



Diagnosis of Isolated Right Ventricular Dysfunction in Systemic Sclerosis Complicated by Pulmonary Embolism: A Multimodality Imaging Approach

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Clinical Image

We report the case of a 72-year-old woman diagnosed with pulmonary (fibrosing interstitial disease) and esophageal (hypotonic, hypokinesia, GERD) Systemic Sclerosis. In 2022 an echocardiogram was performed due to worsening dyspnea, which showed a left ventricle of normal volumes and systolic function, with a D-shape IV septum (A); the right ventricle was severely dilated with moderate systolic dysfunction (FAC=25%; TDI S=8 cm/s). A subsequent cardiac MRI (B) confirmed the severe dilatation of the RV (EDVI 166 ml/m²) with diaphragmatic wall hypokinesia. The bSSFP and CEMRA images documented dilatation of the main trunk of the pulmonary artery (44 mm × 45 mm) and right and left branches. T1 mapping was increased with myocardial ECV within the limits, in line with connective tissue disease with myocardial involvement. In T1-GRE-IR sequences acquired after gadolinium injection we detected junctional and mid-wall distal inferior wall LGE. In 2023 the patient was hospitalized due to progressive-worsening dyspnea: Hemodynamic investigation revealed mild pulmonary hypertension and Sildenafil therapy was started. A control cardiac MRI (C) was performed, revealing a filling defect of the main branch of the left pulmonary artery and of the ipsilateral lower lobar branch which appears hypointense in the LGE images (D), as from endoluminal thrombosis. A lung scintigraphy then showed hypoperfusion of the left lung (E) and an Angio-TC (F) confirmed a partial filling defect of the distal portion of the left main pulmonary artery branch. Follow-up scintigraphy (G) after one week of LMWH therapy showed perfusion improvement of the left lung.

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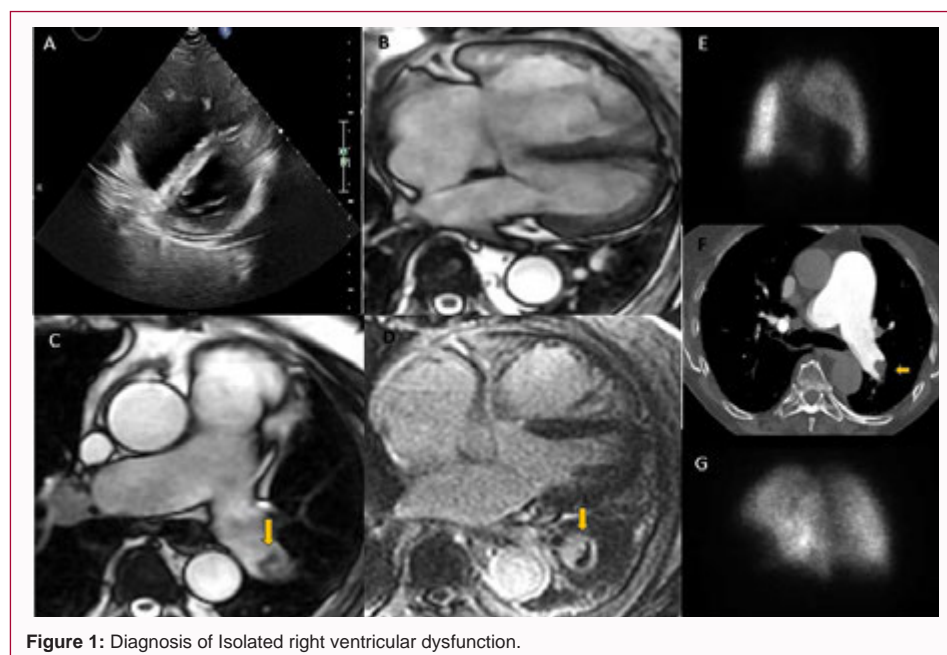


Figure 1: Diagnosis of Isolated right ventricular dysfunction.