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Juvenile Takayasu's arteritis with aortic dissection as initial presentation

Młodzieńcze zapalenie tętnic Takayasu u dziecka z rozwarstwieniem aorty jako pierwszym objawem choroby

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A 6-year-old girl presented for evaluation of low-grade fever, loss of appetite and weight, and breathlessness of six weeks' duration. On examination, blood pressure in left arm was 124/88 mm Hg, and in right arm was 80/58 mm Hg, while pulses in both lower limbs were weak. According to these observations, a diagnosis of Takayasu's arteritis was established. Haemogram was within normal range except for mild anaemia (Hb 9.8 mg/dL), elevated erythrocyte sediment rate (49 mm/h), and raised C-reactive protein (CRP 99 mg/L). Chest radiography was normal. Electrocardiogram revealed left ventricular hypertrophy with sinus tachycardia. Echocardiography showed dilated left atria and ventricle, global hypokinesia with severe impairment of systolic function [ejection fraction (EF) 20%]. Multidetector computed tomography scan (MDCT) of aorta showed mild diffuse narrowing of thoraco-abdominal segment with severe narrowing of distal abdominal segment in infra-renal segment (Figure 1A, B). Total occlusion of right brachio-cephalic trunk and critical ostial stenosis of left common carotid artery were also seen. A spiral flap suggesting aortic dissection (DeBakey type II; Stanford type III) from the descending aorta (T7–T8 vertebral level) to the distal abdominal aorta was also seen, which was further confirmed on contrast aortogram (Figures 1, 2). Therefore, she was clinically diagnosed as Takayasu's arteritis (TA) with aortic dissection (DeBakey type II) based on the European League against Rheumatism (EULAR)/Paediatric Rheumatology European Society (PRES) criteria [1]. Patient succumbed to death because of progressive failure and ventricular tachycardia.

Takayasu's arteritis, first reported by Mikito Takayasu in 1908, causes segmental stenosis, dilatation, or aneurysm formation which generally affects the large vessels [2]. Although it can be diagnosed and classified on the basis of EULAR criteria, assessment of the inflammatory status and pattern of vascular involvement is still a major challenge [3]. Clinically, TA activity can be assessed by acute-phase reactants (ESR or CRP), new bruits, and new angiographic features. TA causes persistent inflammation of the aorta, but dissection is exceedingly rare because of the dense adventitial fibrosis and intimal scarring, though decreased wall elasticity and aortic mobility due to fibrous adhesion between fibrous adventitia and surrounding tissue may be the reason for dissection in TA [4, 5]. Arteriosclerosis, long-term hypertension, dyslipidaemia, and associated autoimmune connective tissue disorders may be other antecedent factors.

In our case, the patient was a young girl admitted to hospital with aortic dissection as the initial presentation and diagnosed with TA after further evaluation. As it was a type III DeBakey dissection, the patient was managed with medical treatment including decongestive therapy. Also, she was in the active phase of the disease; before glucocorticoid therapy and immunosuppressive therapy could be started she succumbed to death because of ventricular arrhythmia. This was also the reason why thoracic endovascular aortic repair was not ventured, because it carries great risks during the active (progressive) stage of TA.

In general, TA patients with severe ischaemic symptoms (limb claudication, heart failure, *etc.*) require endovascular intervention or vascular surgery in addition to medical therapy, although the results are conflicting regarding rates of restenosis and occlusion [6, 7].

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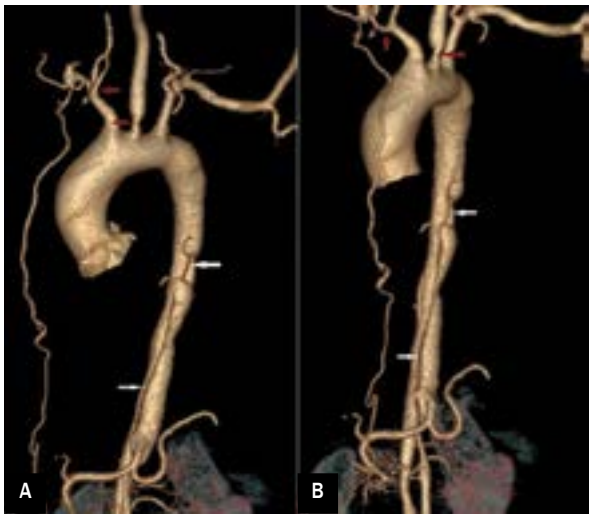

Figure 1A, B. Multidetector computed tomography scan (MDCT) of aorta showing total occlusion of right brachio-cephalic trunk and critical ostial stenosis of left common carotid artery (red arrow), diffuse narrowing of thoraco-abdominal segment with severe narrowing of distal abdominal aorta involving infra-renal segment. Also seen is spiral flap (white arrow) suggesting aortic dissection (DeBakey type II)

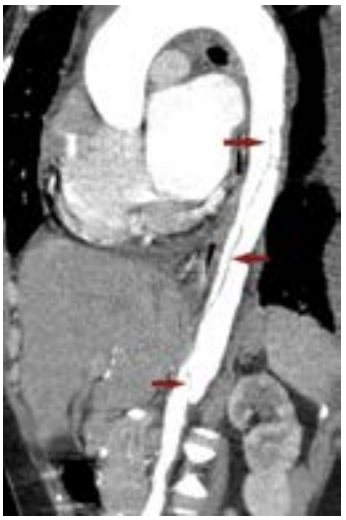

Figure 2. Contrast aortogram showing spiral flap suggesting aortic dissection of descending aorta (DeBakey type II; Stanford type III)

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