## 844

### Evaluating a Pseudo-Wilson Disease Patient: A Previous Undescribed Disease Association of Hallervorden-Spatz Syndrome (HSS) with Anti-m2 Positivity

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**Purpose:** Hallervorden-Spatz Syndrome is a rare neurodegenerative disease with 2 types of clinical presentation: early and late onset. In this subtype, psychiatric, pyramidal and extrapyramidal signs are present. A 56 years old female patient was referred to us to evaluate a possible Wilson's disease, because of liver function tests abnormalities (ALT and AST 2N; alk phos 1,5 N, GGT 8N) associated to recent psychiatric disturbances (mainly depressive type) and extrapyramidal signs (discrete spasticity, dysarthria and dystonia). No family history of neurological or autoimmune disorders was recorded; she was not taking any medication. She underwent extensive work up revealing high levels of anti-mithocondrial antibodies (>1/640) with positive anti-M2. Cholesterol was 240 mg/dl and tryglicerides were 8 times above normal. Liver biopsy showed panlobular steatosis with scarce fibrosis and no inflammation, namely on portal triads. Copper in serum, urine and liver measurements were normal. MRI showed bilateral symmetric hyperintense signal changes in globus pallidus, with surrounding hypointensity on T2-weighted images, the characteristic "eye-of-the-tiger" sign. Therapy was initiated to treat dyslipidemia, including weight loss, and after 6 months there has been a progressive normalization of liver tests (except GGT) and no worsening on the discrete neurological signs. To our knowledge it is the first description of this curious association between an iron metabolism disorder and a possible secondary copper accumulation disease (primary biliary cirrhosis).

## 845

### Spontaneous Retroperitoneal Variceal Bleed: A Case Report and Review of the Literature

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**Purpose:** There have been few reports of intraperitoneal rupture of ectopic varices as a complication of portal hypertension. Most have occurred in male individuals with alcoholic cirrhosis who died as a result of bleeding complications. We present a female patient with primary biliary cirrhosis who developed hemoperitoneum presumed to be from a ruptured retroperitoneal vessel demonstrated on CT which resolved with octreotide therapy. A 69 year-old female with a history of primary sclerosing cholangitis was initially admitted to the hospital for adynamic ileus. Upon presentation her blood pressure was 110/54 and heart rate was 100. She was a nonicteric, thin, elderly female who appeared chronically ill. Her abdomen was grossly distended, tense, tympanitic, and nontender. No rebound or guarding was noted. The liver and spleen were nonpalpable, no fluid wave was appreciated, and hypoactive bowel sounds were present. Initial admission labs were notable for: WBC 5,500 per mm3, hemoglobin 11.9 g/dl, hematocrit 36%, platelets 153, 000, INR 1.2, BUN 9 mg/dl, creatinine 0.6 mg/dl, total protein 7.5 g/dl, albumin 2.3 g/dl, alkaline phosphatase 319 u/l, AST 80 u/l, ALT 55 u/l, total bilirubin 3.2 mg/dl. Abdominal X-ray demonstrated dilation of small bowel loops consistent with adynamic ileus. The patient was initially managed conservatively with nasogastric tube decompression, NPO, and IV fluid with subsequent improvement in her adynamic ileus. However, on the third hospital day the patient's hematocrit dropped from 36.8% to 26.4%. She denied symptoms of gross bleeding and was without tachycardia or hypotension. Her physical exam was now notable for a new fluid wave with shifting dullness and worsening abdominal tenderness. Due to the concern for hemoperitoneum, a dual-phase CT of the abdomen and pelvis was performed. This demonstrated a large volume of hemoperitoneum, and a sentinel clot adherent to the parietal peritoneum consistent with hematoma from a ruptured retroperitoneal vessel. Subsequent abdominal paracentesis confirmed the presence of 2.5-liters of grossly bloody fluid with a hematocrit of 9.5%. Given the evidence for a spontaneous intraabdominal variceal bleed, octreotide was initiated with a bolus followed by continuous intravenous infusion for a total of 72 hours. The patient's serum hematocrit increased steadily throughout the remainder of her hospitalization and she did not require any further intervention including surgery or additional transfusions.

## 846

### Biliary Cyst (aka Choledochal Cyst): A Case Report of an Unusual Cause of End Stage Liver Disease in an 11-year old Female

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**Results:** A previously healthy 11-year old female with a two year history of intermittent sharp epigastric pain was found to have malrotation on barium examination of the upper gastrointestinal tract. At the time of laparoscopic surgical repair, her liver was noted to be grossly cirrhotic. Post-operatively, she developed jaundice and pruritus. On exam, she was jaundiced and had splenomegaly. Her total bilirubin was 14.8 mg/dL, with a conjugated fraction of 6.6 mg/dL, ALT was 75 U/L, AST was 98 U/L, alkaline phosphatase was 590 U/L, albumin was 2.3 g/dL, and INR was 1.22. Evaluation for chronic liver diseases was performed and negative. White blood count was 5.3 k/uL, hemoglobin was 12.3 g/dL, and platelets were 85/uL. She developed ascites and a right-sided pleural effusion; cell count and culture were not suggestive of infection. Abdominal ultrasound demonstrated cirrhosis, splenomegaly, dilated extrahepatic biliary ducts, and portal hypertension. MRCP showed a grossly dilated extrahepatic biliary tree, as well as marked intrahepatic biliary dilation, compatible with a choledochal, or biliary cyst. ERCP was performed, showing a massively dilated biliary tree, with an aberrant low takeoff of the cystic duct and distal stenosis of the common bile duct. Two stents were placed with relief of her obstruction. Her ascitic pleural effusion and ankle edema were controlled with diuretic therapy. After the procedure, her bilirubin showed a downward trend, but she developed recurrent episodes of cholangitis. Repeat liver biopsy demonstrated cirrhosis, with proliferation of bile ducts and neutrophilic infiltration, consistent with biliary cyst and ascending cholangitis. Due to her recurrent symptoms and underlying cirrhosis, she underwent liver transplantation.

**Conclusion:** Congential biliary cysts are irregular dilations of the bile ducts which are due to an abnormality in the ductal wall, possibly ductal stricture and thought to result from pancreaticobiliary malunion, which allows reflux of pancreatic juice into the bile duct, that can cause chemical and inflammatory changes in the duct wall, leading to weakness and dilation. To avoid the complications of these cysts, which include cholelithiasis, cholangitis, pancreatitis, intrahepatic abscess, biliary cirrhosis, portal hypertension and cancer, cysts should be excised surgically. This will involve bile duct resection with hepaticojejunostomy, as well as, partial hepatectomy in patients with intrahepatic involvement. In cases such as ours, with end stage liver disease, or those with Caroli disease, liver transplant is necessary.

## 847

### An Unusual Cause of Fulminant Liver Failure

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**Purpose:** Amyloidosis is a disorder of fibrillary protein deposition in various organ systems and it may be primary (AL) or secondary (AA). Primary amyloidosis can occur alone or in association with plasma cell dyscrasias such as multiple myeloma. Involvement of the liver has been reported in up to 70% of cases with AL amyloidosis. Fulminant liver failure is characterized by rapid development of hepatocellular dysfunction and encephalopathy in a patient with no prior history of liver disease. We report a case of fulminant liver failure due to amyloidosis in the setting of multiple myeloma. A 66-year-old white male with a 50-pack-year history of smoking presented with shortness of breath, atrial fibrillation, and hypotension. His past medical history consisted of hypertension, hyperlipidemia and benign prostatic hypertrophy. Two months prior he had been told of a mild elevation in his liver enzymes. He had not started any new medications in the past six months. His shortness of breath