Mind The Gap: A Case of Autoimmune Pancreatitis

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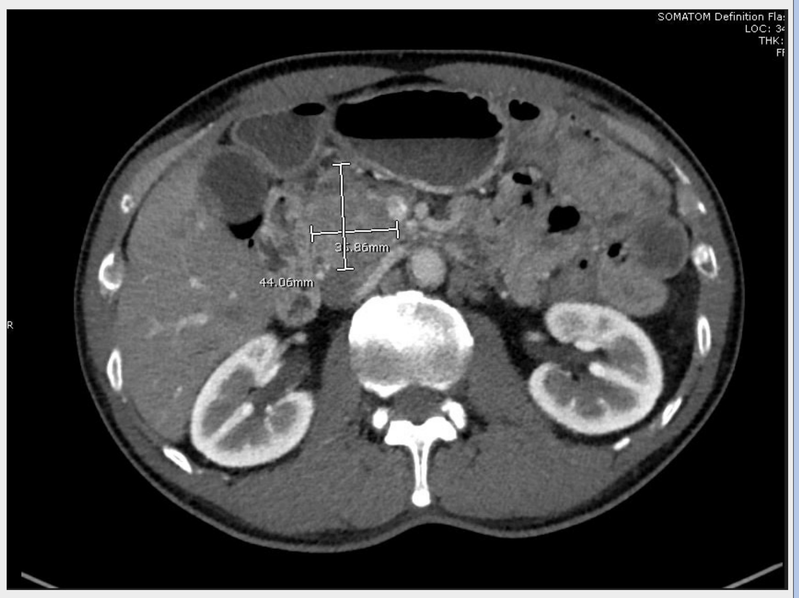
Introduction:

- Autoimmune pancreatitis (AIP) classically presents as painless jaundice, similar to its mimicker pancreatic cancer,
- The diagnosis and distinction between AIP and pancreatic cancer is often very difficult even beyond clinical presentation.
- Despite utilization of laboratory analysis and imaging, it is estimated that around 30% of AIP cases require core biopsy, steroid trial, or surgery to make definitive diagnosis.
- The high mortality associated with pancreatic cancer without treatment can spear-head more aggressive treatment.

Case:

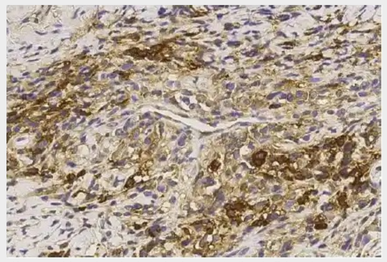
We present a case of a 63 year old male who underwent workup for postprandial abdominal pain and mild elevation of liver associated enzymes with normal bilirubin. He then had MRI imaging, which showed an irregular pancreatic head mass suspicious for malignancy that was further analyzed via EUS and biopsy showing some atypia but no definite evidence of adenocarcinoma. Tumor markers showed elevation of CA 19-9. He was referred to an oncologist and the decision was made that this was most likely pancreatic cancer. He was initiated on modified folfirinox and radiation therapy which led to reduction in size of the pancreatic head mass on repeat imaging. The patient eventually underwent Whipple. Labs after his surgery did not improve and actually worsened in terms of both cholestatic and hepatocellular patterns. The specimen demonstrated storiform fibrosis and lymphoplasmacytic infiltrate consistent with HISORt type 1 AIP as well as positive IgG4 staining. After referral to tertiary care center GI, an IgG4 level was found significantly elevated, consistent with AIP in addition to autoimmune cholangiopathy. Retrospective review of his outside facility imaging showed evidence of aortitis and lymphadenopathy. He had a robust response to steroids and eventually transitioned to rituximab with significant improvement in his clinical and biochemical markers.

Case Continued:

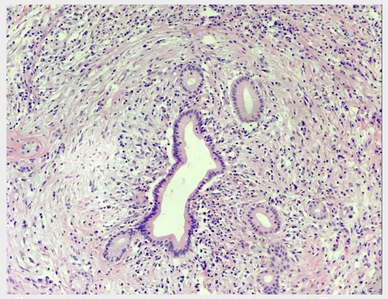


PRETREATMENT LABS	
AST	150
ALT	118
Alk Phos	1404
T. Bili	3.4
D. Bili	2.4
T. Protein	8.7
Albumin	4.1
IgG 4	688
Ca 19-9	21

POSTTREATMENT LABS	
AST	88
ALT	152
Alk Phos	611
T. Bili	1.7
T. Protein	6.8
Albumin	3.7
IgG 4	46



*Immunohistochemical study demonstrating IgG4 + plasma cells



*Demonstrates Lymphoplasmocytic Infiltrate seen in AIP type 1

Conclusion:

It is important to keep autoimmune pancreatitis on your differential for pancreatic mass evaluation. The persistent elevation in this patient's protein gap likely represented his elevated IgG4 level and may have been a clue to the diagnosis well before this patient's interventions. Ultimately, diagnosis is made with the HISORt criteria which is a mnemonic for its components: histology, imaging, other organ involvement and response to therapy. It has been proposed utilizing a combination of IgG4 and CA 19-9 in diagnosis of AIP to distinguish from pancreatic carcinoma with CA19-9 <74 and IgG4 >1.0 in combination showing a 100% PPV and a 96% NPV. This case demonstrates the importance of considering AIP during evaluation of pancreatic head mass to avoid delayed diagnosis, unnecessary treatment and patient harm.

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