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A 19-year-old girl with ostial coronary lesions

Dziewiętnastoletnia pacjentka ze zmianami w obrębie ujścia tętnic wieńcowych

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A 19-year-old girl presented with retrosternal chest pain at rest associated with nausea and vomiting. She had excertional chest pain of three days' duration. She had no past history of hypertension, diabetes or smoking. Her vital signs were stable. She weighed 53 kg and was 156 cm tall. Her body mass index was 21.7 kg/m^2. On physical examination, she had extensive xanthomatosis on the extensor surfaces (knees and elbows), her buttocks and ankles, and swelling and erythema over her first and fifth metatarsophalangeal joints (Figs. 1, 2). There was a history of sudden death in her only sibling, an 18-year-old brother several years earlier. ECG showed diffuse and down-sloping ST segment depression in inferior and precordial leads along with ST segment elevation of 3 mm in avR (Fig. 3). Anti-ischaemic therapy was initiated and the patient was admitted to the Coronary Care Unit. Echocardiography showed global left ventricular ejection fraction of 45% with hypoplastic aortic root without aortic stenosis. Laboratory studies showed elevation of cardiac troponin I and creatinine kinase-MB. Lipid profile was total cholesterol of 570 mg/dL, HDL of 30 mg/dL, LDL of 522 mg/dL. Triglycerides levels were normal. She was scheduled for immediate coronary angiography. Non selective contrast injection in aortic root showed narrowing of aortic root and ostial lesion in the left main artery (Fig. 4). Right coronary injection showed ostial lesion in the right coronary artery. The patient had episodes of chest pain despite medical therapy and was scheduled for urgent coronary artery bypass grafting (CABG). During surgery, both coronary arteries had ostial lesions and CABG was performed. The patient was prescribed lipid-lowering therapy and referred for LDL aphresis on discharge. At six months follow-up, the patient remains well. Multiple xanthomas, high serum levels of LDL, and premature atherosclerosis are indicative of familial hypercholesterolaemia.

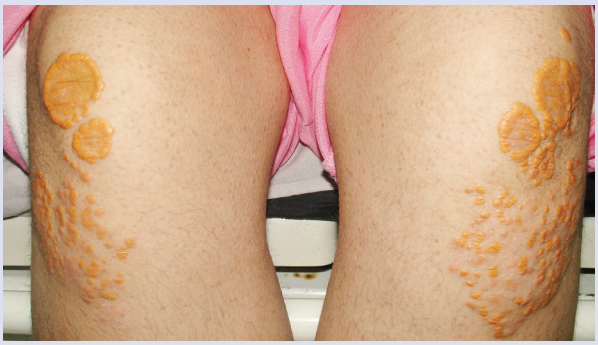

Figure 1. Patient's knees showing diffuse xanthomatic lesions

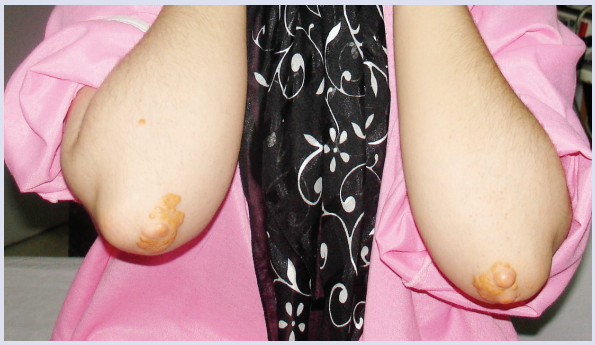

Figure 2. Xanthomas over extensor surface of upper extremities

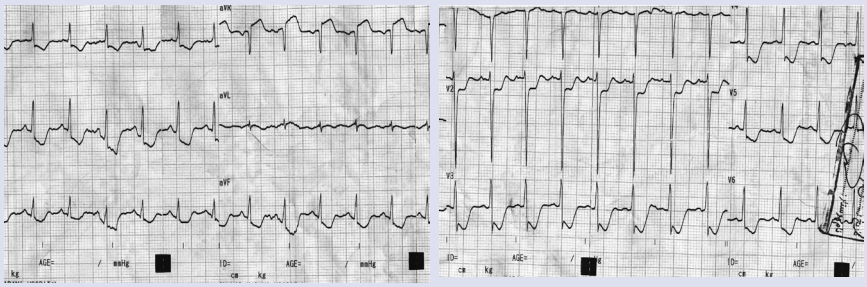

Figure 3. ECG showing diffuse and down-sloping ST segment depression in inferior and precordial leads with ST segment elevation in avR

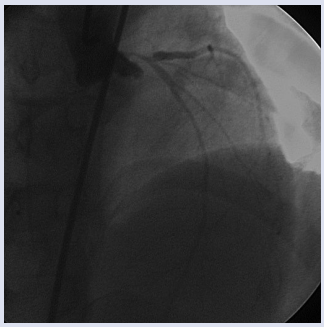

Figure 4. Non selective contrast injection in aortic root showing narrowing of aortic root and ostial lesion in left main artery

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Conflict of interest: *none declared*