

# Difficult diagnosis of Kawasaki disease in a patient with giant coronary artery aneurysms

Choroba Kawasaki u pacjenta z olbrzymimi tętniakami tętnic wieńcowych

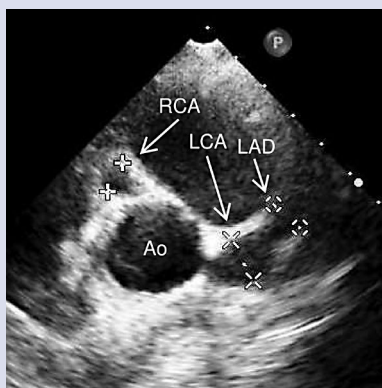
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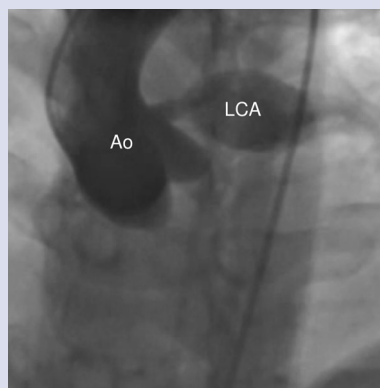
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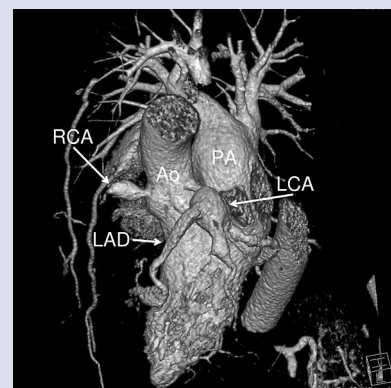
A 14-year-old girl with facial dysmorphism (large, low-set ears, exotropia, and anodontia), hearing impairment, mild mental retardation, and epilepsy in anamnesis without genetic diagnostics, was admitted to the paediatric ward with gastroenteritis, pneumonia, pharyngitis, high persistent fever, and polymorphous rash, without improvement after administration of antibiotics and anti-inflammatory drugs. Because of deterioration of condition and multiorgan failure symptoms, the girl was referred to the intensive care unit, where she was qualified for mechanical ventilation. In addition, suppurative conjunctivitis and reddening of palms were noticed. Despite aggressive antibiotic and anti-inflammatory therapy, her state did not improve. Transthoracic echocardiography revealed left coronary artery (LCA) aneurysm of 10 mm in diameter, left anterior descending artery (LAD) aneurysm of 6 mm in diameter, and right coronary artery (RCA) aneurysm of 5 mm in diameter (Fig. 1), which led to suspicion of Kawasaki disease (KD). After administration of gamma-globulin (2 g/kg) and aspirin (50 mg/kg) her symptoms eventually yielded. Two months after onset of symptoms the aortography showed the following coronary artery aneurysms (CAA): two saccular in RCA (diameters: 8 mm both), saccular in LCA (13 mm), and fusiform in circumflex branch (Cx) (8 mm). Moreover, left common iliac artery and superior mesenteric artery aneurysms were diagnosed. During almost four-year follow-up she has been chronically treated with aspirin (150 mg) and acenocoumarol (INR 2–2.5). Two years after diagnosis heart scintigraphy did not show myocardial perfusion defect (EF 54%) but LCA revealed two-fold expansion during another aortography (25 mm) (Fig. 2). Almost four years after the occurrence of the aneurysms computed tomography revealed saccular aneurysms of LCA: 25 × 17 mm and RCA: 13 × 10 mm, and fusiform aneurysms of LAD: 11 mm and Cx: 12 mm (Fig. 3). Despite such coronary artery malformations, she was almost asymptomatic (10.9 MET in treadmill exercise test). KD, which is acute systemic vasculitis of small- and medium-sized arteries of unknown aetiology, has become the leading acquired heart disease in developed countries. Five of the six principal features have to be present to diagnose KD (persistent fever — mandatory, conjunctival injection without exudate, changes of the mucosa of oropharynx and lips, changes in peripheral extremities, polymorphous exanthema, and cervical lymphadenopathy), or less but with CAA occurrence — incomplete KD (as in our case). Transient CAA are observed in up to 40% of patients in acute phase of KD, but they remain only in about quarter of these patients. Giant CAA, which account for less than 10% of chronic CAA, do not regress and bring the worst prognosis. Thirty-year mortality of giant CAA patients is higher than 10%.



**Figure 1.** Transthoracic echocardiography in acute phase of Kawasaki disease; left coronary artery (LCA; 10 mm), left anterior descending artery (LAD; 6 mm), right coronary artery (RCA; 5 mm) aneurysms; Ao — aorta



**Figure 2.** Aortography two years after onset of Kawasaki disease; left coronary artery (LCA; 25 mm) aneurysm; Ao — aorta



**Figure 3.** Computed tomography four years after onset of Kawasaki disease; left coronary artery (LCA; 25 × 17 mm), left anterior descending artery (LAD; 11 mm), right coronary artery (RCA; 13 × 10 mm) aneurysms; Ao — aorta

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#### Conflict of interest: none declared

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