Chronic thromboembolic pulmonary hypertension complicated by left main compression syndrome

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A 55-year-old-female was admitted with exertional dyspnea (World Health Organization functional class [WHO-FC] III/IV). Clinical evaluation revealed increased N-terminal-pro-brain-type natriuretic peptide (NT-proBNP, 3994 pg/ml) and reduced the six-minute walk test (6MWT, 330 meters). Echocardiography showed enlargement of the right atrium (area, 26.4 cm^2) and right ventricle (RV, 54 mm) with high-velocity tricuspid regurgitation (4.7 m/s), pulmonary artery (PA) dilatation (80 mm), elevated estimated RV systolic pressure (96 mm Hg) and mean pulmonary arterial pressure (mPAP, 56 mm Hg). Electrocardiogram indicated RV hypertrophy and overload (Supplementary material, *Figure S1*). Magnetic resonance imaging ruled out congenital heart defects. The right heart catheterization (the thermodilution method) indicated: mPAP of 58 mm Hg, PA wedge pressure of 12 mm Hg, pulmonary vascular resistance (PVR) of 10.8 Wood units (Wu), and cardiac index of 2.6 l/min × m^2. The patient's pulmonary angiography (Axiom Artis Zee, Siemens, Germany) and computed tomography scan (LightSpeed VCT 64 scanner GE, Chicago, IL, US) revealed a large main PA aneurysm (PAA, 86 mm), along with right and left PAAs (69 mm), and multiple organized thromboembolic lesions in lobar and segmental arteries, which allowed the diagnosis of chronic thromboembolic pulmonary hypertension (CTEPH) (Figure 1A–D). Coronary angiography demonstrated severe left main coronary artery (LMCA) stenosis due to compression by PAA, confirmed by intravascular ultrasound (IVUS, Eagle Eye Platinum, Philips, Netherlands) (Figure 1E, Supplementary material, *Video S1*).

The case was carefully analyzed by the multidisciplinary CTEPH team, and the patient was not qualified for endarterectomy with PA reconstruction due to high risk. The combined pulmonary vasodilators, along with LMCA percutaneous intervention (PCI), were offered for the patient as bridging therapy before lung transplantation (LTx). PCI was performed and the LMCA ostium was expanded with a 4.0 mm × 28 mm drug-eluting stent (Synergy Megatron™, Boston Scientific, Marlborough, MA, US) and post-dilated with a 5.0 mm × 15 mm balloon (Pantera Pro, Biotronik, Berlin, Germany). The final angiogram and IVUS showed good stent expansion with relief of LMCA extrinsic compression (Figure 1F, Supplementary material, *Video S2*). The combined specific treatment was also subsequently introduced. The patient was started on oral riociguat titrated from 3 mg to 7.5 mg daily, along with a continuous subcutaneous infusion of treprostinil titrated to the maximum tolerated dosage (30 ng/kg/min).

A six-month follow-up examination demonstrated a significant reduction of mPAP (47 mm Hg) and PVR (6.25 Wu) without PAA diameter progression. The patient improved to II/III WHO-FC with a 390 m distance at 6MWT and NT-proBNP of 1007 pg/ml.

We describe a rare case of CTEPH-related PAA causing LMCA compression. In CTEPH, PAA seems to be associated with mural thickening, webs, or intramural calcified thrombi [1]. PAA may lead to life-threatening complications, including PAA dissection or acute coronary syndrome [2, 3]. To date, the optimal treatment of PAA has not been established. There are no criteria indicating urgent recon-

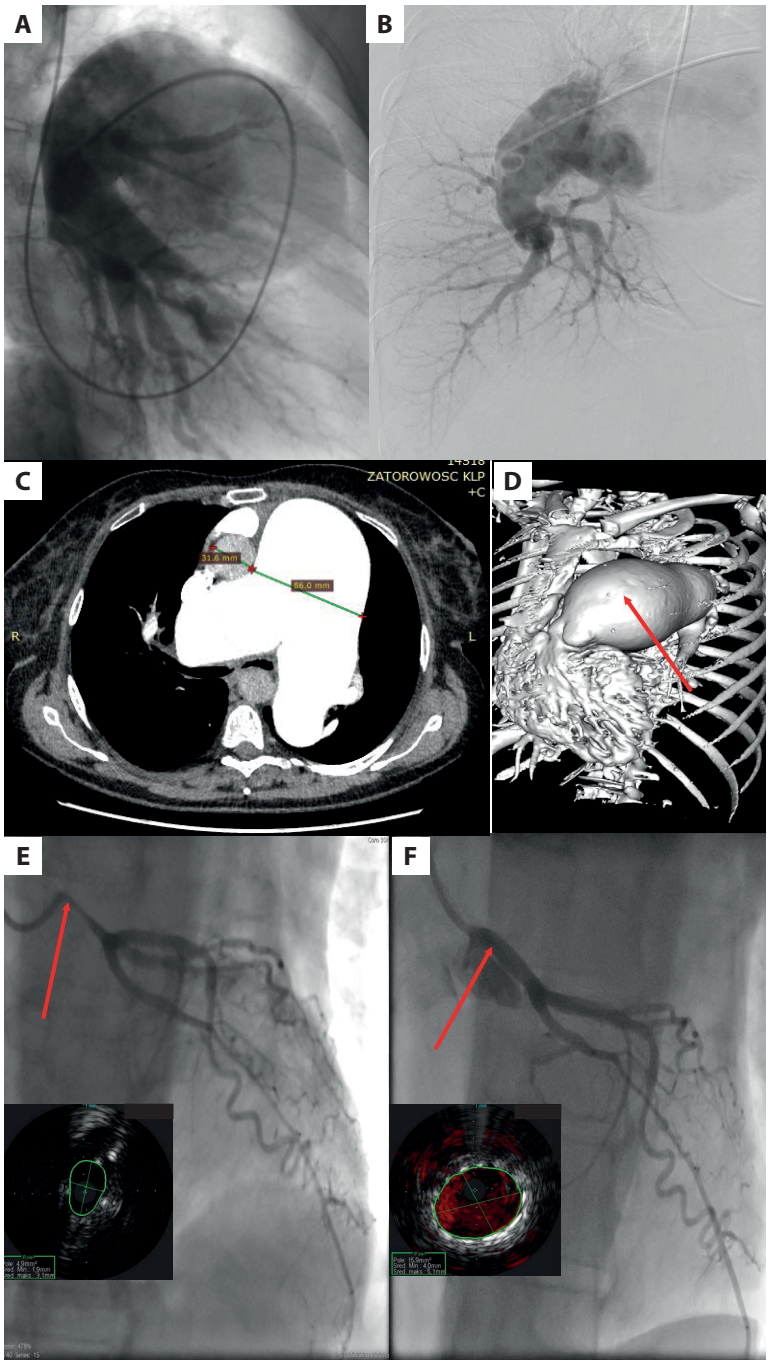

Figure 1. A, B. Angiography of the left (**A**) and right (**B**) pulmonary artery (PA) showing dilatation and calcified thromboembolic lesions. **C, D.** Computed tomography scan (**C**) with volume rendering reconstruction (**D**) showing a pulmonary artery aneurysm (PAA) (the arrow). **E.** Coronary angiography (CA) with an intravascular ultrasound (IVUS) showing critical stenosis of the left main coronary artery (LMCA) attributed to the PAA compression (the arrow). LMCA minimal lumen area (MLA) was 4.9 mm^2. **F.** CA with IVUS after LMCA stenting, with normal LMCA shape and size (the arrow). LMCA MLA after stenting was 15.9 mm^2

structive surgery or waiting for LTx, especially in asymptomatic patients [1, 3]. In the case of LMCA, the accumulated experience suggests stenting might be a first-choice strategy [3, 4]. Recently, treprostinil was proven to increase the exercise capacity in CTEPH [5]. In the presented case, LMCA stenting and pulmonary vasodilators led to the improvement of the patient's hemodynamic status.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

Conflict of interest: None declared.

REFERENCES

1. Kreibich M, Siepe M, Kroll J, et al. Aneurysms of the pulmonary artery. Circulation. 2015; 131(3): 310–316, doi: 10.1161/CIRCULATIONAHA.114.012907, indexed in Pubmed: 25601950.
2. Maj D, Kopiec T, Wieteska M, et al. Acute coronary syndrome due to extrinsic left main compression. Kardiol Pol. 2021; 79(9): 1034–1035, doi: 10.33963/KP.a2021.0045, indexed in Pubmed: 34166517.
3. Florczyk M, Wieteska M, Kurzyna M, et al. Acute and chronic dissection of pulmonary artery: new challenges in pulmonary arterial hypertension? Pulm Circ. 2018; 8(2): 2045893217749114, doi: 10.1177/2045893217749114, indexed in Pubmed: 29251549.
4. Demkow M, Kalińczuk Ł, Kepka C, et al. Left main artery compression by pulmonary artery aneurysm and ostial athero-stenosis of left anterior descending artery in a young female with pulmonary arterial hypertension. Eur Heart J. 2012; 33(20): 2621, doi: 10.1093/eurheartj/ehs195, indexed in Pubmed: 22782942.
5. Sadushi-Kolici R, Jansa P, Kopec G, et al. Subcutaneous treprostinil for the treatment of severe non-operable chronic thromboembolic pulmonary hypertension (CTREPH): a double-blind, phase 3, randomised controlled trial. Lancet Respir Med. 2019; 7(3): 239–248, doi: 10.1016/S2213-2600(18)30367-9, indexed in Pubmed: 30477763.