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Authors: Katarzyna Styczkiewicz, Sabina Mędrek, Agnieszka Kostkiewicz, Marek Styczkiewicz, Michał Włodyka, Magdalena Lipczyńska

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From right ventricle tumor to hypereosinophilic syndrome diagnosis

Katarzyna Styczkiewicz¹, Sabina Mędrek¹, Agnieszka Kostkiewicz², Marek Styczkiewicz¹, Michał Włodyka¹, Magdalena Lipczyńska³

¹Department of Cardiology, Brzozów Specialist Hospital, Subcarpathian Oncological Center, Brzozów, Poland
²Clinical Department of Radiology, Provincial Hospital No. 2, Rzeszów, Poland
³Adult Congenital Heart Disease Department, Institute of Cardiology, Warsaw, Poland

Correspondence:
Katarzyna Styczkiewicz, MD, PhD
Department of Cardiology, Brzozów Specialist Hospital, Subcarpathian Oncological Center, Bielawskiego Street 18; 36-200 Brzozów, Poland
phone: + 48 13 4307940, fax number: + 48 13 4307940
e-mail address: krachwal@interia.pl

Conflicts of interest: none
We describe a case of a 51-year-old woman with no previous cardiac history hospitalized due to severe dyspnoea, weight loss, and lower extremity edema. Physical examination also revealed bilateral pleural effusion and ascites. The initial laboratory tests showed hemoglobin 13.5 g/dL [12.0-16.0], platelet count 219 x 10^3 [150-400] and C-reactive proteine 2.65 mg/L [0-5]. Transthoracic echocardiography (Philips HD15 PureWave, Amsterdam, Netherlands) indicated the presence of hypokinetic left (ejection fraction 30%) and right ventricle (RV), enlargement of the right atrium and a tumor in the RV obliterating most of its cavity (Fig 1A, Supplementary material, video S1). The image was confirmed in transesophageal echocardiography (Supplementary material, video S2). The patient underwent further detailed evaluation, and the peripheral hypereosinophilia (eosinophil count 3.8 x 10^3/uL, 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 diagnostics included Eosinophilic Granulomatosis with Polyangiitis and idiopathic Hypereosinophilic Syndrome (HES)- main differences are shown in supplementary material, table S1. Cardiac magnetic resonance (CMR; Philips Achieva 1.5T) classified the RV tumor as a large thrombus (53x17x12 mm)- Fig 1B and supplementary material, video S3. CMR also described restriction, mainly of the RV with endocardial oedema and fibrosis. HES was diagnosed with possible Löffler endocarditis (the patient did not agree to undergo RV endocardial biopsy). Anticoagulation was started with heparin, which was replaced with warfarin. Standard treatment for heart failure with diuretic, angiotensin-converting-enzyme inhibitor and beta-blocker, was also introduced. HES was treated with steroids. Then hydroxycarbamide was added, which ultimately led to eosinophil count normalization. After 3 months of treatment echocardiography showed RV thrombus resolution and moderate improvement in left (ejection fraction 36%) and RV function with persisting fibrosis (Fig 1C,
Supplementary material, video S4) consistent with CMR imaging (Fig 1D). Currently, the patient is alive in one year follow-up.

HES is a diagnosis of exclusion [1-3]. This rare disorder characterized by unexplained peripheral blood eosinophilia (>1.5 x 10⁹/L) and multi-organ system dysfunction occurs most frequently in young to middle-aged males. Interestingly, according to our best knowledge, this is the first female HES with severe RV involvement 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 three 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-usually which involves the left or both cardiac chambers or, less often, as in our case, predominantly RV. CMR 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 forces us to exclude hypereosinophilia in the case of isolated ventricular mural thrombus, and vice versa, after the detection of an unexplained increased level of eosinophils, one should consider echocardiography screening, and looking for intracardiac thrombi as their presence requires urgent medical treatment.
References:


Figure 1AB. Transthoracic echocardiography (1A) and cardiac magnetic resonance (1B): thrombus obliterating the right ventricle (arrow)

LA- left atrium; LV- left ventricle, RA- right atrium, RV- right ventricle
Figure 1CD. Transthoracic echocardiography (1C) and cardiac magnetic resonance (1D) showing dissolved right ventricular thrombus.
Supplementary materials

Table S1. Main differences between HES and EGPA

Videos: Large mural right ventricular thrombus is visible:

Video S1: Transthoracic echocardiography

Video S2: Transesophageal echocardiography

Video S3: Cardiac magnetic resonance

Video S4: Transthoracic echocardiography after 3 months- the thrombus is dissolved