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Case Report

IgG4-related sclerosing disease with mesenteric and retroperitoneal involvement

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ABSTRACT

IgG4-related sclerosing disease is characterized by infiltration of various tissues and organs, including lung, kidney, pancreas, salivary glands, biliary tree, breast and vessels with IgG4-rich plasma cells. Fibroinflammatory infiltration of these tissues usually progress into fibrosis, eventually resulting in organ failure. In this case report, ultrasonography, computed tomography and positron emission computed tomography features of IgG4-related sclerosing disease involving the retroperitoneum and mesentery are presented together with histopathologic correlation.

1. Introduction

IgG4-related sclerosing disease refers to proliferation and infiltration of IgG4-positive plasma cells within tissues leading to tumefactive lesions, dense lymphoplasmocytic infiltrates and fibrosis [1]. IgG4-related sclerosing disease affects almost all systems and organs in the body including pancreas, lymph nodes, lungs, kidneys, salivary glands, biliary tree, retroperitoneum, mesentery, testis, prostate, breast, aorta, meninges, skin and pericardium [1]. The underlying histopathological features are similar in the setting of IgG4-related sclerosing disease, despite involvement of various body parts. Histopathological sampling is usually necessary for establishing the diagnosis, since IgG4 levels may not be increased in the serum [2]. Awareness for the associated imaging findings in IgG4-related sclerosing disease is critical, as it may be helpful in guiding the diagnosis and management of these patients. In this case report, we present imaging findings of IgG4-related sclerosing disease in a patient with involvement of retroperitoneum and mesentery.

2. Case report

A 35-year-old male patient presented to our hospital with abdominal swelling, abdominal pain, weight loss and fatigue. He had a history of surgical resection of an abdominal mass 4 years ago, which was diagnosed as lipoma per the histopathologic examination. Physical examination of the patient depicted an abdominal mass with palpation. Laboratory tests revealed increased sedimentation rate. Ultrasonography demonstrated a multilobulated mass with heterogeneous echogenicity in the mesenteric and retroperitoneal region. Abdominal computed tomography (CT) examination was performed with a 16-detector CT equipment (Somatom Sensation 16, Siemens, Erlangen, Germany) with administration of 100 ml (ml) of intravenous (IV) contrast agent at a flow rate of 4 ml/s (300 mg/ml Omnipaque, GE Healthcare, Ireland). No oral contrast agent was given. The venous phase of the abdominal CT was acquired 70 s after the triggering threshold of the abdominal aorta (100 Hounsfield Unit) was reached. The body region between the level of the upper diaphragm and ischial tubercle was included in the scanning area. CT demonstrated a large, ill-defined and low attenuating mass in the mesentery and retroperitoneum encircling mesenteric and retroperitoneal vessels (Fig. 1a and b). Positron emission tomography (PET)-CT revealed peripherally avid [18]F-flourodeoxyglucose (FDG) uptake in the mass (Fig. 1c).

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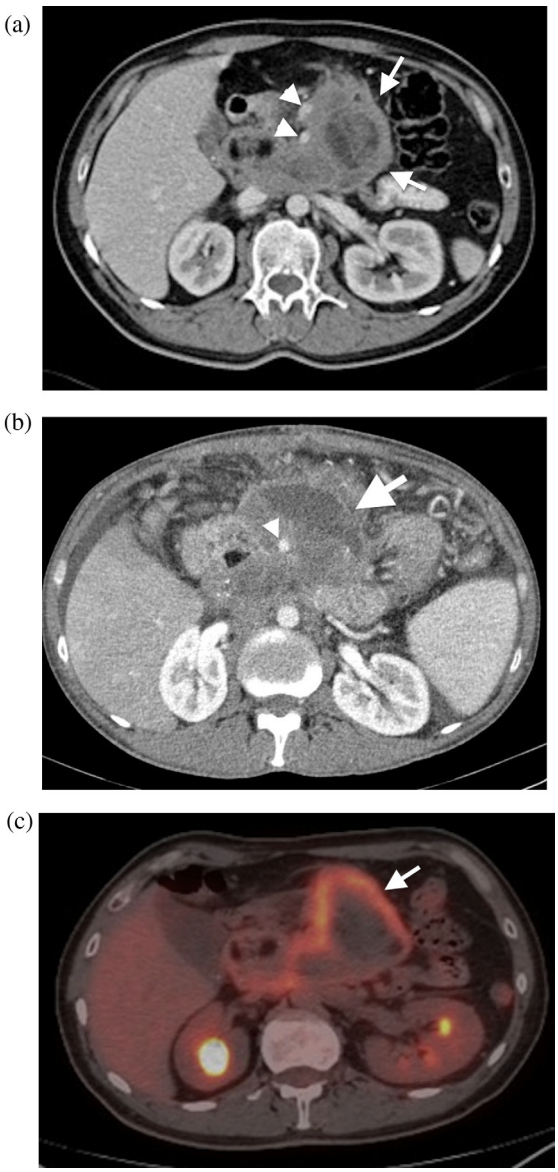

Fig. 1. Mesenteric mass secondary to IgG4-related disease. (a) Axial contrast-enhanced CT demonstrates a fibrofatty mass (arrows) localized in the mesentery, anterior to the abdominal aorta and inferior vena cava. The mass encompasses the mesenteric vessels (arrowheads). (b) Mesenteric mass (arrow) encircles but does not invade superior mesenteric artery (arrowhead). (c) PET-CT reveals avid peripheral enhancement (arrow) in the abdominal mass mimicking malignancy with a SUV max value of 8.1.

The patient underwent abdominal surgery and histopathologic assessment of the resected abdominal mass yielded a diagnosis of IgG4-related sclerosing disease (Fig. 2).

3. Discussion

IgG4-related sclerosing disease is caused by proliferation of IgG4-rich plasma cells and infiltration of eosinophils within various body parts, eventually leading to obliterative phlebitis and fibrosis in the affected regions [3,4]. Pancreas is the most frequently involved organ in the abdomen and the specific entity is also called as type 1 autoimmune pancreatitis. Many disorders previously known as a separate disease such as sclerosing mesenteritis, Mikulicz's syndrome, Küttner's tumor, Riedel's thyroiditis, inflammatory pseudotumor, mediastinal fibrosis, retroperitoneal fibrosis and periaortitis are recently defined as IgG4-related sclerosing disease [1]. The variability of IgG4 concentrations among healthy people and the low specificity of increased IgG4 limits the utility of serum IgG4 measurements in the diagnosis of IgG4-related sclerosing disease. Up to 30% of patients present with serum IgG4 concentrations within the normal range [5]. Therefore, histopathological analysis of biopsy or surgical specimens and immunohistochemical confirmation with IgG4 immunostaining remains the mainstay in the diagnosis. In this regard, it should be kept in mind that IgG4-bearing plasma cells could be found in various inflammatory disorders [1]. A high ratio (>50%) of IgG4-bearing plasma cells to IgG-bearing plasma cells in histopathological specimens is therefore considered highly specific for the diagnosis of IgG4-related sclerosing disease [6,7].

IgG4-related sclerosing disease most frequently presents as a diffuse infiltrative or mass-forming lesion in the body on cross-sectional imaging studies, mimicking other inflammatory and neoplastic disorders. Early diagnosis of IgG4-related sclerosing disease is essential, since organ failure and life-threatening conditions may occur in patients with massive cell infiltration and vascular complications such as thrombosis of small and large-sized vessels. The diagnosis is usually made incidentally due to imaging findings or histopathological assessment of specimens obtained for other indications rather than IgG4-related sclerosing disease. The clinical importance, wide range of involvement sites, incidental detection rates and histopathological challenges necessitate awareness for imaging features in IgG4-related sclerosing disease.

IgG4-related sclerosing disease involving the mesentery and retroperitoneum presents as an infiltrative mass encircling mesenteric vessels [8]. Since IgG4-related sclerosing disease presents as a soft-tissue mass appearance radiologically, malignant disorders should be distinguished from this disorder in order to prevent unnecessary therapeutic interventions. Differential diagnosis of primary mesenteric neoplasms include desmoid tumor, carcinoid tumor, lipoma, schwannoma, smooth muscle tumors and sarcomas [9]. The presence of fat density in the mesenteric mass of our case has raised the suspicion for liposarcoma, however mesenteric vessels were entrapped by the mass without any signs of invasion – which would have been the case in the setting of sarcoma. Absence of radiating strand pattern was helpful for excluding a possible diagnosis of desmoid tumor or carcinoid. Encircling of mesenteric vessels without invasion may suggest lymphoma, however presence of large amounts of fat density and lack of conglomerated appearance in the setting of a large mesenteric mass were against this diagnosis. Distinction between IgG4-related sclerosing disease and other inflammatory or neoplastic disorders is crucial since IgG4-related sclerosing disease demonstrates favorable responds to corticosteroid therapy [10].

In conclusion, IgG4-related sclerosing disease may present with various imaging findings mimicking several disorders throughout the body. Differentiation IgG4-related sclerosing disease from malignancies may be difficult due to solid soft-tissue mass appearance of IgG4-related sclerosing disease. Awareness of imaging findings can be helpful in the diagnosis and management of Ig-G4 related sclerosing disease.

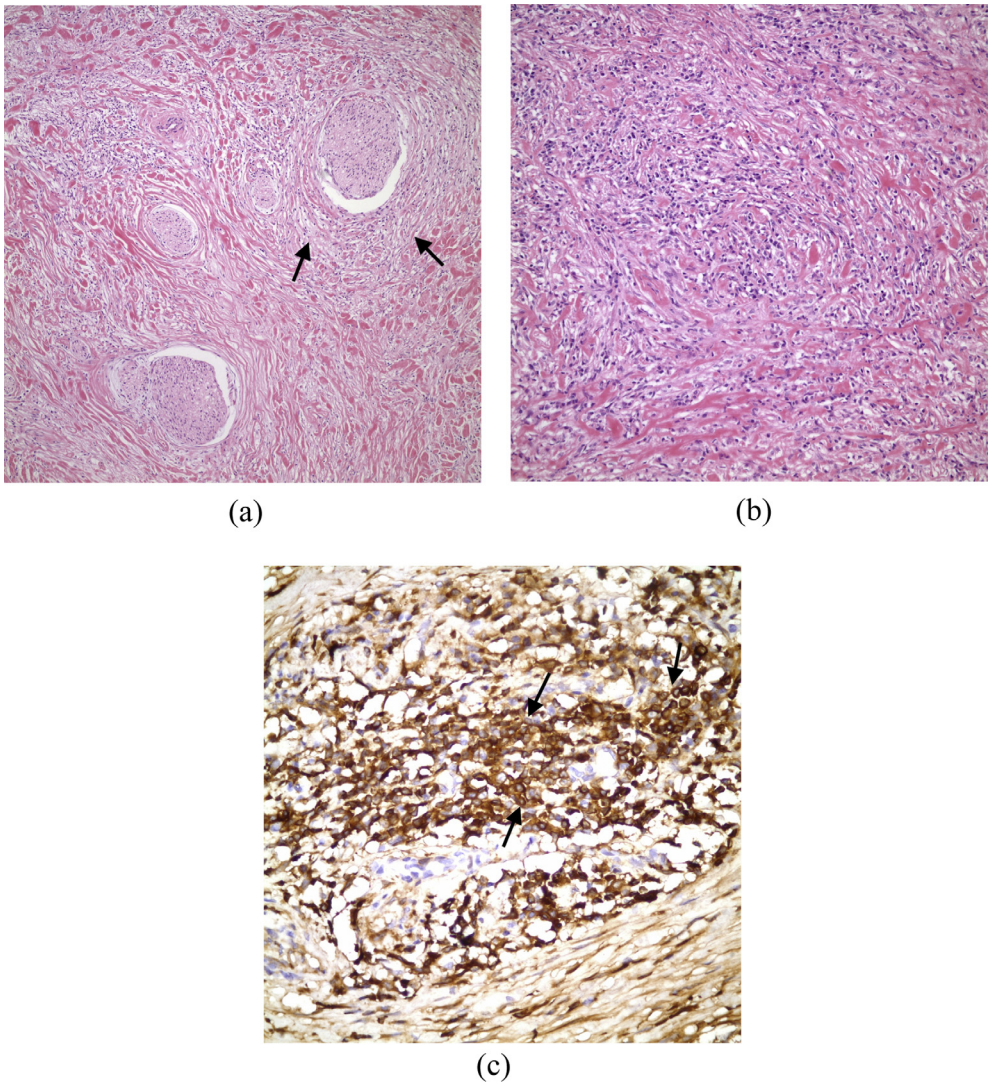

Fig. 2. Histomorphological features diagnostic for IgG4 related sclerosing disease were as follows: (a) Concentric entrapment of vascular structures and peripheral nerves by a fibroinflammatory lesion (black arrows) (H&E, 100×) (b) Myofibroblastic proliferation with storiform pattern and accompanying lymphoplasmacytic inflammatory cells (H&E, 200×). (c) Immunohistochemistry for IgG4 expressed IgG4 positive plasma cells with brown color (black arrows) (E, 400×).

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