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Massive right ventricle thrombus formation in fatal course of hypereosinophilic syndrome — complex diagnostic approach and interventional management

Short title: Right ventricle thrombus in hypereosinophilic syndrome

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Hypereosinophilia is quite frequent finding in clinical practice. Common causes of hypereosinophilia include infectious, allergic, neoplastic and hematological disorders. Other less frequent cause is hypereosinophilic syndrome (HES), characterized by eosinophilic infiltration and damage of various target organs [1]. Therefore, diagnostic approach in hypereosinophilia should specifically focus on a constellation of signs and symptoms presented by patient. The heart involvement in HES is present in 58% of cases and it is potentially-life threating [2]. Present report was aimed to describe a case of heart failure due to large thrombus

formation in the right ventricle (RV) and to discuss the diagnostic approach and interventional management in the fatal course of HES.

64-year-old woman with a history of arterial hypertension, allergy do multiple drugs, recurrent angioedema episodes and eosinophilia observed in the last 12 months was admitted to the hospital due to acute dyspnea with concomitant Quincke oedema. In the last 3–4 months she experienced progressive exertional dyspnoea. Rheumatological, pulmonological and parasitic background of hypereosinophilia were excluded.

On admission the patient presented dyspnoea (predominantly platypnea) with desaturation to 85%. Laboratory tests demonstrated marked eosinophilia 2.07 × 10^9/l (14%), elevated **N-terminal pro B-type natriuretic peptide** (7160.1 pg/ml) and D-dimer concentration, mild respiratory alkalosis with hypoxemia and hypocapnia. Computed tomography pulmonary angiogram excluded pulmonary embolism and revealed a lack of post-contrast enhancement in the RV (Figure 1D), corresponding with the presence of large mass in transthoracic echocardiography (Figure 1A–C). Bone marrow aspiration and trepanobiopsy ruled out hematologic cause of hypereosinophilia. Cardiac magnetic resonance proved its thrombotic character and revealed global subendocardial fibrosis of the RV (Figure 1E, F). Clinical picture was typical for HES.

After a steroid therapy, decrease of eosinophilic count was obtained. 38-day long anticoagulant treatment did not decrease the size of thrombus and heart failure symptoms were still present. The possible treatment pathways, including thrombus aspiration methods [3], were discussed in the multidisciplinary team. Considering the size of thrombus, ineffectiveness of anticoagulants and the high-risk of RV wall damage, cardiac surgery method to evacuate thrombus was recommended.

The operation went initially successful. However, in the postoperative period mechanical ventilation could not be discontinued. The mechanism of above is unclear, but rapid increase in right ventricle outflow after the surgery resulting in pulmonary oedema and damage of pulmonary capillaries should be taken in consideration. Patient developed pneumonia and sepsis which resulted in death.

The presence of intracardiac thrombi was described in other rare systemic diseases [4]. However, there is a limited data considering the presence and management with intra-cardiac thrombotic complications in the course of HES. Zhang et al. [5]reviewed 477 articles about hypereosinophilia and found 33 cases describing this phenomenon. Intra-cardiac thrombi occurred rarely (about 7%), definitely more often in left or both ventricle (91%) and could lead to peripheral embolism. The pharmacological treatment included: steroid and anticoagulant

therapy, hydroxyurea and imatinib. Three patients underwent surgery procedures: 1- died due to septic shock, 2- were alive, but prognosis was poor. Overall mortality rate of the group with intracardiac thrombus was high (27.3%).

This case raises important issues: (1) the diagnosis of HES requires multimodality imaging and management engaging multidisciplinary team; (2) in cases of hypereosinophilia and symptoms suggesting cardiac involvement urgent diagnostic and therapeutic strategy should be applied as (3) regardless of the implemented procedure the presence of a giant intra-cardiac thrombus in the course of HES significantly worsens the prognosis.

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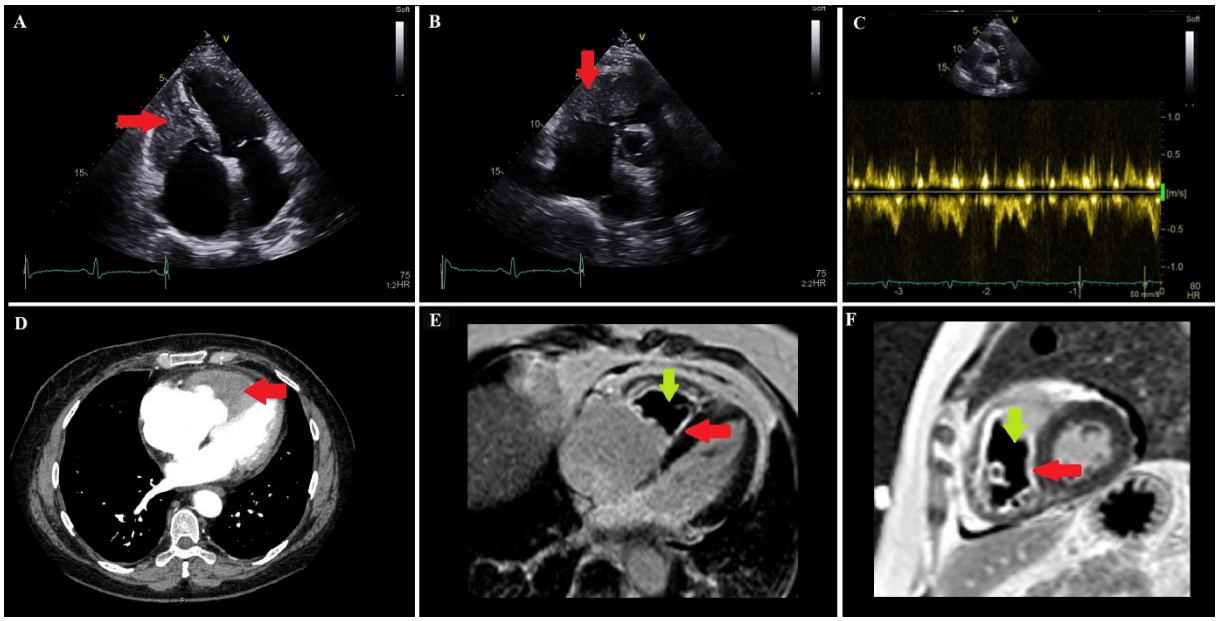

Figure 1. A–C. Transthoracic echocardiography: **A.** apical 4-chamber view **B.** parasternal short axis view: the presence of a large mass filling almost completely the right ventricle cavity and the right ventricle outlow track (the red arrows); **C.** parasternal short axis, pulsed wave Doppler technique: abnormal spectrum of the pulmonary flow with atypical notching. **D.** Computed tomography angiogram: a lack of post-contrast enhancement in the right ventricle. **E, F.** Cardiac magnetic resonance: **E.** four-chamber view; **F.** short axis mid ventricular view. Late gadolinium enhancement. A giant heterogenic mass filling the right ventricle cavity (the green arrows). The global subendocardial fibrosis of the right ventricle (the red arrows)