Amyloidosis-induced tricuspid stenosis mimicking rheumatic heart disease

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A 62-year-old woman who had suffered from dyspnoea on exertion was referred to our hospital. She had no past or family history of medical illnesses. Chest X-ray showed mild cardiomegaly. Serum creatinine and brain natriuretic peptide level were mildly elevated as 1.66 mg/dL and 1,418 ng/L, respectively. Although left ventricular (LV) ejection fraction was preserved, thickened LV myocardium with a restrictive LV filling pattern, suggesting restrictive cardiomyopathy, was noted on echocardiography (Panel A and see Supplementary data online, Video S1). Notably, tricuspid valve was thickened and stenotic, accompanied by mitral stenosis, which could be usually seen in rheumatic heart disease (Panels B–D and see Supplementary data online, Video S2). T2*-weighted magnetic resonance imaging showed no LV myocardial oedema. Subendocardial and diffuse transmural late gadolinium enhancement and a marked expansion of extracellular space on pre- and post-T1 maps, suggesting cardiac amyloidosis, were shown (Panels E and F and see Supplementary data online, Video S3). A cardiac biopsy showed Congo red-positive with apple-green birefringence under polarized light (Panel G). Finally, under the diagnosis of cardiac amyloidosis with bi-valvular involvement, she was treated with melphalan/dexamethasone chemotherapy.

Tricuspid stenosis is uncommon in adults, and in most cases, it is the result of rheumatic heart disease with rheumatic mitral involvement. Amyloidosis involving a significantly stenotic tricuspid valve is also extremely rare. Without careful cardiac imaging and systemic evaluation, this case could have been misdiagnosed as rheumatic valvular heart disease involving the mitral and tricuspid valves.

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IMAGE FOCUS

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