A case of IgG4 related disease presented as constrictive pericarditis

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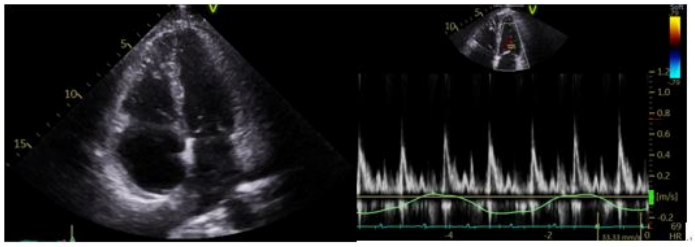
IgG4-related disease(IgG4-RD) is an immune-mediated fibroinflammatory disease affecting multiple organs such as hepatobiliary system, orbits, lungs, kidneys, salivary glands, aorta, and retroperitoneum. Herein, we describe a rare case of IgG4-RD presented as constrictive pericarditis. A 66-year-old man who had been treated as acute pericarditis and persistent AF for generalized edema with abdominal distension, DOE (NYHA class III) for 6 months has been referred to our hospital. NTproBNP was 1245pg/mg, and TTE showed severely reduced LV systolic function (EF 28%) with thickened pericardium. ANA was positive (1:320), and serum PEP showed elevated total protein, gamma levels. Diuretics and steroid treatment was initiated for heart failure, congestive liver cirrhosis and pericarditis. After treatment with steroid and heart failure medication, despite LVEF has been improved to 63%, dyspnea and hepatomegaly have worsened,and TEE showed septal bouncing with suspicious constrictive physiology. After admission for further evaluation, cardiac catheterization was performed and the findings of Kussmaul's sign, square root sign, and 4 chamber pressure equalization were suitable for constrictive pericarditis. Extended pericardiectomy was performed for chronic constrictive pericarditis, and surgical pathology showed increased infiltration of IgG4 positive plasma cells (IgG4:IgG ratio = 60%, 150 IgG4 cells/HPF). Additional tests were performed to confirm the diagnosis. Serum IgG4 was elevated to 269mg/dl, and PET-CT showed increased uptake in the pericardium, ascending aorta and abdominal aorta. According to 2011 comprehensive diagnostic criteria, he was diagnosed with IgG4-RD with constrictive pericarditis and aortitis. The patient has been treated with oral corticosteroid and immunosuppressants at rheumatology department. During 1year follow-up after surgery, the patient remained in remission. Although awareness of IgG4-RD is increasing, there are few cases of IgG4-RD mainly manifesting as pericarditis. The case shows that in patients with constrictive pericarditis, IgG4-RD needs to be included in differential diagnosis,and further evaluation such as serum IgG4 and biopsy.



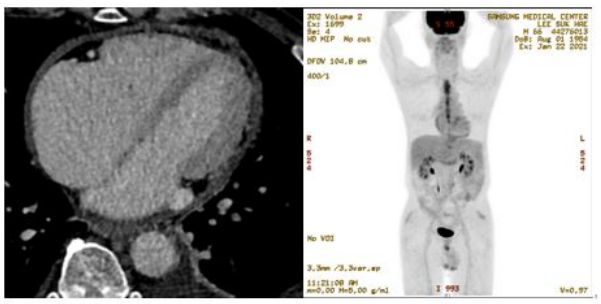


Figure1) 2020.01.31 chest CT: Diffuse pericardial wall thickening.

Figure2) 2021.01.22 PET-CT: Mild increased uptake in the pericardium, ascending thoracic aorta and proximal aortic arch.

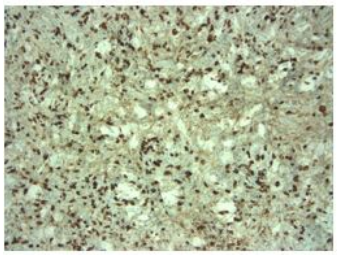


Figure 3) 20.12.28 Heart, pericardium, pericardiectomy pathology.