From a tumor in the right ventricle to hypereosinophilic syndrome diagnosis

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A 51-year-old woman with no previous cardiac history was hospitalized due to severe dyspnea, weight loss, and lower extremity edema. Physical examination also revealed bilateral pleural effusion and ascites. The initial laboratory tests showed hemoglobin levels of 13.5 g/dl (reference range, 12.0–16.0 g/dl), platelet count of 219 ×10^3/µl (reference range, 150–400 ×10^3/µl), and C-reactive protein levels of 2.65 mg/l (reference range, 0–5 mg/l). Transthoracic echocardiography (Philips HD15 PureWave, Amsterdam, the Netherlands) showed hypokinesis of the left (ejection fraction 30%) and right ventricle (RV), enlargement of the right atrium, and a tumor in the RV obliterating most of its cavity (FIGURE 1A, Supplementary material, *Video S1*). The image was confirmed by transesophageal echocardiography (Supplementary material, *Video S2*). The patient underwent further detailed evaluation, and the peripheral hypereosinophilia (eosinophil count, 3.8 ×10^3/µl, percentage of total leukocytes, 39.4% [1%–5%]) was noted. Eosinophilic infiltration in pleural effusion and the bone marrow was detected. Secondary causes of eosinophilia were excluded (parasites, allergies, reactive eosinophilia, malignancy). Differential diagnosis included eosinophilic granulomatosis with polyangiitis and idiopathic hypereosinophilic syndrome (HES)—the main differences are shown in Supplementary material, *Table S1*. Based on cardiac magnetic resonance (CMR) (Achieva 1.5T, Philips), the RV tumor was classified as a large thrombus (53 ×17 ×12 mm) (FIGURE 1B and Supplementary material, *Video S3*). Restriction, mainly of the RV with endocardial edema and fibrosis, was also observed on CMR. Hypereosinophilic syndrome was diagnosed with possible Löffler endocarditis (the patient did not agree to undergo RV endocardial biopsy). Anticoagulation was started with heparin and then switched to warfarin. The standard treatment for heart failure with a diuretic, angiotensin-converting enzyme inhibitor, and β-bloker, was also introduced. Hypereosinophilic syndrome was treated with steroids. Then hydroxycarbamide was added, which led to normalization of the eosinophil count. After 3 months of treatment, echocardiography showed RV thrombus resolution and moderate improvement in the function of the left ventricle (ejection fraction 36%) and RV with persisting fibrosis (FIGURE 1C, Supplementary material, *Video S4*) consistent with CMR imaging (FIGURE 1D). Currently, the patient is alive in 1 year follow-up.

Hypereosinophilic syndrome is a diagnosis of exclusion.[1-3] This rare disorder characterized by unexplained peripheral blood eosinophilia (>1.5 ×10^3/µl) and multiorgan system dysfunction occurs most frequently in young to middle-aged men. Interestingly, according to our best knowledge, this is the first case of HES with severe RV involvement in a woman described in the literature. Cardiac manifestation occurs in about 50% of HES cases and is the major cause of morbidity.[1] It is classified in 3 stages: myocardial necrosis caused by eosinophilic infiltration, thrombotic formation, and a fibrotic stage with the development of restrictive cardiomyopathy. The hallmark echocardiographic feature is the obliteration of the ventricular apex by the mural thrombus, which usually involves the left or both cardiac chambers or, less often, as in our case, predominantly the RV. Cardiac

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Received: November 16, 2019.
Review accepted: December 4, 2019.
Published online: December 4, 2019.
Kardiol Pol. 2020; 78 (1): 89-90
doi:10.33963/KP.15084

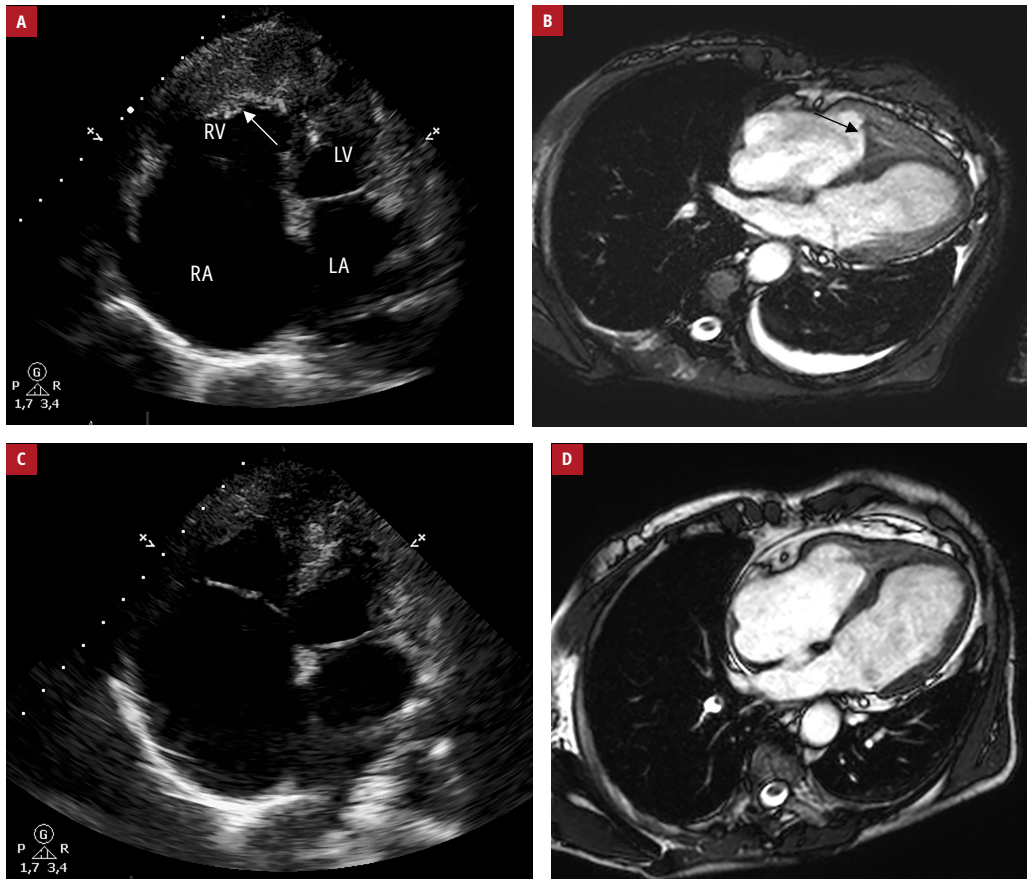

FIGURE 1 Transthoracic echocardiography (**A**) and cardiac magnetic resonance (**B**) showing thrombus obliterating the right ventricle (arrows). Transthoracic echocardiography (**C**) and cardiac magnetic resonance (**D**) showing dissolved right ventricular thrombus

Abbreviations: LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

magnetic resonance in our patient confirmed RV thrombus, inflammation (an increased T2 signal was present), and myocardial fibrosis on late gadolinium enhancement imaging, all of which confirmed Löffler endocarditis.

In conclusion, the association of HES with cardiac thrombosis requires exclusion of hypereosinophilia in the case of an isolated ventricular mural thrombus, and vice versa, after the detection of unexplained increased eosinophil counts, one should consider echocardiography screening and look for intracardiac thrombi, as their presence requires urgent medical treatment.

SUPPLEMENTARY MATERIAL

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

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

HOW TO CITE Styczkiewicz K, Mędrek S, Kostkiewicz A, et al. From a tumor in the right ventricle to hypereosinophilic syndrome diagnosis. Kardiol Pol. 2020; 78: 89-90. doi:10.33963/KP.15084

REFERENCES

1 Jin X, Ma C, Liu S, et al. Cardiac involvements in hypereosinophilia-associated syndrome: case reports and little review of the literature. Echocardiography. 2017; 34: 1242-1246.

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

3 Lipczyńska M, Klisiewicz A, Szymański P, et al. [Not only after myocardial infarction – left intraventricular thrombus in the Churg–Strauss syndrome]. Kardiol Pol. 2010; 68: 836-837.