

Churg-Strauss syndrome with endocardial injury, clot formation in heart's chambers, and neurological complications

Zespół Churga-Strauss przebiegający z uszkodzeniem wsierdza, wytworzeniem skrzepin w jamach serca i powikłaniami neurologicznymi

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A 63-year-old female patient with a history of hypertension and bronchial asthma was admitted to the hospital due to a five-day history of muscle weakness in the upper and lower limbs, balance disorders, and cyanosis of the distant phalanges of the hands and feet with subungual petechiae (Fig. 1). No pathological changes of the heart and lungs could be heard on auscultation. A cranial contrast computed tomography did not reveal any pathologies of the central nervous system (CNS). Laboratory abnormalities included leucocytosis (13.8 G/L) and elevated concentrations of D-dimers (1371 ng/mL), troponin T (0.64 ng/mL), and C-reactive protein (8.52 mg/L). A transthoracic echocardiography (TTE) did not reveal any contractility abnormalities. A non-physiological structure (0.8 × 1.9 cm) could be observed in the left atrium, attached to the posterior mitral leaflet and a paramural structure (2.0 × 1.5 cm) was detected at the apex of the left ventricle (LV). Both pathological findings suggested the presence of blood clots (Fig. 2). Subcutaneous enoxaparin 80 mg b.i.d., acetylsalicylic acid 75 mg q.d., and ramipril were administered. On the third day the patient reported evident right limbs weakness, particularly in the upper limb, and a seriously limited right visual field. The consulting neurologist diagnosed the right hemiparesis, subtle left-sided pyramidal symptoms, and impaired cognitive function. Magnetic resonance imaging revealed recent ischaemic foci in the white matter of the frontal, parietal, and occipital lobes, as well as in the white matter of the semioval centre. No flow was visualised in the A1 section of the right anterior cerebral artery. TTE performed on day 4 did not reveal the previously observed pathological structure on the mitral leaflet. Presumably it was a source of the embolism, leading to ischaemic changes in the CNS. The LV clot still could be observed. Laboratory tests confirmed persistent leucocytosis (13.56 G/L) and severe eosinophilia (8.14 G/L, 60%). An initial diagnosis of Churg-Strauss syndrome (Eosinophilic granulomatosis with polyangiitis) was made. Prednisone 80 mg q.d. was added to the therapy. A marked decrease in leukocyte and eosinophil count was observed in the following days (leukocytes to 4.46 G/L; eosinophils to 0.08 G/L, 0.2%). The neurological status of the patient improved considerably. Warfarin was added to the treatment and enoxaparin was discontinued. The dose of prednisone was reduced gradually. No marked reduction in size of the LV clot was documented on several control TTE examinations. The patient was discharged in good general condition. She was prescribed prednisone, warfarin, and ramipril. The LV clot still could be observed on TTE two months after the discharge, but it was no longer visible at five months. The dose of prednisone was reduced gradually, down to 5 mg per day one year after discharge and was discontinued four months thereafter. Only discrete neurological symptoms persisted by that time. Consecutive peripheral blood smears and eosinophil count were normal.



Figure 1. Cyanosis of fingers with subungual petechiae



Figure 2. Transthoracic echocardiography: blood clots in the left atrium and ventricle

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Conflict of interest: none declared

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