Case Presentation
A 27-year-old female with no medical history presented with one month of progressively worsening jaundice and a 30-pound weight loss. She was found to have conjugated hyperbilirubinemia with an acute cholestatic hepatitis. Ultrasound of the liver was unremarkable and MRCP showed normal appearing bile ducts. A liver biopsy was significant for features of cholestasis and ductopenia suggestive of vanishing bile duct syndrome. A computed tomography scan of the chest revealed a large complex anterior mediastinal mass with extensive lymphadenopathy (Figure 1). A core needle biopsy of the right supraclavicular lymph node was obtained and pathology consistent with Hodgkin lymphoma. She was initiated on induction therapy with improvement in her liver enzymes.

Etiologies of Vanishing Bile Duct Syndrome
Infection: Hepatitis C, CMV, sequalae of ascending cholangitis
Autoimmune Conditions: Primary biliary cholangitis, immune cholangitis, sarcoidosis
Transplant Associated: Acute and chronic liver rejection, graft versus host disease
Malignancy: Hodgkin Lymphoma as a paraneoplastic syndrome
Medications: Antibiotics, ibuprofen, anabolic steroids

Diagnostic Criteria: pathologic diagnosis defined by a paucity of interlobular bile ducts, defined as less than 50% of portal areas with a bile duct on biopsy.

Conclusions
• Patients with intrahepatic cholestasis and unrevealing initial workup should be considered for liver biopsy (Figure 2).
• Vanishing bile duct syndrome is a pathologic diagnosis with several potential etiologies. A thorough workup must be completed to determine the underlying cause.

ERCP
Cholestatic Pattern of Liver Injury:
Elevation in ALP greater in magnitude than AST and ALT

Ultrasonography
Mechanical Obstruction
Extrahepatic Cholestasis:
Evaluate with MRCP vs ERCP

Intrahepatic Cholestasis:
Obtain autoimmune workup including AMA, ASMA, ANA

Treat underlying cause

Alkaline phosphatase persistently >2 times upper limit of Normal?

Liver Biopsy

REFERENCES
5. Reau, N. Hepatic Ductopenia and Vanishing Bile Duct Syndrome. In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2018.