A Diagnostic Conundrum: Pancreatic Retroperitoneal fibrosis masquerading as a metastatic pancreatic tumor.

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Introduction

- Retroperitoneal fibrosis (RPF) is a rare condition characterized by inflammation and the progressive development of a fibrotic mass in the retroperitoneal space.
- Due to the wide variety of presentations, diagnosis remains challenging, difficult, and often delayed. This case highlights an unusual presentation of this rare entity.

Case Description

- A 72-year-old female, with a history of hypothyroidism, presented with epigastric pain and distention of 6 weeks' duration. Pain was associated with fatigue and 24 lbs of weight loss over 6 months. She denied vomiting, jaundice, anorexia, and bloody stool.
- Physical examination was notable for diffuse abdominal tenderness and ascites without stigmata of chronic liver disease. Blood work revealed normal CBC, CMP, and hepatitis panels.
- A CT scan of the abdomen revealed multiple hypodense, ill-defined masses scattered throughout the liver. There was associated soft tissue fullness of the pancreas with CBD and IHD dilatation and a moderate volume of ascites.
- Follow up MRI showed a mass in the root of the mesentery which appeared to be originating from the pancreas, along with focal fatty infiltration of the liver.
- CA 19–9, CEA, and CA 125 were unremarkable.
- She underwent a liver biopsy which showed steatohepatitis without evidence of malignancy.
- A biopsy of a suspected pancreatic mass showed evidence of acute inflammation and was negative for malignancy, with some areas of chronic inflammation and fibrosis.
- Repeat EUS-FNB returned with nonmalignant findings. Ascitic fluid cytology was negative for malignancy.

Continue…,Case Description

- During this course of time, she has had numerous doctor visits, including multiple hospitalizations, without a definitive answer. Portal HTN with ascites was thought to be secondary to cryptogenic cirrhosis versus NASH versus autoimmune. She developed multiple episodes of spontaneous bacterial peritonitis, intra abdominal venous thrombosis.
- Repeated CT abdomen 3 months later showed extensive desmoplastic reaction and fibrosis in the root of the mesentery, obliterating the superior mesenteric vein and splenic vein and producing cavernous transformation of the portal vein.
- Serum IgG and IgG4 were elevated to 1723 and 1568.9, respectively. ANA-specific antibodies, ANCA, and cardiolipin antibodies were negative. An IgG4 immunohistochemical analysis was performed but remained inconclusive with an average of 6 IgG4 and 22 IgG plasma cells per HPF due to insufficient sample tissue. ESR, CRP, and ANA were elevated.
- The mass-forming lesion was diagnosed as IgG4-related retroperitoneal fibrosis. Infectious and malignant etiologies for RPF were excluded.
- Treatment with prednisone was started with a very good clinical response. Inflammatory markers and immunoglobulin were improved. She was regularly followed up. Cellcept was started as a steroid sparing agent. Follow-up CT a year later showed a stable mass.

Figure

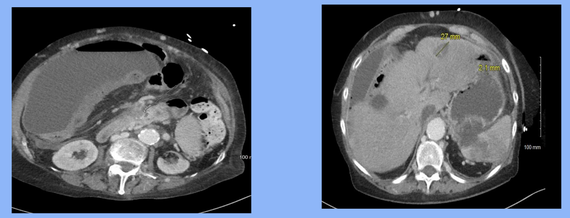

Figure : *CT scan of the abdomen showing multiple hypodense, ill-defined masses scattered throughout the liver. There was associated soft tissue fullness of the pancreas with CBD and IHD dilatation and a moderate volume of ascites.*

Discussion

- *Retroperitoneal Fibrosis is a rare fibroinflammatory disorder involving multiple organ systems.*
- *It is idiopathic in about two-thirds of cases; however, it may also be associated with other conditions, such as autoimmune diseases, atherosclerotic aortic disease, medications, malignancies, infections or radiotherapy.*
- *IgG4-related disease is now increasingly recognized as a cause of previously categorized idiopathic RPF.*
- *Diagnosis is by tissue biopsy, which demonstrates a dense lymphoplasmacytic infiltrate, IgG4-staining plasma cells.*
- *CT and MRI scans are important tools in the diagnosis. PET-CT is emerging as a recognized tool in identifying etiology, in assessing steroid responsiveness and in monitoring this disease.*
- *It is debatable whether a tissue biopsy is necessary for definitive diagnosis in a patient who has imaging studies demonstrating characteristics features of RPF like ours.*
- *Steroids are the mainstay of treatment. They are very effective in inducing remission.*

Conclusion

- *RPF encasing the SMA presenting as a pancreatic mass without ureteral or periaortic involvement has rarely been documented.*
- *In this case, the mass effect of the fibrosis resulted in portal hypertension and highlights the difficulty in diagnosing an exceedingly rare condition masquerading initially as a pancreatic malignancy.*
- *This patient had a protracted course involving multiple sub-specialties, numerous endoscopic procedures, and biopsies.*

Reference

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