Intracardiac emboli as first presentation of cardiac AL amyloidosis

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A 74-year-old patient presented at the Emergency Room with exercise-induced shortness of breath. Clinical examination revealed arterial hypertension (167/110 mmHg), decreased breathing sounds, and bilateral oedema. Electrocardiogram showed low voltage (Panel A) and aspecific repolarization changes. Chest X-ray confirmed bilateral pleural effusion. D-dimers (7644 ng/mL) and NTproBNP (9650 pg/mL) were elevated. Transthoracic echocardiography demonstrated a concentric hypertrophic left ventricle with myocardial sparkling, poor systolic function (ejection fraction 28%), dilated left atrium, and restrictive filling pattern. A large thrombus was seen in the left atrium (Panel B) and left ventricle (Panel C). Transoesophageal echocardiography demonstrated a left atrial thrombus passing through a patent foramen ovale (Panel D) and thrombotic material in the right atrium (Panel E) originating from the inferior caval vein. Deep venous thrombosis was confirmed in the superficial femoral vein. Angio-CT did not show pulmonary embolism. The patient was immobilized and treated with low-molecular-weight heparin. Four days later, left-sided thrombus had disappeared. Cerebral MRI demonstrated recent parieto-occipital infarction without neurological symptoms. Laboratory diagnosis of cardiac AL amyloidosis was suggested by elevated serum-free light chains: λ, 181 mg/L (ref: 5.7–26.3); κ, 20.3 mg/L (ref: 3.3–19.4); ratio: 0.11 (ref: 0.26–1.65), suspicion of clonal lambda fraction on serum immunofixation, monoclonal free lambda chains on urine immunofixation and urine lambda secretion: 16.4 mg/dL (ref: 0–1). Bone marrow showed a small clone of abnormal plasmacells (3%) (Panel F). Flow cytometry could confirm the abnormal clone (lambda) plasma cells. Characteristics of multiple myeloma such as hypercalcaemia, bone pain, or lytic bone lesions were not present. Subsequently, coronary angiography was performed and myocardial right ventricular biopsy performed. There was non-obstructive coronary artery disease. Histological diagnosis of primary (AL) amyloidosis was confirmed on the congo red stain. Treatment was initiated with oral antiocoagulants, thalidomide, and dexamethasone; serum-free lambda levels diminished subsequently.