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Case Report

IgG4 porta-hepatis pseudotumor mimicking a hilar cholangiocarcinoma ☆

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ABSTRACT

A 60-year-old female with a BRCA2 mutation and a history of breast cancer presented with diffuse abdominal pain and elevated liver enzymes. Imaging revealed a porta-hepatis mass, prompting consideration of hilar cholangiocarcinoma or breast cancer metastasis. Further investigation including biopsy and ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography findings were inconsistent with malignancy, leading to investigation of non-neoplastic causes. Elevated IgG4 levels suggested IgG4-related disease, a mass-forming fibroinflammatory condition. This case demonstrates IgG4-related disease exclusively impacting the portal vein and underscores the importance of considering IgG4-related disease in the differential diagnosis of hepatic masses.

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Introduction

IgG4-related disease (IgG4-RD) is a chronic inflammatory condition characterized by tissue infiltration with IgG4-positive plasma cells, fibrosis, and elevated serum IgG4 levels. This systemic disease can affect any organ, presenting a broad spectrum of clinical manifestations that mimics malignancies and other inflammatory conditions. While the pancreas and hepatobiliary system are the most involved sites, exclusive involvement of the portal vein has not been documented. In this case report, we present a unique instance of IgG4-RD manifesting solely affecting the portal vein.

Case presentation

A 60-year-old female with a medical history significant for BRCA2 mutation and breast cancer presented with diffuse abdominal pain and elevated liver enzymes. Abdominal ultrasound (Fig. 1) showed a heterogeneous porta-hepatis mass with internal color Doppler flow and cavernous transforma-

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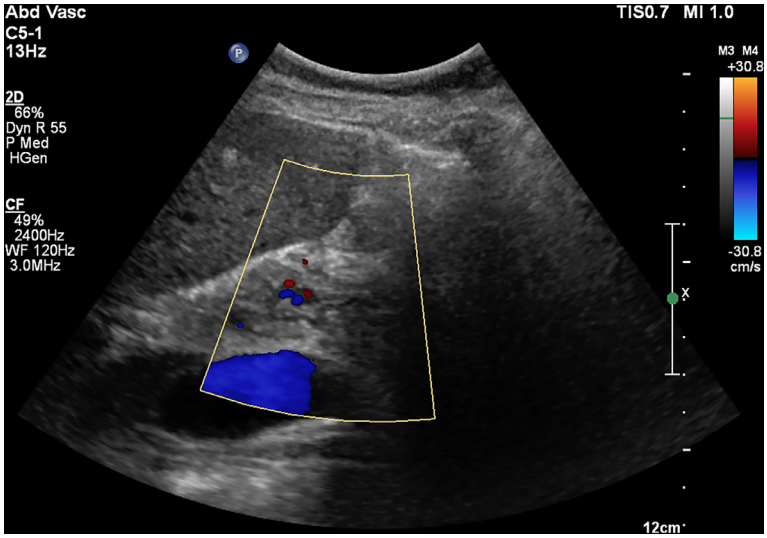

Fig. 1 – Abdominal ultrasound demonstrates a heterogenous porta-hepatis mass.

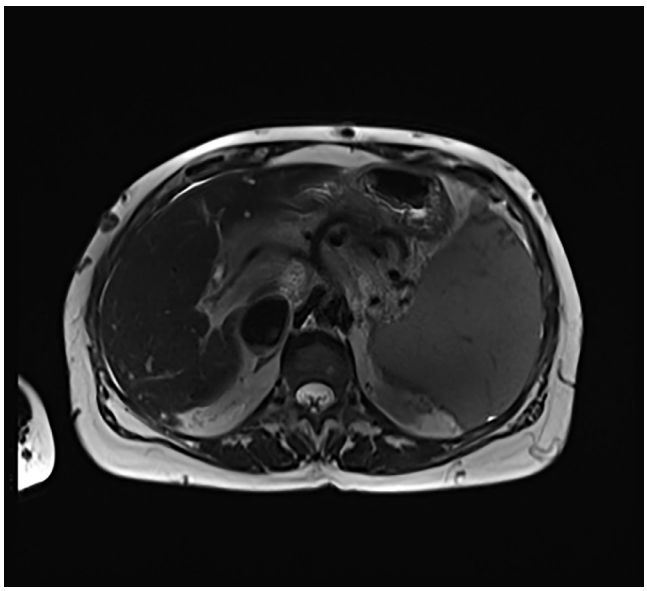

Fig. 2 – Axial T2 MRI demonstrates a T2 hyperintense, mildly enhancing porta-hepatis mass.

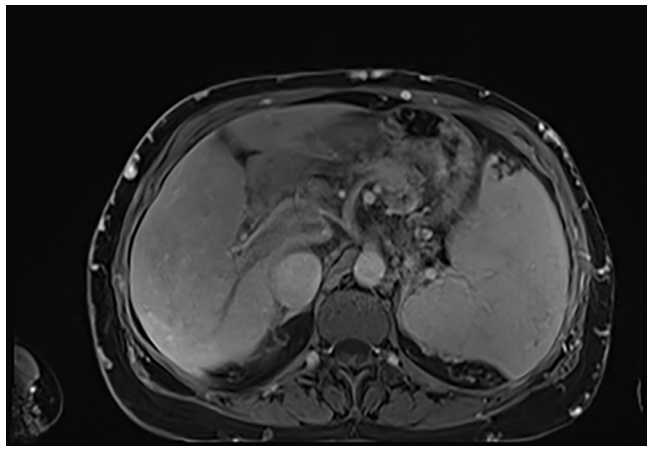

Fig. 3 – Postcontrast MRI shows the mass occluding the portal vein and encasing the common hepatic artery and common bile duct without upstream dilatation.

tion of the portal vein concerning for malignancy. Follow-up magnetic resonance imaging (MRI) demonstrated an ill-defined T2 hyperintense porta-hepatis mass, compressing the portal vein with subsequent perisplenic and perigastric collateral formation and associated splenomegaly (Figs. 2 and 3). Based on these findings, concern for hilar cholangiocarcinoma versus breast cancer metastasis was raised. Interestingly, the mass compressed only the portal vein, and encased the more rigid structures, including the common bile duct, without occlusion or upstream dilatation often seen with cholangiocarcinoma.

A CT-guided biopsy of the hilar mass was performed and demonstrated dense fibrotic tissue without evidence of malignancy. ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) was performed, showing low-grade uptake of 2.5 SUV max (Fig. 4), inconsistent with either hilar cholangiocarcinoma or breast cancer metastasis. Serum tumor markers including cancer antigen 125, cancer antigen 19-9, carcinoembryonic antigen, alpha-fetoprotein, and chromogranin A were within normal limits.

This led investigators to explore non-neoplastic etiologies. Tests for various autoimmune conditions, including anti-mitochondrial antibodies and anti-smooth muscle antibodies, were within normal limits. A serologic assay for IgG showed elevated total IgG, normal IgG1-3, and elevated IgG4 levels at 181 mg/dl, suggesting IgG-4 related disease (IgG4-RD).

Discussion

IgG4-RD is a multiorgan fibroinflammatory condition characterized by diffuse or focal inflammatory mass forming reac-

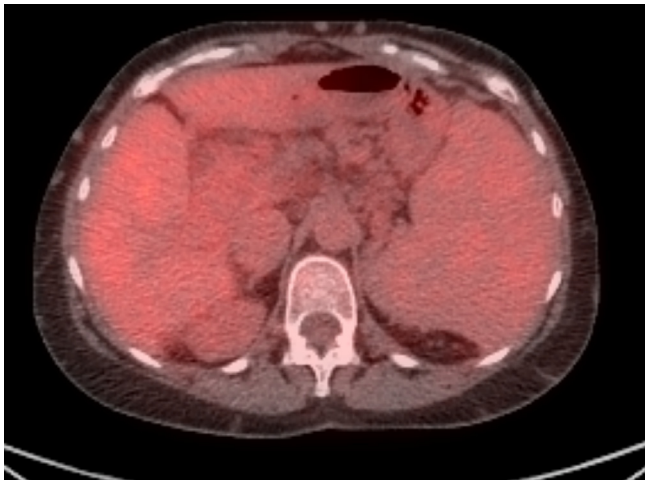

Fig. 4 – No significant ^{18}F-FDG uptake seen on ^{18}F-FDG PET-CT.

tion with fibrosis and dense lymphoplasmacytic infiltrate rich in IgG4 cells and elevated IgG4 levels [1]. Sarles et al first described such a case in 1961 in a patient with pancreatitis associated with hypergammaglobulinemia. Elevated IgG4 levels later emerged as a diagnostic marker for patients with autoimmune pancreatitis [2,3]. Although the pancreas is the most frequently cited affected primary organ, IgG4-RD can impact almost any organ, with multiple organ involvement observed in 60%-90% of patients. Following the pancreas, the hepatobiliary system is the most affected [4,5]. IgG4-related sclerosing cholangitis is the most common extra-pancreatic manifestation; with imaging typically demonstrating soft tissue encasing the common bile duct walls, long strictures, and upstream dilatation. Other hepatobiliary manifestations include hepatic inflammatory pseudotumor and hepatopathy [6].

Diagnosis of IgG4-RD is based on clinical and imaging findings, serological studies, and histopathological analysis. Up to 70% of patients have elevated serum IgG4 levels above 1.4 g/L, though normal levels do not exclude the diagnosis [7]. In patients with elevated serum IgG4 levels, subsequent levels can be trended to assess for response to treatment. CT and MRI can detect organ involvement, often revealing diffuse or localized swelling, masses, or nodules, but cannot differentiate between specific causes [8]. Further evaluation of organ involvement can be performed with ^{18}F-FDG PET/CT, though it is not routinely used due to challenges in interpretation from normal variants and physiological uptake [6]. In this case, ^{18}F-FDG PET/CT was helpful in reducing suspicion of carcinoma given the patient's history of cancer.

Updated diagnostic criteria for IgG4-RD was proposed by Umehara et al. in 2020 and consists of three domains; 1) Clinical and radiological features, 2) Serological diagnosis, and 3) Pathological diagnosis. Clinical and radiological features include "one or more organs with diffuse or localized swelling, or a mass or nodule characteristic of IgG4-RD," with lymph node swelling omitted in cases with single organ involvement. To fulfill criteria in domain 2, serum IgG4 levels must be greater than 135 mg/dL. Pathological diagnosis is the last domain, and must be positive for 2 of the 3 criteria; 1) dense lymphocyte and plasma cell infiltration with fibrosis, 2) ratio of IgG4-positive plasma cells/IgG-positive cells greater than 40% and the number of IgG4-positive plasma cells greater than 10 per high powered field, and 3) typical tissue fibrosis, particularly storiform fibrosis, or obliterative phlebitis. Diagnosis is considered "definite" in patients who meet all 3 domains, "probable" in patients who meet clinical criteria and pathological features, and "possible" in patients who meet clinical and serological criteria [9]. Our patient met all three criteria, thus the diagnosis for IgG4-RD was made.

Early recognition of IgG4-RD with prompt initiation of treatment is critical to reduce the damage to the affected organ systems. Treatment for IgG4-RD typically includes glucocorticoids, with a response usually observed within 1 month of initiation. Glucocorticoid-sparing therapy, such as rituximab, can be used, particularly in patients with chronic conditions like diabetes, hypertension, and osteoporosis, to limit the negative effects from long-term steroid use [7]. Also, patients often need treatment for complications from IgG4-RD. In this patient, pharmacologic management for gastric varices and periodic paracentesis for ascitic fluid accumulation were initiated in addition to daily prednisone. While IgG4-RD cases involving the biliary ducts mimicking cholangiocarcinoma have been reported, this case exclusively affecting the portal vein highlights the importance of considering IgG4-RD in the differential diagnosis of porta-hepatic masses.

Conclusion

This unusual presentation illustrates the diagnostic challenges posed by IgG4-RD, particularly when it mimics more common hepatic pathologies. Through this report, we emphasize the importance of considering IgG4-RD in the differential diagnosis of isolated porta-hepatic masses, thereby contributing to a broader understanding of the disease's diverse presentation.

IRB approval

The subject has been acknowledged and approved by the IRB.

Patient consent

Approval was obtained prior to case report submission.

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