Pulmonary embolism as a clinical manifestation of right ventricular intimal sarcoma

Alicja Genc[1], Izabela Pisowodzka[1], Monika Lica-Gorzynska[2], Andrzej Łoś[3], Jan Rogowski[3], Marcin Fijałkowski[1]

1 1st Department of Cardiology, Medical University of Gdańsk, Gdańsk, Poland
2 Department of Cardiology, District Hospital, Chojnice, Poland
3 Department of Cardiac and Vascular Surgery, Medical University of Gdańsk, Gdańsk, Poland

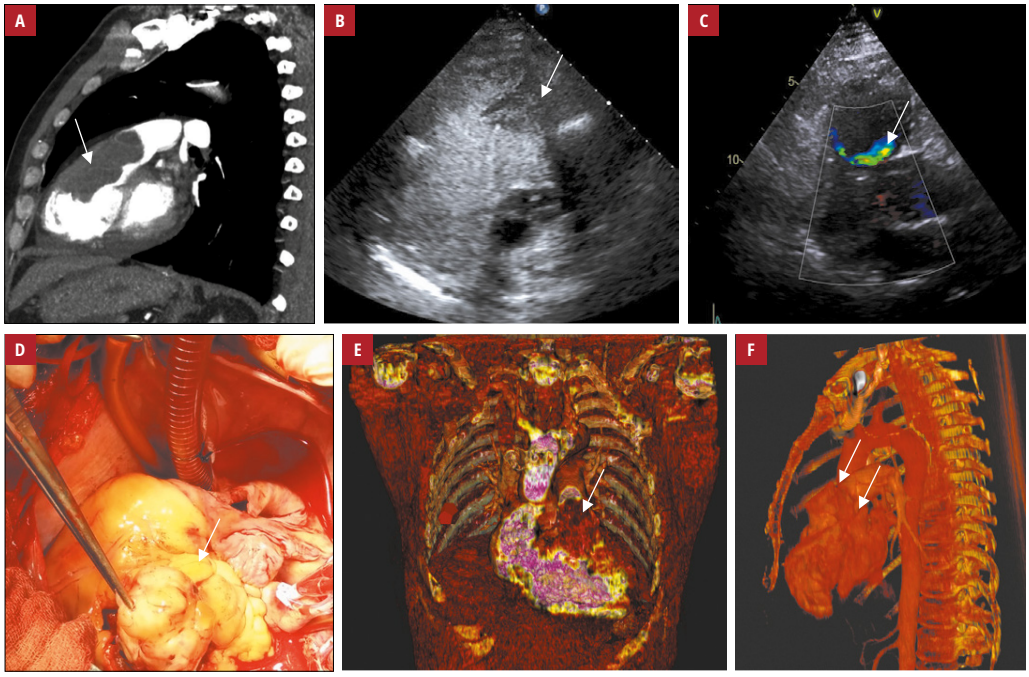

FIGURE 1 **A** – sagittal computed tomography scan showing a hypodense neoplasm filling the pulmonary trunk and the right ventricular outflow tract (arrow); **B** – contrast transthoracic echocardiography showing the negative contrast effect in the right ventricular outflow tract (arrow); **C** – color Doppler echocardiography showing residual blood flow in the right ventricle (white arrow); **D** – photography done during the surgery showing the excised tumor (white arrow); **E** – 3D visualization (Vesalius3D, PS-Medtech, Amsterdam, the Netherlands) from computed tomography scans without contrast showing neoplasm filling the pulmonary trunk and the right ventricular outflow tract (arrow); **F** – 3D visualization (Vesalius3D) from computed tomography scans with contrast showing the residual blood flow (arrows) caused by the huge neoplasm filling almost the entire right ventricle.

Correspondence to:
Alicja Genc, MD,
1st Department of Cardiology,
Medical University of Gdańsk,
ul. Dębinki 7, 80-952 Gdańsk,
Poland, phone: +48 58 349 25 00,
email: alicja.genc@gumed.edu.pl
Received: December 17, 2020.
Revision accepted:
January 20, 2021.
Published online:
January 27, 2021.
Kardiol Pol. 2021; 79 (3): 348-349
doi:10.33963/KP.15787

A 47-year-old man was admitted to the hospital with a 2-month history of increasing dyspnea and fatigue and a 2-week history of edema of the right leg. He denied weight loss, fever, and signs of infection. Laboratory tests showed slightly elevated C-reactive protein (8.9 mg/l; reference range, <5 mg/l) and D-dimer (0.72 μg/ml; reference range, <0.5 μg/ml) levels. The patient underwent computed tomography angiography, which showed a large hypodense mass in the right atrium and the pulmonary trunk (PT). Transthoracic echocardiography showed that both atria and the right ventricle (RV) were enlarged, indicating RV pressure

overload, with moderate tricuspid regurgitation. Based on the clinical and radiological features, pulmonary embolism (PE) was diagnosed as the most likely disease. The patient was prescribed dabigatran 150 mg twice a day. Six weeks later, he was readmitted to the hospital with symptoms of RV failure. Computed tomography revealed a massive hypodense neoplasm, filling the RV and the proximal segment of the PT and allowing only a narrow parietal blood flow (FIGURE 1A). Transthoracic echocardiography showed a high RV pressure overload, a negative contrast effect in the RV outflow tract and PT, and a residual contrast within the right atrium and RV (FIGURE 1B; Supplementary material, *Videos S1* and *S2*). Another color Doppler echocardiogram confirmed the residual flow (FIGURE 1C). Because the patient's dyspnea and presyncope were triggered by minimal physical activity, urgent cardiac surgery was performed. A huge tumor was excised, which filled the RV and invaded the myocardium (FIGURE 1D). Furthermore, De Vega tricuspid annuloplasty was done. Histopathological examination corresponded with intimal sarcoma, and this was confirmed by the Harvard University laboratory. The patient was then referred for adjuvant oncological treatment.

Pulmonary artery sarcoma (PAS) is a rare and extremely malignant tumor, first reported in 1923 by Mandelstamm as quoted by Mussot et al.[1] It often mimics pulmonary vascular disease, and therefore, some cases are initially misdiagnosed as PE, which can lead to a 3- to 12-month delay before a correct diagnosis is established.[1,2] The analyzed median (SD) survival was 36.5 (20.2) months for patients who undergo a curative resection and 11 (3) months for those with an incomplete resection.[2] The tumor appears to arise from multipotential mesenchymal cells of the muscle anlage of the bulbus cordis, the embryologic structure that gives rise to the PT.[3] Most often, the tumor is found in the PT and its branches; nevertheless, it can extend proximally into the RV as its outflow tract originates from the same structure (up to 100% and 25% of cases, respectively).[3,4] Radiological images may reveal differences between PAS and thromboembolic disease, as the former is generally bulky, unilateral, and located centrally.[3] Nevertheless, PE may also coexist with tumors, both malignant and benign.[5]

We report here on a patient with a massive tumor and hardly any flow in the RV who required urgent surgery, and it was vital that we were able to differentiate between PE and PAS. This case confirms the usefulness of a multimodal approach, especially with the use of contrast echocardiography and 3D visualization (FIGURE 1E and 1F; Supplementary material, *Videos S3* and *S4*).

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

NOTE This case report was presented as an abstract at the 13th Polish Society of Cardiology International Congress on September 26 to 28, 2019 in Katowice, Poland.

CONFLICT OF INTEREST None declared.

HOW TO CITE Genc A, Pisowodzka I, Lica-Gorzynska M, et al. Pulmonary embolism as a clinical manifestation of right ventricular intimal sarcoma. Kardiol Pol. 2021; 79: 348-349. doi:10.33963/KP.15787

REFERENCES

1 Mussot S, Ghigna MR, Mercier O, et al. Retrospective institutional study of 31 patients treated for pulmonary artery sarcoma. Eur J Cardiothorac Surg. 2013; 43: 787-793.

2 Blackmon SH, Rice DC, Correa AM, et al. Management of primary pulmonary artery sarcomas. Ann Thorac Surg. 2009; 87: 977-984.

3 Cox JE, Chiles C, Aquino SL, et al. Pulmonary artery sarcomas: a review of clinical and radiologic features. J Comput Assist Tomogr. 1997; 21: 750-755.

4 Bleisch VR, Kraus FT. Polypoid sarcoma of the pulmonary trunk: analysis of the literature and report of a case with leptomeric organelles and ultrastructural features of rhabdomyosarcoma. Cancer. 1980; 46: 314-324.

5 Szerszyńska A, Nowak R, Łaskawski G, Fijałkowski M. Recurrent pneumonia and pulmonary embolism in a young patient as a presentation of right ventricular myxoma. Kardiol Pol. 2019; 77: 63.