

A case of IgG4-related pericarditis

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ESC curriculum 6.6 Pericardial disease • 2.2 Echocardiography

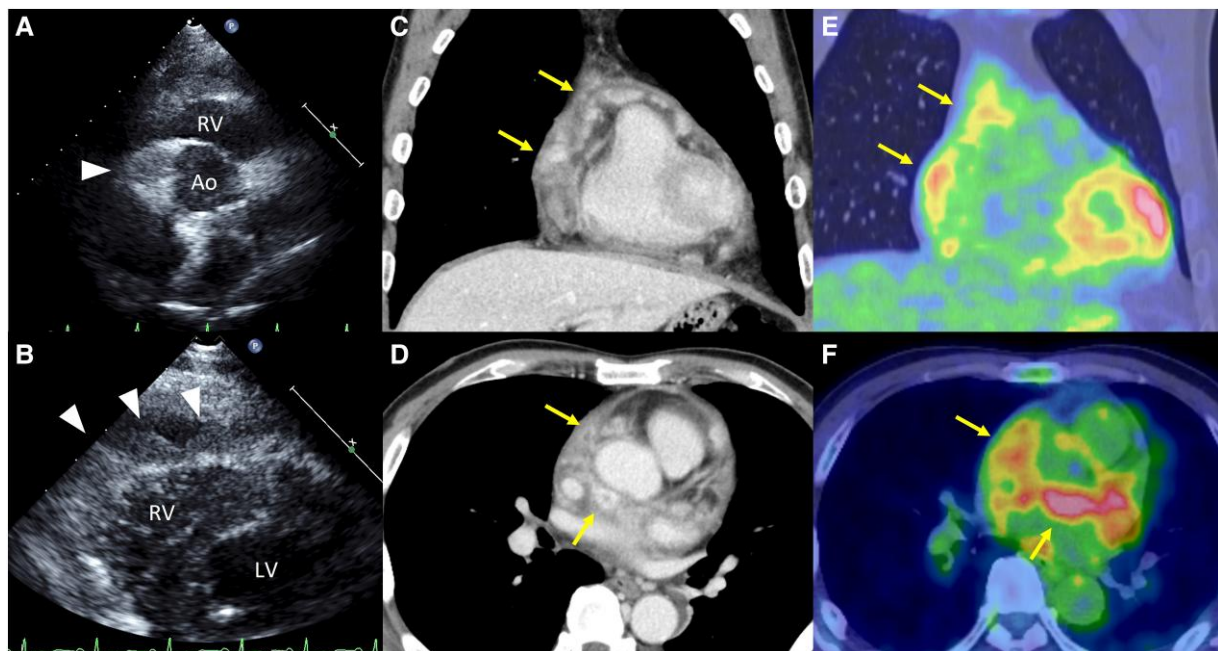


Figure 1 (A and B) Transthoracic echocardiography shows hyperechoic lesion around the aortic root (A, arrowhead) and multiple nodules on the visceral pericardium (B, arrowheads). (C and D) Contrast-enhanced computed tomography shows diffuse thickening of pericardium, with a small amount of pericardial fluid present. Contrast-enhanced nodules are visible along the visceral pericardium and in the soft tissue around the aortic root, coronary arteries, and on the dorsal aspect of the left atrium (arrows). (E and F) ^{18}F -fluorodeoxyglucose positron emission tomography/computed tomography. ^{18}F -fluorodeoxyglucose uptake coincident with mass and nodules, indicating active inflammation (arrows). Ao, aorta; LV, left ventricle; RV, right ventricle.

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Case description

A 66-year-old man was transferred to our hospital with a diagnosis of pericarditis that was refractory to treatment using non-steroidal anti-inflammatory drugs and colchicine. At admission, body temperature was 36.5°C, and blood pressure was 142/81 mmHg. A 12-lead electrocardiogram revealed a heart rate of 90 beats/min in sinus rhythm and negative T waves in I, II, aVL, and V 5–6. Transthoracic echocardiography revealed a small amount of pericardial effusion, a hyperechoic lesion around the aortic root (Figure 1A), and multiple nodules in the pericardium (Figure 1B, arrow heads). Contrast-enhanced computed tomography (CT) revealed diffuse pericardial thickening without calcification. Multiple contrast-enhancing nodules were present along the pericardium and soft tissue around the aortic root, coronary artery, and dorsal left atrium (Figure 1C and D, arrows). ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography/CT revealed focal FDG uptake consistent with the lesions detected by contrast-enhanced CT (Figure 1E and F, arrows). A pericardial biopsy was performed to differentiate lymphoproliferative disorders and revealed fibrosis and infiltration of inflammatory cells, including lymphocytes and plasma cells, with no malignant findings. The laboratory test indicated increased IgG4 (415, 11–121 mg/dL). Oral prednisolone at a dose of 40 mg/day (0.6 mg/kg/day) was initiated based on the IgG4-related diseases classification criteria. No other organs were involved. After 2 weeks of treatment, the inflammatory response became negative, serum IgG4 level decreased (112 mg/dL), and chest pain and

pleural effusion promptly subsided. Transthoracic echocardiography revealed that the pericardial effusion had disappeared and the nodules along the pericardium had shrunk.

IgG4-related disease is a multi-organ involved systemic disease and pericardial involvement mostly presents as pericarditis and pericardial thickening.^{1–3} Early diagnosis and intervention are recommended; irreversible fibrosis may lead to constrictive pericarditis in the advanced stages of the disease. IgG4-related disease should be considered in the differential diagnosis of unexplained pericarditis.

Consent: The author/s confirm that written consent for submission and publication of this case report including the image(s) and associated text was obtained from the patient in line with COPE guidance.

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Data availability

The data underlying this article are available in the article.

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