Question 6 – October 5

A 72 year old gentleman with a longstanding history of chronic pancreatitis presents with a two-week history of jaundice. On further questioning, he reports previous episodes of jaundice and abdominal pain. He does not drink alcohol and does not have a history of IBD. He had a colonoscopy 6 months ago which was unremarkable.

Examination: Tired and slightly frail appearing gentleman who is clinically jaundiced.

Lab values: Hgb 11.4, WCC 6.9, Platelets 240,000, Total Protein 6.9, Albumin 3.7, Total Bilirubin 9.6 (7.2/2.4), AST 33, ALT 38, ALP 508, CA 19-9 normal

CT scan demonstrating changes consistent with chronic pancreatitis and intrahepatic biliary dilation. No pancreatic head mass is noted. ERCP demonstrated significant proximal extrahepatic bile duct stricture with diffuse intrahepatic biliary dilation. Cytology of the stricture reveals benign tissue. Which one of the following should be done next?

A. Order IgG4 levels
B. Start ursodeoxycholic acid
C. Start oral prednisone therapy
D. MRCP
E. EUS to evaluate pancreatic head

Answer: A

This is a case of IgG4 associated cholangitis and is an extrapancreatic manifestation of type 1 autoimmune pancreatitis. This gentleman presents with recurrent pancreatitis and an ERCP showing a distal bile duct stricture. His history is concerning for IgG4 associated cholangitis which presents with painless jaundice, proximal extrahepatic/intrahepatic biliary strictures and pancreatic changes. IgG4 levels should be checked and bile duct biopsies or pancreas FNA (if done) may be positive for IgG4 with a lymphoplasmacytic infiltrate.

Reference:

Immunoglobulin G4-associated cholangitis: clinical profile and response to therapy. Ghazale A, et al. Gastroenterology. 2008;134(3):706.