Case Snippets

Castleman’s disease in a patient with celiac disease

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A 72-year-old man with celiac disease was doing well on gluten-free diet. Five years later, he developed dyspepsia, fever and weight loss. CT scan and laparoscopic biopsies of small mesenteric lymph nodes clinched the diagnosis of Castleman’s disease. He started on chlorambucil; unfortunately he died few months after the diagnosis was made. [Indian J Gastroenterol 2007;26:187]

Most patients with celiac disease develop satisfactory response to gluten-free diet. Unsatisfactory response raises the possibilities of poor dietary compliance, doubt about the diagnosis, presence of other coincidental diseases, ulcerative jejunitis, or development of small bowel lymphoma.

A 72-year-old gentleman, who had been diagnosed as celiac disease 5 years earlier, had responded well histologically and symptomatically to gluten-free diet. He started losing weight, developed dyspepsia, fever with night sweats, and had one episode of melena. Clinical examination was unremarkable except for ankle edema. Blood count, liver function tests, urea and electrolytes, and serum albumin were normal; platelets were 498 X 10^9/L (normal 150-400) and serum albumin was 23 g/L (30-50). Gastro duodenoscopy showed discrete healing ulcers in the second part of duodenum; histology showed partial villous atrophy.

Further blood tests showed hemoglobin 10.8 g/dL (13.5- 18.0), platelets 471 X 10^9/L (120-400), MCV 79 fl (80-98), neutrophils 1.8 X10^9/L (2.0-7.5), ESR 27 in first hour (<20), C-reactive protein 58 mg/L (0-8), ferritin 60.3 µg/L (10-350), vitamin B12 and folate levels were normal, and beta2 microglobulin level was 4.3 mg/dL (normal <2.6). Bone marrow biopsy was normal.

Barium follow-through showed ileum having multiple folds, which was described as “jejunization” of the ileum, and some narrowed loops of jejunum. With a possibility of ulcerative jejunitis, he was started on azathioprine and prednisolone. He showed some improvement in his weight but otherwise remained unchanged. Contrast-enhanced CT scan showed small mesenteric lymph nodes. Laparoscopic excision of the mesenteric lymph nodes was subsequently done; histology showed plasma cell type of multicentric Castleman’s disease.

Our hematologist started him on oral chlorambucil, he did not respond to treatment and passed away after a brief stay in hospital.

Castleman’s disease is a rare disorder characterized by benign lymph node enlargement. Its possible etiology has been mentioned as autoimmune, human herpes virus 8 infection (HHV-8) and uncontrolled inflammation. Castleman’s disease has not been reported earlier in patients with celiac disease. Localized Castleman’s disease (about 90% of patients) is classified into hyaline-vascular and plasma cell types; the former is usually asymptomatic. Patients with the plasma cell type present with constitutional symptoms, with 50%-90% having anemia, leukocytosis, hypoalbuminemia, and hypergammaglobulinemia. There is an abnormal IL-6 expression by cells in the germinal center and elsewhere in the lymph nodes in plasma cell type localized disease. The treatment is surgical removal.

Multicentric Castleman’s disease (MCD), with more than one site of disease with nodal involvement, can present with fever and weight loss, hepatosplenomegaly, high ESR, and hypergammaglobulinemia. Multicentric type has been associated with plasma cell disorders, POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes), autoimmune diseases, venous thromboses and bony abnormalities. Progression to lymphoma is higher in patients with multicentric type; 15-fold increase in lymphoma has been noticed in patients with HIV-positive MCD.

There is no standard treatment for the multicentric type. Various methods of treatment like radiotherapy, surgery, chemotherapy, steroids, anti IL-6 antibody and rituximab, a monoclonal antibody to CD20, have been reported to be effective. Therapy with gancyclovir has been reported to cause remission of HHV-8 and HIV-associated MCD.

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Spontaneous rupture of intrahepatic biliary ducts with biliary peritonitis

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Spontaneous rupture of intrahepatic biliary ducts is a rare cause of acute abdomen due to biliary peritonitis. We report a 92-year-old woman with 48-h history of upper abdominal pain, nausea and vomiting, and peritoneal signs. CT scan showed free fluid in the abdomen and mild dilatation of the common bile duct. Exploratory laparotomy showed bile in the abdominal cavity with leakage from a ruptured bile duct radicle in segment 3, as confirmed on intraoperative cholangiography. She underwent cholecystectomy, choledochotomy with removal of gallstones, repair of the perforation with primary suture, and placement of a T-tube. She had an uneventful recovery.

Spontaneous rupture of the hepatic duct is an uncommon and usually fatal cause of peritonitis. Only 40 cases with spontaneous perforation of the common bile duct (CBD) have been reported so far. Cholelithiasis, choledocholithiasis, and tumor obstruction of the ampulla have been reported as possible etiologies. However, spontaneous rupture of the intrahepatic bile duct is rare. We present a case with biliary peritonitis due to spontaneous rupture of intrahepatic bile duct in whom the diagnosis was made intraoperatively.

A 92-year-old woman was admitted with 48-h history of generalized abdominal pain, nausea and vomiting. Clinical examination revealed abdominal rebound tenderness and absence of bowel sounds. She was found to have mild icterus and severe dehydration. She was stabilized with resuscitative measures. Laboratory findings showed conjugated hyperbilirubinemia (total bilirubin 4.8 mg/dL, direct 3.1) and normal alkaline phosphatase. Plain X-ray abdomen did not show evidence of pneumoperitoneum. Ultrasonography revealed generalized free fluid in the abdomen and CBD dilatation. CT scan showed dilatation of the left lobe intrahepatic bile duct in addition.

At exploratory laparotomy, free intra-peritoneal bile was found. No rupture was found in the gall bladder or extrahepatic biliary tract. A perforated superficial biliary radicle in segment 3 of the liver was the source of the free bile. The gall bladder contained calculi; CBD was mildly dilated (10 mm) with no calculus in it. There was no stenosis of papilla or pancreatic mass. Cholecystectomy was done with exploration of CBD. Intraoperative cholangiography showed dilatation of the left hepatic bile duct with leakage from the intrahepatic biliary radicle of segment 3 (Fig). The CBD was drained with a T-tube and the ruptured biliary radicle was repaired with 3/0 prolene sutures. The right subhepatic space and pelvic region were drained. T-tube cholangiography on postoperative day 10 confirmed free passage of contrast into the duodenum. The tube was removed on postoperative week 6 after check cholangiography and normal liver function tests.

Peritonitis due to bile contamination commonly occurs post cholecystectomy because of imperfect clipping of the cystic duct. Other causes include post liver transplant, spontaneous hepatic rupture in pregnancy, and trauma to the extrahepatic biliary system at surgery. Perforation of the CBD is rare and etiologies include increased intraductal pressure, calculus erosion, and necrosis of the duct wall secondary to thrombosis. Spontaneous perforation of extrahepatic ducts is very rare. Only six patients with biliary peritonitis secondary to spontaneous intrahepatic duct rupture have been reported so far. The causative factors are calculus disease of the biliary ducts, stenosis of the papilla and Caroli’s disease. In our patient the reason for the spontaneous rupture of intrahepatic bile duct was not clear. It is possible that a calculus in the CBD may have caused its dilatation and then passed into the duodenum.

The left intrahepatic biliary ducts, especially those in segment 3, may be more prone to spontaneous rupture. This may be because the biliary system on the left side is closer to the surface than on the right side. In conclusion, in a patient with biliary peritonitis with intact gallbladder, meticulous investigation of whole biliary tract, particularly the left hepatic lobe, should be performed.
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Acute hemorrhagic ascites: look beyond the gastrointestinal tract

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A 33-year-old alcoholic man presented with acute abdominal pain and hemorrhagic, high serum-ascitic albumin gradient ascites, accompanied by hyperkalemia and azotemia. Spontaneous rupture of urinary bladder was diagnosed. The patient recovered uneventfully with conservative management. [Indian J Gastroenterol 2007;26:189-190]

The causes of acute abdomen with hemorrhagic ascites include acute necrotizing pancreatitis, cirrhosis with ruptured hepatocellular carcinoma, mesenteric ischemia with bowel gangrene, among others. Spontaneous rupture of urinary bladder is a rare cause of hemorrhagic urinary ascites.1 In most patients an underlying pathology is implicated; idiopathic rupture is rare, and often follows heavy alcohol ingestion the preceding night.

A 33-year-old man, chronic alcohol consumer, presented with acute abdominal pain for 2 days, which started the morning after an alcohol binge. Evaluation at another hospital revealed normal serum amylase and abdominal ultrasonography except for gross ascites. Paracentesis drained 1.5 liters of hemorrhagic fluid. He had persistent generalized abdominal pain, obstipation, oliguria and hematuria. He had no fever, jaundice, gastrointestinal bleed or altered sensorium. He had renal stone diagnosed 6 months back.

He was in acute distress; abdomen was distended with diffuse and rebound tenderness, and ascites was present. Bowel sounds were absent. Other systemic, urogenital and per rectal examination was unremarkable.

Investigations: Normal hemoglobin and white blood cell counts. Serum creatinine was 7.5 mg/dL (normal 0.5-1.6) and serum potassium was 5.8 mmol/L (3.8-5.4). There were tall-tented T waves on EKG. Arterial blood gas study revealed high-anion-gap metabolic acidosis. Liver biochemistry, serum amylase and lipase were normal. X-ray chest and abdomen were unremarkable. Ultrasonography with Doppler was normal, except for gross ascites. MRI revealed no evidence of pancreatitis.

Ascitic fluid examination showed cell count of 300 cells/mm³ (20% polymorphs, 80% lymphocytes), glucose 88 mg/dL (serum glucose 130), protein 1.0 g/dL (serum protein 6.2), albumin 0.5 g/dL (SAAG 3.0) and lactate dehydrogenase 109 U/L (serum 194). Culture and cytology of ascitic fluid were negative. Urine examination revealed plenty of RBCs with no WBC or casts.

He was managed with intravenous fluids and antibiotics, and hemodialysis was done for hyperkalemia. He developed obstructive voiding symptoms and had retention of urine. Foley’s catheter drained about 3 liters of blood-tinged fluid, following which the abdominal distension subsided. Repeat ultrasonography revealed minimal free fluid and renal functions started improving.

The possibility of intraperitoneal rupture of bladder was considered. The ascitic fluid-serum creatinine ratio was >1.0 (i.e. 6.3:4.4 mg/dL). Retrograde cystography revealed contrast leak from the dome of the bladder after instillation of 200 mL contrast (Fig). He was managed with catheter drainage and intravenous antibiotics, recovered uneventfully and was discharged with improving azotemia. He has remained asymptomatic for the last 1 year.

Fig: Retrograde cystourethrography – Contrast leak from dome of bladder into peritoneum (arrows)

The pathophysiology of rupture of urinary bladder in patients abusing alcohol is distinct.2
The bladder becomes markedly over-distended from the diuretic effect of alcohol. Patients are variably obtunded, which clouds their sensory cues to void. The scenario of being awakened from sleep with sudden onset of abdominal pain supports the concept of an atraumatic rupture. In most cases the rupture is intraperitoneal, as the bladder expands at its superior portion and eventually tears at the thinnest portion, the dome.1

Clinical features include diffuse abdominal pain and bilateral shoulder pain or hiccups due to diaphragmatic irritation. Most have inability to void and dysuria. The sudden relief of pain and distension on draining small amounts of blood-stained urine following catheterization, raises the possibility of bladder rupture.3,4 Voiding cystourethrography is the investigation of choice. Ascites:serum creatinine ratio of >1.0 is suggestive of intraperitoneal urinary leak.5 The delay in presentation and diagnosis results in significant reabsorption of urea and creatinine through the peritoneal surface and manifests as azotemia. Raised serum levels of ammonia and potassium have also been reported.4

Surgical repair of the bladder has shown good results. Conservative management is also advocated.

The presence of high-SAAG ascites in our patient caused diagnostic confusion with underlying liver disease in a person who has a history of alcoholism.

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Eosinophilic cholangiopathy – a report of two cases

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Eosinophilic cholangiopathy is an unusual and benign form of biliary disease characterized by peripheral blood eosinophilia and cholangitis. Dramatic response to steroids is the hallmark of the disease. We present two cases of eosinophilic cholangiopathy.

Peripheral blood eosinophilia and eosinophilic infiltration of the gastrointestinal tract and hepatobiliary system may be seen in association with a number of conditions,1-5 but may occasionally occur in isolation. When the hepatobiliary system is involved, a radiological and clinical picture compatible with cholangitis, cholecytitis or hepatitis may be observed.

Case 1: A 13-year-old boy with history of bronchial asthma, on investigation for persistent fever, was diagnosed to have an abscess in the left lobe of liver with stricture of the common bile duct (CBD) on abdominal CT. This was confirmed by ERCP (Fig), during which the stricture was dilated, brushings obtained for cytology, and bile culture and stenting was done. Despite the stent, the boy had recurrent attacks of abdominal pain and fever. He was given multiple courses of antibiotics and a complete course of anti-tuberculous drugs, with no relief. Liver biopsy revealed marked eosinophilia, periportal fibrosis and bile duct damage; lymph node and bone marrow biopsy also showed plenty of eosinophilic infiltrate. Peripheral eosinophilia associated tissue eosinophilia in the liver, lymph nodes and bone marrow raised the suspicion of eosinophilic cholangiopathy. Echocardiogram was normal. He was given a course of albendazole and diethylcarbamazine with no change. Subsequently, he was started on 1 mg/Kg of prednisolone.

At review after a month he was well with no symptoms. His absolute eosinophil count decreased to 400 cells/mm³ and liver function tests normalized. Repeat ERCP (Fig) showed resolution of the bile duct stricture and of the intrahepatic biliary dilatation. While

Fig: ERCP of Case#1. ERCP at the time of diagnosis shows a long irregular stricture of common bile duct and distal common hepatic duct, with thin cystic duct and small gall-bladder (left panel). Dilatation of proximal common hepatic duct, right and left hepatic ducts, and some second order branches, is noted. Repeat ERCP after therapy with steroids (right) shows resolution of bile duct narrowing and of proximal biliary dilatation.
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6 months, exclusion of other causes of eosinophilia, organ system involvement attributable to eosinophilic infiltration – were met in both the patients. Schrulen et al.\(^5\) reported a patient with HES, bowel disease resembling chronic inflammatory bowel disease, and cholangiopathy resembling primary sclerosing cholangitis, which responded clinically and symptomatically to hydroxyurea, the drug of choice in HES.

Obstructive jaundice in association with eosinophilic cholangitis has been reported.\(^6\)\(^,\)\(^8\)\(^,\)\(^9\) In all these patients the diagnosis was made after surgery for presumed malignancy. Our patient\(^1\) also had an ill-defined mass at laparotomy and biopsy was not suggestive of any specific pathology probably because the patient was on steroids. ERCP picture suggestive of primary sclerosing cholangitis, as in our patient, has also been reported.\(^1\)

The treatment of eosinophilic cholangitis has varied from conservative watchful waiting to the use of steroids and hydroxyurea. The treatment of eosinophilic gastroenteritis has varied from short repeated courses of steroids to long-term low-dose steroid therapy or the use of steroid-sparing agents such as montelukast, ketotifen or sodium cromoglycate.\(^7\)

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