Case report

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

Xiaolong Liu a, Shanshan Yu a, Chunyao Yu a, Yanan He a, Jifeng Zhang a, Ying Jiang b, Ping Li a,*

a Department of Radiology, the Second Affiliated Hospital of Harbin Medical University, Harbin 150086, Heilongjiang, China
b Department of Pathology, the Second Affiliated Hospital of Harbin Medical University, Harbin 150086, Heilongjiang, China

Received 17 November 2014; accepted 14 January 2015
Available online 28 February 2015

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 confirmed by pathology 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 venous distention. 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
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 extrapancreatic 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 nodular lesions, or organ dysfunction; (b) a serum IgG 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-
related sclerosing disease invading the trachea and superior vena cava (SVC) in mediastinum. In this patient, mediastinal mass in the intratracheal and right lower paratracheal area and enhanced mass on the SVC wall could be seen in CT images.

IgG4-Related sclerosing mediastinitis, like other IgG4-related sclerosing disease, usually respond to glucocorticoids typically with symptomatic improvement, reductions in the size of masses or organ enlargement [6]. Therefore, it is crucial to differentiate IgG4-related sclerosing mediastinitis from other mediastinum lesions in order to select the proper treatment. As for our case, there are some differential diagnoses of IgG4-related sclerosing disease in posterior mediastinum on CT scans. The vessel encasement or narrowing and enhancement pattern seen from sclerosing mediastinitis may be different from paraspinal disease such as nerve sheath tumors and abscess. Nerve sheath tumors typically appear as well-marginated, smooth, rounded, or elliptical masses, with or without intraspinal canal extension and enlargement of neural foramina. Paraspinal abscess of bacterial osteomyelitis or spinal tuberculosis usually presents relative hypoattenuation and is complicated with the destruction of vertebral bone. In addition, lymphoma and other causes of lymph node enlargement may result in mass in this region. Involvement of posterior mediastinal nodal groups is much more common with non-Hodgkin lymphoma, this was seen in 10% of cases [16-18]. CT scans may manifest extranodal involvement and extra thoracic adenopathy and may be helpful in differential diagnosis. The mass-like imaging features in our case may be related to the mass-forming characteristics of IgG4-related sclerosing disease, which can manifest inflammatory pseudotumor in many organs [7,19].

CT scans play an important role in evaluating not only the size and content of posterior mediastinum lesions but also the involvement of organs and vasculature when considering surgical strategies [20]. Based on the imaging characteristics, enhancement patterns and demographics, the differential diagnosis of posterior mediastinum lesions can be narrowed down [21]. However, malignancy, benign and inflammatory posterior mediastinum lesions may have overlapping imaging characteristics, this makes single imaging index insufficient to identify the IgG4-related sclerosing disease. And serum IgG4 level as well as histological examination is usually required to confirm the diagnosis. But CT findings can still play an irreplaceable role in the detection, differential diagnosis and pretherapeutic assessment of IgG4-related sclerosing disease of the posterior mediastinum.

References


