



Multiple nodules under the pericardium in a patient with IgG4-related disease

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Received: 18 May 2020 / Revised: 17 August 2020 / Accepted: 1 September 2020 / Published online: 7 September 2020
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Presentation

A 73-year-old male was found with lung nodule during a regular checkup. He had a history of asthma and hypertension for many years. Besides, he had undergone a nasal polypectomy before. Physical examinations showed that the right submandibular gland was swollen and hard. The laboratory tests indicated increased erythrocyte sedimentation rate (33 mm/h, 0–15 mm/h), IgG (22.49 g/L, 7–17 g/L), IgG4 (15600 mg/L, 80–1400 mg/L), T-IgE (6040 KU/L, 0–60 KU/L), and normal high-sensitive C-reactive protein (1.51 mg/L, 0–8 mg/L) level. Antinuclear antibody was positive (1:160, membrane pattern), while other autoantibodies were negative. A whole-body ¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) positron emission tomography/CT (PET/CT) was performed (Fig. 1a) and revealed enlargement and diffusely elevated uptake of bilateral submandibular glands (especially right side), patchy ¹⁸F-FDG-avid lesions in the prostate gland, a hypermetabolic nodule in the left lower lobe of the lung, and

multiple nodular lesions in the pericardium and mediastinum, as well as thickness and elevated uptake of bilateral iliac arteries and circumferential soft tissue (Fig. 1c, d, e, f, g, and h). Histopathological findings of the right submandibular gland biopsy revealed dense lymphoplasmacytic infiltration and fibrosis (Fig. 1i). Immunohistochemical staining demonstrated that IgG(+) (Fig. 1j) and IgG4+ cells were 30/HPF (Fig. 1k), IgG4+/IgG+ < 10%, CD3(+), CD35(+), CD21(+), and CD20(+). The patient was diagnosed with IgG4-related disease (IgG4-RD) according to the 2019 ACR/EULAR classification criteria [1]. He was treated with prednisolone 40 mg/day and cyclophosphamide 50 mg every other day. Two weeks later, prednisolone was tapered gradually until maintained at 7.5 mg/day. After treatment, serum IgG4 level declined to 4240 mg/L and he repeated ¹⁸F-FDG PET/CT, which showed complete remission of all affected lesions (Fig. 1b).

Discussion

IgG4-RD is a multi-organ involved systemic disease and pericardial involvement mostly presents as pericarditis and pericardial thickening [2–4]. To our knowledge, pericardial nodules are unusual and there is no such report before. The same as commonly affected organs in IgG4-RD, pericardial nodules respond well to glucocorticoids.

Funding This work was supported by CAMS Initiative for Innovative Medicine (2017-I2M-3-001) and the National Natural Science Foundation of China (No. 81771757).

Compliance with ethical standards

Disclosures None.

Patient consent Obtained.

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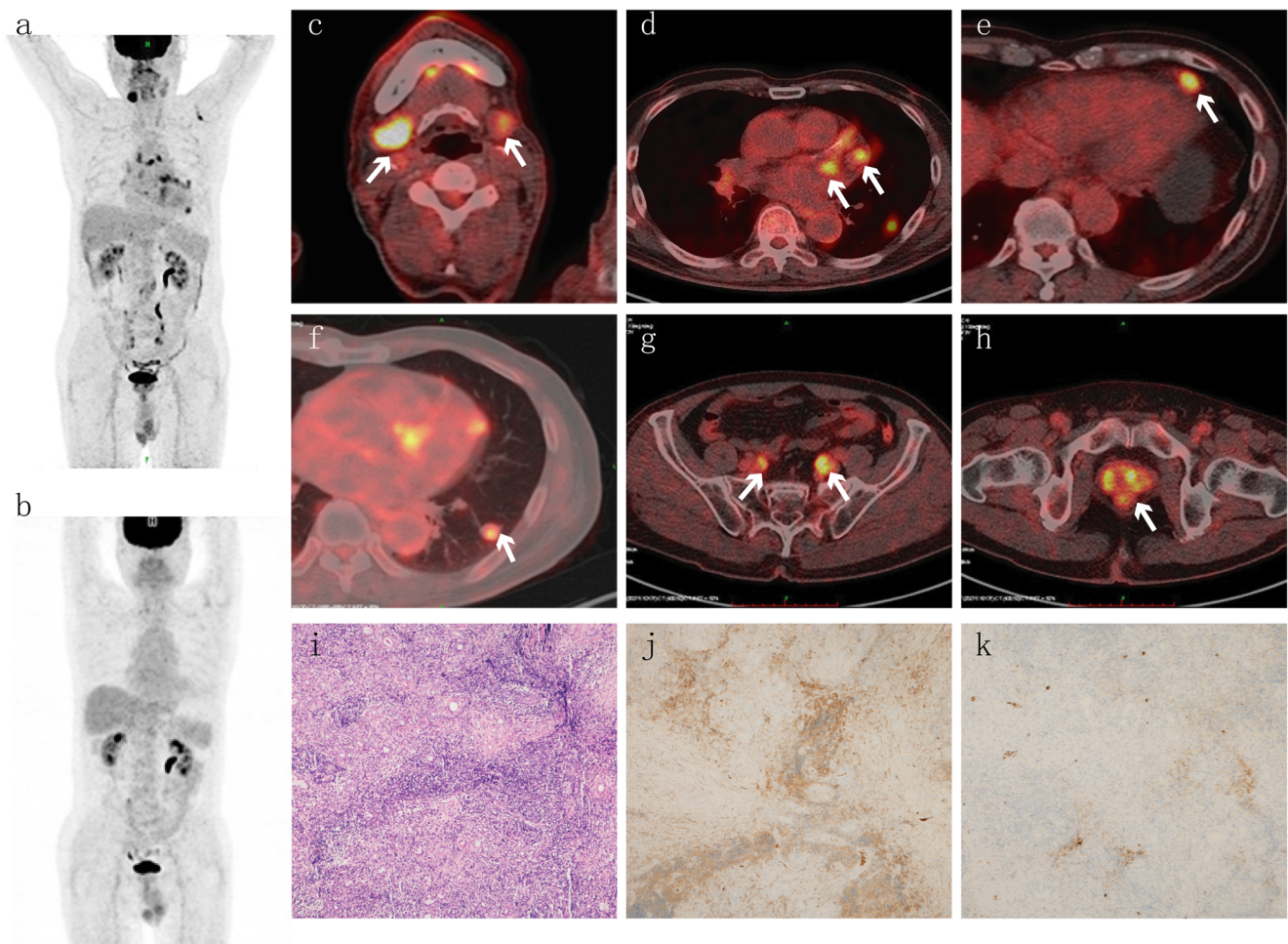


Fig. 1 Imaging and immunohistochemical findings of the patient with IgG4-related disease. ^{18}F -fluorodeoxyglucose (^{18}F -FDG) positron emission tomography/CT (PET/CT) showed the whole-body lesions (a) and complete remission of affected lesions after treatment (b). ^{18}F -FDG PET/CT showed the enlargement and diffusely elevated uptake of bilateral submandibular glands (c, arrows), multiple nodular lesions in the pericardium and mediastinum (d and e, arrows), a hypermetabolic nodule in the

left lower lobe of the lung (f, arrow), thickness and elevated uptake of bilateral iliac arteries and circumferential soft tissue (g, arrows), and patchy ^{18}F -FDG-avid lesions in the prostate gland (h, arrow). Hematoxylin and eosin (HE) staining revealed dense lymphoplasmacytic infiltration and fibrosis ($\times 40$ magnification) (i). Immunohistochemical staining showed the IgG-positive plasma cells ($\times 40$ magnification) (j) and IgG4-positive plasma cells ($\times 40$ magnification) (k)

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