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A rare presentation of IgG4 related disease as a gastric antral lesion: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Immunoglobulin G4 related disease is a recently recognized systemic fibro-inflammatory disorder affecting virtually every organ in the body, characterized by lympho-plasmacytic dense infiltrates rich in IgG4 positive plasmacytes along with storiform fibrosis, inconstantly associated with elevated serum IgG4 levels. Few cases of Immunoglobulin G4 related disease occurring solely in the stomach have been published.
PRESENTATION OF CASE: We herein present a rare case of a 57 year old male patient presenting with an incidentally discovered asymptomatic pre-pyloric submucosal gastric lesion confused with a gastro-intestinal stromal tumor with failed endoscopic biopsy attempts due to tumor mobility. The patient underwent wedge resection of the lesion which was diagnosed postoperatively as Immunoglobulin G4 related disease.
DISCUSSION: Immunoglobulin G4 related disease presenting as a solitary lesion in the stomach is a very rare condition. It should be kept in the differential diagnosis of a submucosal mass or polyp. The treatment is medical with systemic steroid therapy.
CONCLUSION: Obtaining a tissue biopsy is of extreme importance to avoid unnecessary surgery.

1. Introduction

Immunoglobulin G4 related disease (IgG4RD) is a recently recognized systemic fibro-inflammatory disorder affecting virtually every organ in the body: the pancreas, biliary tree, stomach, kidneys, retroperitoneum, prostate, aorta, lymph nodes, meninges, thyroid, salivary glands, lungs, breast, pericardium and skin [1,2]. Histologically, irrespective to disease site, it is characterized by lympho-plasmacytic dense infiltrates rich in IgG4 positive plasmacytes along with storiform fibrosis and obliterative phlebitis [3]. IgG4RD is associated but not always with elevated serum IgG4 levels. The clinical manifestation depends on the site of occurrence, the infiltrates causing swelling, enlargement, thickening or nodules in the affected organs [4]. It typically affects the pancreas causing autoimmune pancreatitis (AIP) with diffuse IgG4 deposition in adjacent organs such as the stomach [3]. IgG4 related sclerosing disease can be associated in the stomach with gastritis and gastric ulcer [5,6], atypically and very rarely it may present as a focal submucosal gastric mass [7]. We herein present a rare case of IgG4RD presenting as an isolated submucosal pre-pyloric mass. This work has been reported in line with the SCARE criteria [8].

2. Case presentation

A 57 year old man was referred to our clinic for a gastric antral lesion discovered incidentally on a thoraco-abdominal CT scan performed on a routine follow up of prostate cancer. The patient was feeling well and he denied any nausea, vomiting, abdominal pain, fatigue or satiety. His past medical history was remarkable for hypertension and prostate cancer operated 5 years ago and classified as T3N1 with R0 resection, treated post-operatively by

Abbreviations: AIP, autoimmune pancreatitis; BMI, body mass index; CA19.9, cancer antigen 19.9; CDC, comprehensive diagnostic criteria; CEA, carcino-embryogenic antigen; CT, computer tomography; HPF, high power field; IgG4RD, IgG4-related disease; GIST, gastro-intestinal stromal tumor; SMA, smooth muscle antigen; FNA, fine needle aspiration.

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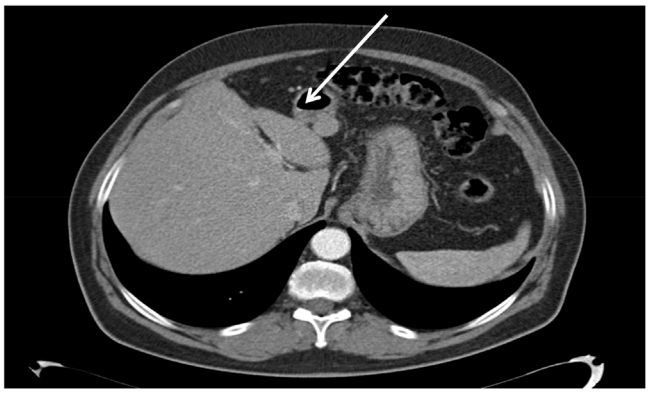

Fig. 1. Abdominal tomography examination showing the well-defined gastric antral lesion.

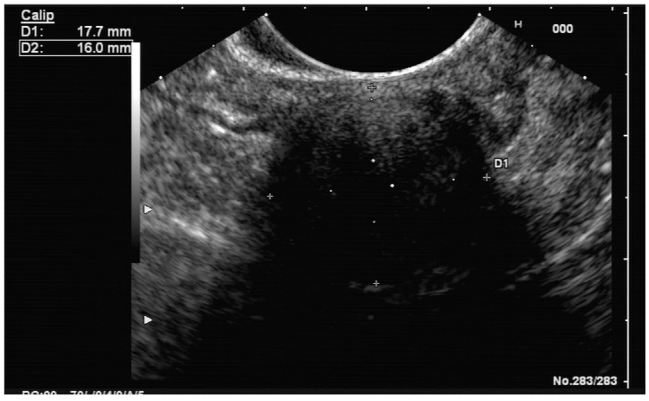

Fig. 2. Endoscopic ultrasound examination of the mass showing the heterogeneous oval shaped intra-parietal lesion, measuring 17.7 × 16 mm on large diameters.

hormonal ablation therapy. Physical examination showed a comfortable obese man with a BMI of 31 kg/m^2, blood pressure of 130/70 mm Hg, heart rate of 80 bpm. The abdomen was soft, non-tender and not distended. There was no palpable mass, no enlarged liver nor spleen. His laboratory examination tests were normal with a hemoglobin level 14 g/dl, platelet count: 409,000/mm^3, leucocyte count: 7100/mm^3 with neutrophils, lymphocytes and monocytes at 58.1%, 30% and 8.2% respectively, LDH: 179 U/L. Liver function tests were all normal. Abdominal CT showed an 18 mm well defined lesion at the gastric antrum with arterial enhancement, suggestive of GIST (Fig. 1). An endoscopic ultrasound (EUS) was performed using a linear-array US scope (EG-3870UTK, Pentax Medical & Hitachi Hi Vision 8500 US machine). On endoscopic vision, the lesion was identified under the Incisura Angularis on the lesser curvature of the stomach. EUS revealed an intraparietal lesion developing from the muscularis propria, measuring 17.7 × 16 mm on largest diameters, oval, with sharp margins and regular edges, mainly hypoechoic, with heterogeneous content (Fig. 2). This lesion extended into the extra-digestive area without infiltrating other structures. No lymphadenopathy was noted. A trial of EUS guided FNA, using the 22 G ProCore Needle from Cook (EchoTip® ProCore™; Cook Endoscopy, IN, USA) was attempted but ended in failure due to the hypermobility of the lesion and its difficult location. A retrospective review of the scans done earlier showed the same mass to be present since 4 years.

In front of a very slow evolution, the patient consented to undergo a laparoscopic partial gastrectomy, for a wedge resection of a probably benign tumor.

The laparoscopic exploration of the abdomen showed absence of peritoneal carcinomatosis and of liver metastases. The mass

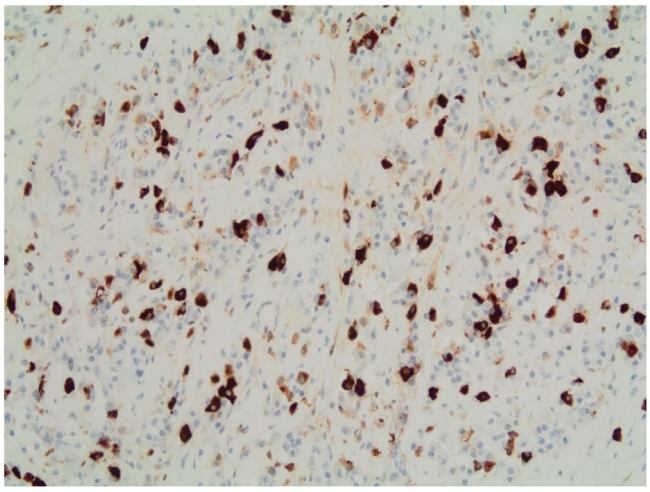

Fig. 3. Immuno-histochemical staining for IgG4 revealing the presence of numerous positive plasmacytes.

was identified at the level of the lesser curvature. The stomach was mobilized after division of the lesser omentum. A mechanical laparoscopic wedge resection using GIA was avoided due to the risk of having a sort of "Hourglass" stenosis at the body of the stomach. A small laparotomy incision was done and the stomach was exteriorized through a protective bag. A wedge resection of the small curvature with safety margins was done. A frozen section showed negative margins. The gastric edges were approximated using 2 layers of continuous suture by vicryl 2.0, then by silk 2.0 sutures.

The post-operative course was uneventful and the patient was discharged on the 5th post-operative day, with a mashed diet regimen.

The definitive histopathologic examination report demonstrated the homogenous extra-mucosal lesion of 18 mm to be composed of collagen and an inflammatory infiltrate of numerous lympho-plasmacytes.

On immuno-histo-chemistry analysis, the tumor stained negative for anti CD117 (c-Kit protein), SMA, CD34, S100. Numerous inflammatory cells stained positive for anti CD138 along with anti-IgG4 with a ratio of IgG4/IgG total >40%, and a number of IgG4 positive cells >50/HPF, which proved the tumor to be an IgG4-related inflammatory pseudo-tumor (Fig. 3).

Serum IgG4 level done 45 days post-surgery was: 118 mg/dL (normal 14–126 mg/dL) and after 5 months: 128 mg/dL. At 1 year post-surgery, the patient was doing well, with no signs of recurrence on abdominal computed tomography examination.

3. Discussion

IgG4 is the least abundant IgG in healthy individuals (< 5% of total IgG) [9]. It is linked to other autoimmune diseases like pemphigus vulgaris, pemphigus foliaceus, idiopathic membranous glomerulonephritis, and thrombotic thrombocytopenic purpura [10]. In 2001, Hamano et al linked AIP to elevated serum concentration of IgG4 [11]. Kamisawa et al detected severe or moderate infiltration by IgG4 positive plasma cells, along with storiform fibrosis in the pancreas, peri-pancreatic tissue, biliary tree, salivary glands, lymph nodes, colonic and gastric mucosa, and recognized the AIP as being a manifestation of a systematic autoimmune fibro-inflammatory process. The disease is similar to sarcoidosis in the way that one or multiple organs can be affected and they share the same histopathologic features, it can affect [10]. In opposite to most of the autoimmune diseases, IgG4 RD affects mostly men above 50 years of age [12]. IgG4RD is excessively misdiagnosed because of