Unusual cause of pericardial effusion: IgG4-related disease

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DESCRIPTION

A 53-year-old woman presented to a hospital with a history of general malaise, significant weight loss and chest discomfort for 6 months. She had no significant past medical history. Her initial investigations revealed elevated inflammatory markers, normocytic anaemia, mildly increased cardiac biomarkers and an elevated eosinophil count. Baseline chest radiograph showed bilateral pleural effusions, and transthoracic echocardiography revealed a small pericardial effusion, mild impairment of left ventricular function and moderate mitral regurgitation. Differential diagnosis included viral myocarditis and an autoimmune disease. Patient subsequently underwent cardiac MRI (CMRI) for further evaluation. This revealed significant thickening and active inflammation of the pericardium (figure 1). Late gadolinium enhancement (LGE) series demonstrated extensive circumferential LGE of the pericardium and linear subendocardial LGE of the left ventricle in a non-coronary distribution (figure 2). These findings in the setting of hypereosinophilia were concerning for an idiopathic eosinophilic syndrome. Patient underwent a pericardial biopsy via mediastinoscopy for further evaluation. Histology demonstrated abundant eosinophils seen at the inner surface of the pericardial cavity. IgG and IgG4 staining were performed, and over 40% of plasma cells were positive for IgG4. After a multi-disciplinary discussion, a diagnosis of IgG4-related disease was made. Patient has been maintained on

Figure 2 Extensive circumferential late gadolinium enhancement (LGE) of the pericardium (white arrowheads) as well as linear subendocardial LGE of the left ventricle in a non-coronary distribution (white asterisk).

CD20 monoclonal antibody with good response. She remains under close follow-up.

IgG4-related disease is a rare, fibroinflammatory entity. It is a difficult condition to diagnose as it can present with a myriad of clinical features suggestive of a neoplastic or an inflammatory process. This disease may affect multiple organs and, as a result, can present with various, non-specific symptoms, which may lead to a delay in diagnosis and treatment.¹ Serum IgG4 levels coupled with typical biopsy findings, including abundant lymphocytes, fibrosis, IgG4-plasma cells and obliterative phlebitis in the setting of a relevant clinical presentation are diagnostic of IgG4-related disease. Treatment consists of steroids and steroid-sparing agents, such

Figure 1 Significant thickening and active inflammation of the pericardium as evidenced by the presence of oedema on T2-weighted Short-T1 Inversion Recovery (STIR) images (white arrows).

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Learning points

- ▶ Cardiac MRI should be considered in suspected autoimmune or inflammatory condition involving the myocardium or pericardium.
- ▶ It is important to consider an idiopathic eosinophilic syndrome in the presence of an elevated eosinophil count.
- ▶ Specialist input is required once diagnosis of IgG4-related disease is made, and inappropriate treatment may result in relapse and increased morbidity.

as CD-20 monoclonal antibody (rituximab). The natural history of this disease remains uncertain.²

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