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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Young woman with idiopathic supradiaphragmatic mid-aortic syndrome and hypertension successfully controlled with a beta-blocker–calcium antagonist fixed-dose combination

Młoda kobieta z nadprzeponowym zespołem aorty i nadciśnieniem tętniczym skutecznie leczonym preparatem złożonym beta-adrenolityk–antagonista wapnia

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Mid-aortic syndrome (MAS) is a rare cause of hypertension. A 19-year-old woman was admitted to the Hypertension Department for evaluation after elevated systemic blood pressure (BP) was disclosed 3 years previously with values up to 220/120 mm Hg and treated unsuccessfully with metoprolol. She was asymptomatic, with a negative family history for hypertension. Physical examination was unremarkable except BP values 184/106 mm Hg at admission. ABPM revealed 24-h mean BP 156/90 mm Hg and night mean BP 136/79 mm Hg. BP load was 96.7/100% (Fig. 1). Routine laboratory work, ECG, chest radiogram and echocardiography was normal with no evidence of coarctation. Renal ultrasound showed normal kidneys with incorrect spectrum of renal arteries blood flow without features of renal artery stenosis. Computed tomography angiography (CTA) showed normal aortic arch and its branches. The descending aorta showed smooth narrowing in its middle segment from 21 mm to 15 mm with poststenotic dilatation. Three-dimensional (3D) CTA revealed 3.4 cm — long thickening of aortic wall with 70–80% stenosis of aortic lumen, started 9 cm above diaphragm (Fig. 2). The Adamkiewicz's artery could not be identified. Magnetic resonance angiography (MRA) confirmed the presence of a mass with extensive area of signal void (likely massive calcification) in aortic wall narrowing aortic lumen to 8 × 10 mm (Figs. 3, 4). Abdominal aorta and renal arteries were normal, as well as both suprarenal glands. Ankle-brachial index (R: 0.96, L: 0.93) indicated normal arterial flow in lower extremities. Biochemical tests for pheochromocytoma were negative. Surprisingly, plasma renin activity (PRA 0.19 ng/mL/h) was low and plasma aldosterone level (1421.4 pg/mL) was normal. Potential secondary causes of restricted *vasulitis* (tuberculosis, syphilis, autoimmunological diseases) and calcification (abnormal calcium-phosphate metabolism) were excluded. Antinuclear antibodies, ANA, cANCA, pANCA, QuantiFERON-TB Gold and VDRL tests, serum calcium, phosphate and parathormone level were within normal range. Idiopathic supradiaphragmatic MAS with secondary hypertension was diagnosed. As the female patient was of reproductive age and had low PRA, antihypertensive therapy with a fixed dose combination of bisoprolol 5 mg with amlodipine 5 mg was introduced. HBPM and second ABPM after 4 weeks showed good BP control: 24-h mean BP 128/69 mm Hg and night mean BP 125/67 mm Hg and BP load was 8.5/2.4% (Fig. 5). Uncontrolled hypertension, kidney survival at risk, claudication and intestinal ischaemia are indications for surgical or endovascular treatment of MAS. As this was not the case here, a decision was made to delay surgical correction, with regular follow-up for BP control, renal function and aortic stenosis lesions.

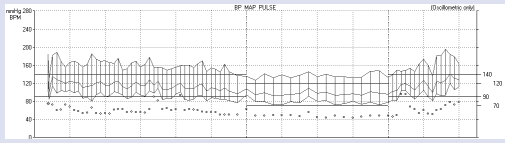

Figure 1. Initial ABPM

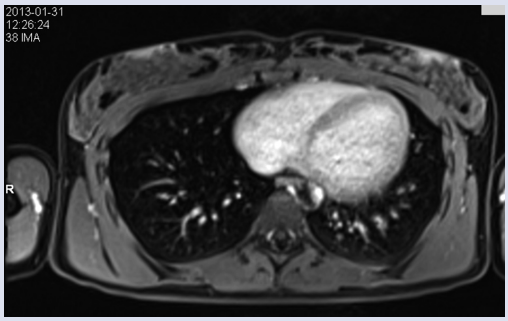

Figure 4. MRA: transverse section revealed aortic stenosis

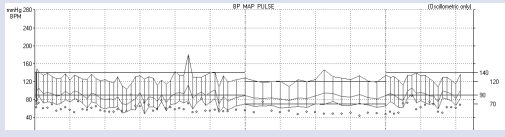

Figure 5. Second ABPM

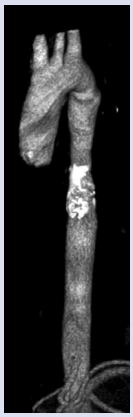

Figure 2. CTA of aorta; 3D reconstruction

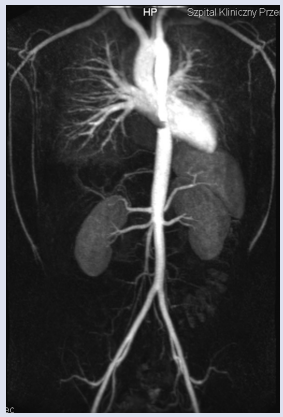

Figure 3. MRA of aorta and its branches

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Conflict of interest: *A. Tykarski — lectures for Merck*