year-old girl paediatric-onset Behçet's disease case with a widespread narrowing of the carotid, vertebral, renal, subclavian arteries, and the abdominal aorta. Arterial stenotic lesions mimicking Takayasu arteritis, though fairly reported in adults, are previously not described in paediatric-onset Behçet's disease.

Treatment of vascular Behçet's disease includes combination therapy of systemic corticosteroid and immunosuppressants. Recently, recombinant interferon-alpha 2a and anti-TNF-α antiagents have been reported as an alternative treatment for management of vascular Behçet's disease.[1] There is no agreement for anticoagulation in case of thrombosis related to Behçet's disease. Due to risk of recurrences, immunosuppressants should be the first choice of drug before surgical excision of thrombosis. Presence of arterial stenotic lesions indicates a worse prognosis and requires long-term immunosuppressive therapy. The treatment choices of abdominal aortic stenosis include surgical patch aortoplasty, thoracoabdominal bypass, and endovascular angioplasty with stenting; however, there are risks for thrombosis of the graft and relapses of aneurysms at the site of bypass.

Conclusion

Behçet's disease should be kept in mind in vascular findings in childhood, even if there is no classical clinical triad of Behçet's disease. The stenosis of the abdominal aorta was described in our paediatric-onset Behçet case for the first time.

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Prof. Dr Filiz Ekici: Concept/design, data analysis/interpretation, drafting article, critical revision of article.

Assoc. Prof. Dr Elif Çomak: Data analysis/interpretation.

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