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IgG4-related disease mimicking pancreatic cancer: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Most patients with pancreatic masses pose a diagnostic challenge when a benign lesion is suspected, and often, resection is needed before a benign diagnosis is confirmed.
PRESENTATION OF CASE: A 57 years old male patient presented with a pancreatic head mass, obstructive jaundice and submandibular lymph node enlargement. He also had a history of recurrent eye pain and redness, skin lesions, and benign prostatic hypertrophy. MRI showed a pancreatic head mass with double duct sign, aortic thickening, bilateral renal lesions, diffuse lymph node enlargement, and prostatic enlargement. FDG-PET/CT demonstrated abnormal uptake corresponding to the MRI lesions, and there were elevated IgG4 levels on blood investigations. Biopsy of an inguinal lymph node revealed infiltrates with IgG4 plasma cells, consistent with the diagnosis of IgG4 disease. The patient was treated with IV steroids and showed significant improvement.
DISCUSSION: IgG4 related disease is a rare entity that is characterized by lesions that show heavy infiltration with IgG4 positive plasma cells, storiform fibrosis, and obliterative phlebitis. The pancreas is the most commonly involved organ, but several other organ systems are involved, and this helps in clinical suspicion of the diagnosis. A biopsy from any easily accessible site that shows the characteristic histological features is sufficient for diagnosis. Patients respond quickly to steroids, but recurrence is frequent.
CONCLUSION: IgG4 related disease is a rare cause of pancreatic tumorous lesions that need a high index of suspicion for diagnosis and should be differentiated from pancreatic neoplastic lesions.
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1. Introduction

Most patients with a pancreatic mass pose a diagnostic challenge when a benign lesion is suspected, and often, resection is needed before a benign pathology is confirmed. IgG4 Related Disease (IgG4-RD) is a rare cause of pancreatic mass lesions that should be kept in the differential diagnosis, as a pancreatic biopsy may be avoided if the disease is suspected and identified.

The patient in this case report was treated at our General Hospital, which is an academic medical center and part of a Medical Corporation. This manuscript has been reported following the SCARE guidelines [1].

2. Presentation of the case

We are presenting the case of a 57 years old male patient who was referred to us with a pancreatic head mass and obstructive jaundice. He is a known case of hyperuricemia, gouty arthritis, chronic renal impairment, Diabetes Mellitus, and hypertension.

His condition started about two years before this presentation with recurrent nasal obstruction and eye dryness and itching, with multiple visits to the emergency department. He was diagnosed as allergic rhinitis and conjunctivitis and was maintained on antihistamines and eye drops. Six months after onset, he devel-

Abbreviations: IgG4, immunoglobulin-G4 related disease; FNAC, fine needle aspiration cytology; IgG, immunoglobulin G; BPH, benign prostatic hyperplasia; AST, aspartate aminotransferase; ALT, alanine aminotransferase; MRI, magnetic resonance imaging; MRCP, magnetic resonance cholangio-pancreatography; GFR, glomerular filtration rate; AFP, alfa feto protein; CEA, carcinoembryonic antigen; HBV, hepatitis B virus; HCV, hepatitis C virus; EUS, endoscopic ultrasound; FDG-PET, fluorodeoxyglucose positron emission tomography; AIP, autoimmune pancreatitis.

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oped bilateral submandibular swellings, more on the left side, and cervical ultrasound examination showed enlargement of multiple cervical lymph nodes with an appearance most likely representing proliferative lymphadenopathy. Fine needle aspiration cytology (FNAC) did not show any evidence of malignancy, and the diagnosis of reactive lymphadenopathy was confirmed. A few months later, the patient presented to the dermatology clinic with hyperpigmented skin lesions in the groin and hyperkeratotic lesions in both feet and received topical treatments with no improvement. He was kept on regular follow up and was seen by hematology to rule out the possibility of malignancy, and multiple investigations were done, including a serum immunoglobulin levels that showed a high IgG level of >2000, but no diagnosis was reached at that time. The patient again frequently presented to the emergency department with recurrent symptoms of eye pain and redness and was treated with antihistamines and antibiotics. He also developed obstructive urinary symptoms, and was diagnosed with benign prostatic hypertrophy (BPH) and was started on tamsulosin.

One month before his presentation, routine lab tests showed high AST and ALT levels, and he was referred to Gastroenterology. By the time he was seen he had developed jaundice, dark urine, and pale stools, with 8 Kg unintentional weight loss (over four weeks) and mild dilatation of the biliary system on ultrasound examination. MRI/MRCP (Fig. 1) revealed dilated biliary and pancreatic ducts (double duct sign) with a pancreatic head mass, multiple wedge-shaped lesions in both kidneys, significant aortic thickening suggestive of aortitis/Peri-aortitis, and cervical, mediastinal and intra-abdominal lymph node enlargement. His liver function tests were significantly abnormal (total bilirubin: 151, direct bilirubin: 113, ALP: 251, ALT: 30, AST: 50), and renal functions tests still showed renal impairment with a GFR of 51 ml/min, but the blood cell counts were normal. Serology for HBV and HCV were negative, and his AFP, CA 19-9, CA 15.3, and CEA were within normal. Total IgG was elevated at 2560, with elevation in all the subclasses as follows: IgG1: 1420, IgG2: 831, IgG3: 265, IgG4: 1980 (mg/dl). C3 and C4 were within normal, and his urine analysis showed no proteinuria. Endoscopic ultrasound (EUS) showed enlarged mediastinal lymph nodes and a bulky pancreatic head suggestive of pancreatic malignancy or autoimmune pancreatitis, and EUS guided FNAC samples taken from both sites showed only a few atypical cells, with no evidence of malignancy or metastatic carcinoma. An FDG PET/CT scan (Fig. 2) showed intense circumferential uptake in the abdominal aorta corresponding to the MRI findings, and suggestive of aortitis. Hypermetabolic enlarged lymph nodes were seen above and below the diaphragm as well as an irregular small pulmonary infiltrate, and the lacrimal and major salivary glands were also hypermetabolic. The pancreatic head mass and the renal cortical lesions seen on MRI showed intense FDG uptake as well, and the overall picture was suggestive of lymphoma.

The patient was discussed in the hepatobiliary multidisciplinary meeting with the principal differential diagnoses reached being lymphoma or Immunoglobulin-4 related disease (IgG4-RD), and lymph node biopsy was recommended. He underwent inguinal lymph node biopsy. Histopathological examination of the lymph node, which measured 1.8 cm in maximum dimension, showed many lymphoid follicles with prominent germinal centers consistent with reactive follicular hyperplasia. In addition, there was a marked expansion of paracortical areas with a dense plasma cell infiltrate. Areas of fibrosis including fibrotic thickened blood vessels were noted. The inflammatory infiltrates extended into para-nodal tissue. Immunohistochemistry studies confirmed a reactive nature of the lymphoid follicles (CD10+, BCL6+, and BCL2-). The plasma cells-infiltrate was CD138+, most of which were IgG positive cells and more than 30 IgG4 positive cells were seen per high power field. Kappa and Lambda immunohistochemical stains showed no light chain restriction. The overall morphology and Immunohistochemistry were consistent with IgG4-related disease (Fig. 3).

These findings confirmed the diagnosis of IgG4-RD with multiple manifestations including a pancreatic lesion, aortitis and peri-aortitis, bilateral renal lesions, diffuse lymph node involvement, lacrimal and salivary gland lesions, and cutaneous manifestations, with a possible pulmonary lesion. The patient was started on intravenous pulsed steroids (Methylprednisolone 500 mg daily) for three days, followed by oral prednisolone 40 mg daily and he showed a rapid clinical response. The bilirubin level dropped to 54 mmol/l at four weeks. Eight months after the onset of treatment, the patient was asymptomatic, with complete resolution of the lymphadenopathy, jaundice, skin lesions, allergic and conjunctival manifestations, and the obstructive urinary symptoms.

3. Discussion

IgG4 related disease is a recently described entity, first designated in 2003, that includes variable organ manifestations, many of which used to have different groupings and nomenclatures [2]. The diagnosis relies on the characteristic pathological features that are identical in all of the involved organs and include: heavy plasmacytic infiltrates with IgG4 producing plasma cells, storiform fibrosis, and obliterative phlebitis [3]. The possible mechanisms for etiology and pathogenesis include autoimmunity (whether being an autoimmune disorder itself or a down-regulatory mechanism for another autoimmune entity), and allergy, but this is still not fully understood [3,4].

The disease affects many organs and organ systems in the body, and this leads to variable presentations that may seem nonspecific or unrelated, contributing to the long delay before diagnosis, as seen in this patient who was diagnosed after two years of follow up and investigations. However, the most common manifestations are autoimmune pancreatitis, salivary gland disease, orbital/lacrimal gland disease, and retroperitoneal fibrosis [5,6]

This patient presented at the age of 57 years, which is consistent with the epidemiology of the IGG4-RD as it mainly affects middle-aged and elderly males, except in limited cervicofacial disease, where men and women are affected equally [7]. At presentation, multiple organ involvement is present in 60–90% of the cases, and most of the patients have lymphadenopathy, most frequently involving the cervical, mediastinal, and retroperitoneal lymph nodes. However, some patients may present initially with lymphadenopathy only, as seen in this patient, making the diagnosis difficult, even with a biopsy. Weight loss is common and progressive during the long workup stage despite the patient being generally well with no systemic manifestations, as evident in this patient. Allergic symptoms were also noted in this patient and as have been described in many patients with IGG4-RD [7,8].

Autoimmune pancreatitis is the typical and most common manifestation. It can present with diffuse enlargement of the gland or as in our case, with a pancreatic mass, making differentiation from pancreatic cancer difficult [9]. Closely associated is IgG4 related sclerosing cholangitis, which should be distinguished from primary sclerosing cholangitis and cancer. The diagnosis is challenging but is mainly based on identifying the characteristic histological features in other organs due to the difficulty in obtaining adequate biopsies from the bile ducts [10].

Salivary and lacrimal gland involvement is also common in IgG4-RD. It may present with dryness, enlargement, and tumors, and is one of the causes of proptosis and orbital pseudotumor [11]. Lacrimal gland involvement most likely explains the recurrent eye symptoms in our patient, especially that the PET scan showed

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