

Image in cardiology

## Fibrotic Periaortitis Infiltrating Into the Aortic and Mitral Valves

### Periaortitis fibrosa con infiltración en las válvulas aórtica y mitral

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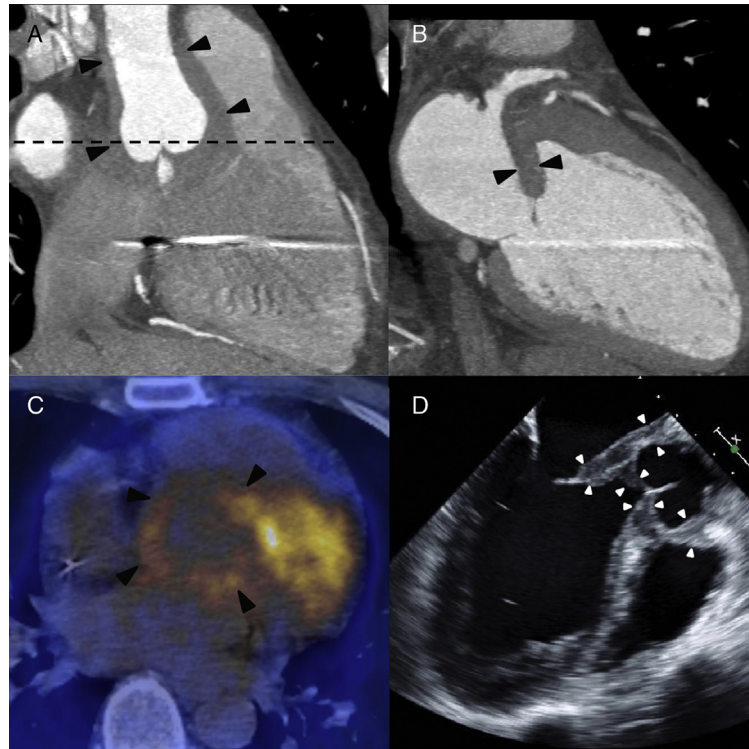


Figure 1.

A 54-year-old man had been admitted to another hospital with syncope and underwent implantation of an intracardiac defibrillator for idiopathic ventricular tachycardia due to induction of monomorphic ventricular tachycardia on electrophysiologic test. Under consideration for nonspecific aortitis by transthoracic echocardiography and positron emission tomography (PET), high-dose prednisolone (0.5 mg/kg/day) had been initiated and then tapered to 5 mg/day for 10 months. He was referred to our clinic because of persistent dyspnea on exertion. Laboratory examination revealed an elevated erythrocyte sedimentation rate of 31 mm/h, an elevated C-reactive protein level of 2.98 mg/dl, and an IgG4 level of 47 mg/dl. Coronal computed tomography images demonstrated an aortic wall with surrounding soft tissue mass (Fig. 1A, arrowheads) and an extremely thickened anterior mitral leaflet (Fig. 1B, arrowheads). The PET showed increased <sup>18</sup>F-fluorodeoxyglucose uptake (Fig. 1C, arrowheads) in the aortic root at the level of the dotted line (Fig. 1A), proximal ascending aorta, and aortic arch. Transesophageal echocardiography revealed a continuous, severe thickening of the aortic root, aortic valve, and anterior mitral leaflet (Fig. 1D, arrowheads). Biopsy of the aorta was performed to rule out infective or neoplastic aortitis and histopathologic examination revealed focal lymphoplasmacytic infiltration and fibrosis. The presumptive diagnosis was IgG4-related periaortitis, and high-dose prednisolone (1 mg/kg/day) was restarted to control active inflammation. In parallel with clinical improvement in symptoms such as dyspnea, elevations in ESR and CRP levels gradually reverted to normal. Three months later, follow-up PET demonstrated marked decrease of previous FDG uptake.

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