

CARDIAC INVOLVEMENT WITH IgG4-RELATED DISEASE, REPORT OF TWO CASES

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Cardiac involvement in IgG4-related disease (IgG4-RD) is relatively rare; multiple cases of possible heart involvement with IgG4-RD have been reported, but uncertainty persists and partially resides in the lack of clear criteria defining IgG4-RD cardiac disease. We report two cases of IgG4-RD with heart involvement.

Our first patient was a 59-year-old Caucasian female with lymphocytic thyroiditis, enlargement of the oculomotor muscles on CT imaging, and orbital biopsy consistent with inflammatory sclerosing pseudotumor. She was treated with corticosteroids with complete resolution of the symptoms. Two years later in 2006 she had recurrence of the symptoms; the initial biopsy was reviewed and was characterized as IgG4-RD. In 2008 experienced substernal discomfort, had a negative stress tests and no coronary artery calcium deposits on CT scan. The patient developed first degree heart block and 4 days later had acute chest pain, with a repeat negative stress test. Echocardiogram showed moderate cardiomegaly. She later developed third degree heart block and underwent pacemaker placement. PET CT scan revealed increased FDG uptake in the oculomotor muscles, the sinoatrial node, and the left ventricle. She was treated successfully with corticosteroids.

Our second case was a 28-year-old African American female who presented with left arm pain and decreased palpable pulses. Her CT angiography revealed a partial occlusion of the left subclavian artery, fusiform dilation of the aortic arch, and a left carotid artery aneurysm. The aneurysms were surgically corrected. Six months later she developed swelling on the left side of her neck and diagnosed with a recurrent carotid aneurysm. Echocardiogram and cardiac MRI detected thickening of the mitral valve annulus and anterior leaflet, suggestive of pseudotumor. Histopathology of the aortic tissue removed during the aortic aneurysm repair surgery was consistent with IgG4-RD. She also had elevated serum IgG level (2300 mg/dl) and IgG4 levels (231 mg/dl). The patient received treatment with Rituximab and prednisone followed by decrease of the lesions in the mitral valve.

Discussion:

Both patients had multi-organ involvement, characteristic histology for IgG4-RD and response to corticosteroids, plus rituximab. Cardiac biopsy was not performed, but the constellation of findings was suggestive of IgG4-RD. IgG4-RD cardiac involvement is rare, but potentially fatal. Further research and development of organ specific criteria for diagnosis, as well as defining the role of PET imaging along with tissue biopsy, would be helpful in timely recognizing and treating this condition.