Sacrococcygeal teratoma with anorectal malformation

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A 7-month-old child presented with imperforate anus, penoscrotal hypospadia and transposition, and a midline mucosa-lined perineal mass. At surgery the mass was found to be supplied by the median sacral artery. It was excised and the anorectal malformation was repaired by posterior sagittal anorectoplasty. Histologically the mass revealed well-differentiated colonic tissue. The final diagnosis was well-differentiated sacrococcygeal teratoma in association with anorectal malformation. [Indian J Gastroenterol 2003;22:27]

Key words: Perineal mass

A wide spectrum of malformations associated with anorectal anomalies have been reported and have been organized into the VATER or VACTERL complex. However, congenital perineal masses have been rarely reported in association with anorectal anomalies.¹

A seven-month-old male child was referred for treatment of an imperforate anus. The child had had a loop sigmoid colostomy constructed at birth. On examination, in addition to the imperforate anus, he had a 4 cm x 4 cm x 2 cm fleshy perineal/sacrococcygeal mass that appeared to be lined by mucus-secreting mucoa (Fig). Anterior to this mass was a penoscrotal hypospadia and penoscrotal transposition. Ultrasonography of the abdomen was normal. Distal cologram revealed a supralevator anorectal anomaly with rectourethral fistula; there was no communication with the perineal mass.

Posterior sagittal anorectoplasty combined with excision of the mass was done. The mass was attached to the coccyx, supplied by the median sacral artery and was separate from the blind rectal pouch. Histological examination of the excised mass revealed well-differentiated colonic tissue.

Sacrococcygeal teratomas are the most common perineal masses in newborn babies. These tumors are attached to the coccyx and the main blood supply is from a middle sacral artery. The degree of differentiation can vary from anaplasia to a well-differentiated cell line.² The features of the case under review are consistent with a sacrococcygeal teratoma, viz., a congenital perineal mass supplied by the median sacral artery, attachment to the coccyx and differentiation into colonic tissues.

Anorectal malformations have been found in association with sacrococcygeal teratomas¹ but there is no instance of an imperforate anus reported with a colonic duplication although a colonic duplication with rectourethral fistula has been reported.³

Embryologically, the sacrococcygeal teratoma could be the cause of the high anorectal anomaly and the associated penoscrotal transposition and hypospadia.¹

References


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Acute mesenteric venous thrombosis complicating endoscopic variceal sclerotherapy with absolute alcohol

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We report two patients, one with liver cirrhosis and another with extrahepatic portal vein obstruction, who developed acute mesenteric vein thrombosis following endoscopic variceal sclerotherapy with absolute alcohol. Both patients recovered after emergency laparotomy and resection of gangrenous bowel loop. [Indian J Gastroenterol 2003;22:27-29]

Key words: Esophageal varices, portal hypertension

Acute mesenteric venous thrombosis (MVT) is a potentially lethal complication following endoscopic variceal sclerotherapy (EST); less than 20 cases have been reported in English literature¹ and none from

Fig: Imperforate anus and perineal mass. The blind sacrum is also clearly visible
India. We report two patients, one with liver cirrhosis and another with extrahepatic portal vein obstruction (EHPVO), who developed acute MVT following EST. This complication has not been reported earlier in EHPVO.

Case 1: A 17-year-old boy with post necrotic liver cirrhosis with portal hypertension was admitted with bleed 10 weeks back from esophageal varices. He was managed by injection sclerotherapy with 1% polidocanol as sclerosant. On admission, the patient was pale and mentation was normal. Abdominal examination revealed mild splenomegaly and presence of free fluid. Investigations: hemoglobin 2.7 mg/dL, serum bilirubin 1.9 mg/dL and prothrombin time four seconds beyond control. Ultrasonography had shown liver cirrhosis, dilated portal vein (15 mm) and patent mesenterico-spleno-portal axis.

The patient was resuscitated with intravenous fluids, packed cells and fresh frozen plasma. Octreotide infusion (25 μg/hour) was started. Esophago-gastro-duodenoscopy (EOD scope) showed two columns of grade III esophageal varices with red color signs; stomach and duodenum were normal. EST was performed with absolute alcohol (2 mL in each column) with a combination of intra and paravasical technique. The patient tolerated the procedure well and did not bleed subsequently. Two days following the procedure he complained of continuous dull pain in the abdomen. On examination the abdomen was soft with no tenderness, and bowel sounds were sluggish. The pain increased in severity with abdominal distension, and frank signs of peritonitis were evident by the next day. X-ray showed dilated bowel loops with few air-fluid levels. Abdominal paracentesis yielded dark serosanguinous fluid.

Exploration revealed segmental MVT with bowel gangrene. A one-meter long segment of jejunum 15 cm from the duodenaljejunal flexure was gangrenous, with thrombus in the mesenteric veins. The superior mesenteric artery and its branches were patent and arterial pulsations well felt. The gangrenous bowel and adjacent congested segment was resected and end-to-end anastomosis performed. In view of liver cirrhosis with deranged coagulation parameters, systemic heparinization was not initiated. Postoperative period was uneventful. He is well at a follow up five months later.

Case 2: A 25-year-old lady with EHPVO was admitted with history of melena since six days. She had undergone central splenorenal shunt eight years back for the management of esophageal variceal bleed and was asymptomatic in the intervening period. On admission, she was pale and sensorium was normal. Abdominal examination revealed an upper midline scar of surgery. Investigations: hemoglobin 5.7 g/dL, platelet count 4.3 X 10^9/mm³. Other hematological indices, coagulation profile, and liver and renal biochemical tests were normal. Ultrasonography showed thrombus in the extrahepatic portal vein extending into the right and left branches and proximally up to the splenorenal confluence with cavernoma formation. Liver echotexture was normal.

The patient was resuscitated with fluids and three units of blood transfusion. Emergency EOD scope showed four columns of grade III esophageal varices with red color signs; stomach and duodenum were normal. EST was performed with absolute alcohol (2 mL in each column) with a combination of intra and paravasical technique. The patient developed severe pain in the abdomen and distension on the second day. There was frank evidence of peritonitis: severe abdominal tenderness with guarding and absent bowel sounds. X-ray showed dilated bowel loops with few air-fluid levels. At laparotomy, a one-meter-long segment of proximal jejunum was gangrenous. There was thrombosis in the mesenteric veins though the arteries were patent. The infected bowel was resected and end-to-end anastomosis was done. The patient was started on heparin 5000 units i.v. six hourly postoperatively. She developed anastomotic leak that responded to conservative management. She is well at follow up three months later.

While the exact pathogenesis is not known, it is postulated that altered venous flow, endothelial damage and hypercoagulable state following EST may promote local venous thrombosis and its propagation into the splenic venous system. Local inflammatory response after sclerotherapy may be an initiating event. Retrograde flow through collateral pathways and/or abnormal responses of perivenous lymphatic vessels to the sclerosant may also account for these changes.1 Distant histologic effects due to sclerosant, including intimal damage, medial destruction and increased fibrosis and microthrombi in the portal and splenic veins have been reported earlier.34 In two prospective studies where the portal venous system was assessed by ultrasound as well as angiography, no evidence of thrombosis or alteration in the caliber of the portal venous system was noted.1

The alternative explanation proposed is that a dilute sclerosant may induce a hypercoagulable state. This has been shown with dilute sodium tetradecyl sulfate, which causes selective inhibition of protein C and promotes platelet aggregation.2 Vasopressin infusion, which causes splanchic vasoconstriction, may aggravate the hypercoagulable state and promote thrombosis in the splenic bed, as can be inferred by the number of patients with vasopressin treatment from amongst the cases with acute MVT reported till date.13 One of our patients was on octreotide infusion for control of variceal bleed; a mechanism similar to vasopressin might have been operational in the thrombogenesis. EHPVO in the second patient possibly suggests an inherent predisposition to venous thrombosis. Raised platelet count (4.9 x 10^9/mm³) following previous splenectomy for shunt surgery was another contributing factor.

References
Endoscopic management of anal protrusion of ventriculo-peritoneal shunt

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A 2-year-old male child, who was operated on 18 months earlier for tuberculous meningitis with hydrocephalus by placement of a ventriculo-peritoneal shunt, presented with the lower end of the shunt tube coming out through the anus. Colonoscopy showed the shunt tube coming out through the colon 22 cm from the anal opening. The cranial end, along with a malfunctioning valve, were disconnected surgically, and the shunt was removed endoscopically using a pediatric flexible colonoscope. [Indian J Gastroenterol 2003;22:29-30]

Key words: Bowel perforation, hydrocephalus

Abdominal complications of ventriculo-peritoneal shunts are seen in 5%-47% of cases.1 The main complication is peritonitis without perforation; other complications include transient intestinal ileus, CSF ascites, pseudocyst formation and perforation of various viscera. Perforation of the bowel is very rare, occurring in less than 0.1% of cases.2

A 2-year-old boy presented with a glistening white thin tube coming out through the anus. He had been operated on for post-tuberculous meningitis hydrocephalus at the age of 5 months; a standard ventriculo-peritoneal shunt had been placed. Four revision surgeries were done subsequently, wherein the malfunctioning ventriculo-peritoneal shunts were removed and new shunts were placed. Each time, the peritoneal ends were positioned on either side of the abdomen through a midline incision. There were no complaints of fever, diarrhea, convulsions, vomiting or hydrocephalus. On examination, his fontanelles were lax and there were no signs of raised intracranial pressure. The head circumference was 48 cm. The abdomen was soft and non-tender. The shunt retracted the next day.

The cranial end, along with a malfunctioning valve, were first disconnected surgically. The peritoneal end was localized by means of flexible pediatric colonoscope to a site 22 cm from the anal opening (Fig). With the help of a snare, the protruded shunt was extracted under direct vision; it was not obstructed.

Fig: Perforation of ventriculo-peritoneal shunt tube through colonic wall. The tube is seen in the lumen

The site of perforation in the colonic wall was not visible because of fecal matter. There was no evidence of peritonitis post-procedure; there was normal passage of feces in the postoperative period. Intravenous antibiotics were administered.

Subsequently a ventricular chamber was inserted, which was aspirated at regular intervals, thus reducing the intracranial pressure. When CSF studies following chamber aspiration revealed sterile CSF, a new ventriculo-peritoneal shunt was placed. The child was discharged asymptomatic.

The pathogenesis of intestinal perforation by ventriculo-peritoneal shunts is unclear and various mechanisms have been suggested:3 foreign body reaction; pressure necrosis; relatively sharp and stiff end of shunt tube causing perforation; poor general condition of the patient with weakening of the intestinal wall.

Management depends on the nature of the complication. In our case, there were no abdominal signs or symptoms. Therefore, disconnection of the cranial end followed by simple removal of the catheter endoscopically was done. Only two cases of endoscopic removal of the peritoneal end of such perforating shunt tubes have been reported so far.4,5

Thus, if bowel perforation in patients with ventriculo-peritoneal shunts is recognized before it produces serious complications, it can be treated using the simple procedure we described.

References