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Case report

An IgG4-related sclerosing mediastinitis in posterior mediastinum: CT findings☆

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Abstract

Immunoglobulin G4 (IgG4)-related sclerosing disease, an uncommon disease entity, is known to involve various organs. To our knowledge, few reports have been presented on IgG4-related sclerosing diseases involving the mediastinum, especially the posterior mediastinum. We present a case of IgG4-related sclerosing disease of the posterior mediastinum with imaging findings on computed tomography (CT).

Keywords: IgG4-related sclerosing disease; Mediastinitis; Computed tomography

1. Introduction

IgG4-related sclerosing disease is rare and is known to involve various organs including the salivary gland, breast, lung, pancreas, bile duct, retroperitoneum, kidney, urethra and so on [1–3]. Its pathogenesis remains undefined, and it is composed of many disorders that have specific histopathologic and serologic features [4]. Few reports have been presented on IgG4-related sclerosing disease involving the mediastinum, and it is mainly delineated in the clinical and pathological findings [5,6]. Therefore, we present a case of IgG4-related sclerosing disease involving the posterior mediastinum confirmed by pathology with imaging findings on CT in this paper.

2. Case report

A 54-year-old man was admitted to our hospital with a two-month history of cough with white sputum, shortness of breath, back pain, anorexy and fever occasionally. The physical examination revealed facial swelling and jugular varicosity. The laboratory tests showed mild leucopenia (1320/μL) with a descended neutrophil percentage (26.2%) and eosinophil leukocytosis (1740/μL) with an elevated eosinophil percentage (34.6%). The serum IgG4 level was not available in this case. However, elevated Immunoglobulin E (IgE) value (680.0 IU/mL) was detected. Carbohydrate antigen (CA125, CA153 and CA199), squamous cell carcinoma antigen (SCCA), carcinoembryonic antigen (CEA) and alpha-fetoprotein (AFP) were all within the normal reference range. Serum antinuclear antibodies (ANAs) spectrum was negative. Plain and contrast-enhanced chest 128-multi-slice CT (Optima CT660, GE Healthcare, Milwaukee, WI, USA) scans demonstrated a mass in the posterior mediastinum (Fig. 1). No significantly clinical and radiological abnormality

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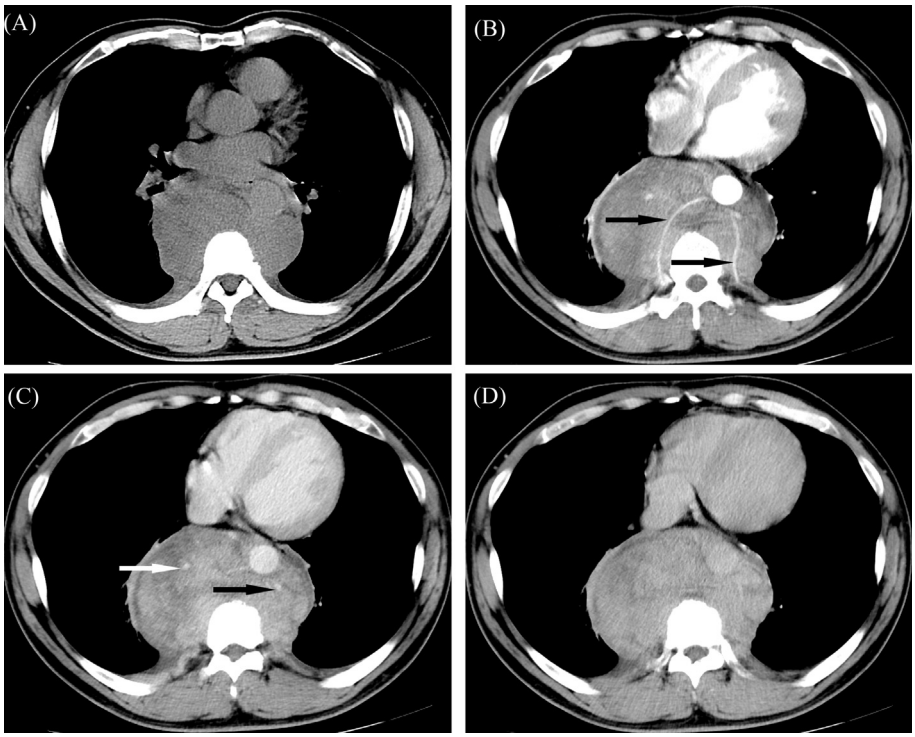

Fig. 1. CT images acquired in a 54-year-old man with IgG4-related sclerosing disease in posterior mediastinum. (A) Non-enhanced chest CT image demonstrates an irregular mass in posterior mediastinum, with inhomogeneous attenuation and well-defined margin. Slight bilateral pleural effusion is visible. (B) Contrast-enhanced CT in the arterial phase shows the mass locates posterior and lateral to the thoracic aorta. The thoracic aorta, intercostals artery (black arrows), azygos vein and hemiazygos vein are encased and squeezed shift by the mass. (C, D) Contrast-enhanced CT in the venous phase shows the azygos vein (black arrow) and hemiazygos vein (white arrow) with narrowing. The mass presents a gradually heterogeneous enhancement.

was found in other organs. The laboratory test results and radiologic findings were somewhat confusing and non-specific because they can suggest malignancy, benign and inflammatory diseases [7]. Hence, the patient underwent percutaneous biopsy of the posterior mediastinum lesion under ultrasonography guidance. The pathologic specimen showed lymphocyte and plasmacyte infiltration as well as fibrosis (Fig. 2A and B). On immunohistochemical staining (Fig. 2C and D), conspicuous IgG4 rich-lymphoplasmacytic infiltrates (IgG4 positive cells > 50/HPF) could be seen. What's more, the IgG4+/IgG+ cell ratio >40%. Therefore, all these pathologic results confirmed the diagnosis of IgG4-related sclerosing disease.

3. Discussion

First described in association with autoimmune pancreatitis (AIP) in 1995 [8] and manifested to be found in extra-pancreatic organ in 2003 [9], IgG4-related sclerosing disease is currently considered to be a systemic fibro-inflammatory disorder characterized by tumefactive lesions, prominent IgG4 rich-lymphoplasmacytic infiltrate, and storiform fibrosis with or without elevated serum IgG4 levels [10]. Because clinical symptoms and histopathologic features vary with lesion location and the recognition of this disease is not enough, its received diagnostic criteria have not yet been established [11]. Umehara et al. [11] recently proposed a comprehensive diagnostic criteria: (a) Organ swelling, mass or nodularlesions, or organ dysfunction; (b) a serum IgG4 concentration > 135 mg/dl; and (c) histopathological findings of >10 IgG4 cells/high power field (HPF) and an IgG4+/IgG+ cell ratio > 40%. According to the criteria, IgG4-related sclerosing disease is classified as definite, probable, possible, and denial. A definite diagnosis should fulfill criteria (a) + (b) + (c); probable diagnosis: (a) + (c); possible diagnosis: (a) + (b). As the absence of serum IgG4 level examination, 'probable' IgG4-related sclerosing disease can be diagnosed to our patient. In recent studies frequency of high IgE is surprisingly (86%–87%) [12,13] in AIP patients, which may be equal to frequency of high IgG4 (73.3%–94.3%) [14,15]. What's more, peripheral eosinophilia is also not rare in IgG4-related sclerosing disease in AIP patients [12]. Both of these two signs can be seen in our case. The pathogeneses of elevated eosinophil percentage and IgE level are still poorly understood in patients with IgG4-related sclerosing disease. As these two signs are common in allergic procedure, Kuruma et al. [12] speculate that allergic mechanisms may be concerned with the occurrence of IgG4-related sclerosing disease.

So far, there have been only two reports of IgG4-related sclerosing disease arising in the mediastinum. In 2007, Inoue et al. [5] reported the first case of IgG4-related sclerosing mediastinitis that showed IgG4-positive plasma cells infiltrated into the fibrous tissue and a high serum IgG4 level, and CT scans demonstrated a homogeneous and diffuse mass within the mediastinum involving bilateral main bronchi with severe narrowing. Moreover, this case was treated successfully by steroid therapy. Noh et al. [6] presented a case of IgG4-