maldevelopment of the dorsal mesogastrium or acquired ligamentous laxity due to multiparity in women.1,2 Clinically it presents as a fairly painful mass with subacute abdominal or gastrointestinal complaints, or as a mobile abdominal mass diagnosed incidentally or with signs of "acute abdomen" due to splenic torsion and infarction.1,3 Imaging studies assist in the preoperative diagnosis.1,2

Splenectomy is recommended for acute torsion of spleen,1,3 for congestive splanomegaly and secondary hypersplenism,1 and even in asymptomatic adults.4 Splenectomy is recommended in the very young, to avoid post splenectomy sepsis.1

Kalikankan Dutta, Manoranjan Kar, Rabin Mandal
Department of Surgery, Calcutta National Medical College and Hospital, Kolkata 700 014

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Correspondence to: Dr Kar, Assistant Professor, Flr 3B, 1/34A, Khudiram Bose Sarani, Kolkata 700 037

Malignant fibrous histiocytoma of mesocolon

Malignant fibrous histiocytoma of the mesentery is very rare; so far only two cases have been documented in literature.

A 50-year-old man presented with a lump in the upper abdomen since 3 months. On examination, a vertically oval mass was present in the upper abdomen intraperitoneally. Systemic examination was otherwise normal. At laparotomy, a lobulated mass 25 cm x 20 cm in size was removed from the transverse mesocolon. Histology was suggestive of malignant fibrous histiocytoma of the mesentery. The postoperative period was uneventful. The patient received chemotherapy.

Malignant fibrous histiocytoma is the most frequent sarcoma in adults and can involve the mesentery rarely. The diagnosis is often delayed till the disease is an advanced stage, because of the non-specific nature of the symptoms.1 Malignant fibrous histiocytoma of the small bowel mesentery2 and of the sigmoid mesocolon were reported in literature as successfully resected, with favorable prognosis.3

G Mallikarjuna Rao, M Janaki,*
P Haricharan, P Nagendra Nath
Departments of Surgery and *Pathology, Kurnool Medical College Hospital, Kurnool 518 002 (AP)

Spontaneous enterocutaneous fistula – a rare presentation of Crohn’s disease in India

Crohn’s disease is common in the Western world, but is considered rare in developing countries like India.1,2 In the cases reported from India, presentation with enterocutaneous fistula is very rare.

A 30-year-old man presented with history of lump in the right iliac fossa and pain in the right lower abdomen since 2 months, with no history of fever, cough, diarrhea or GI bleed, and no symptoms suggestive of peritonitis or intestinal obstruction. Ultrasonography 10 days earlier had shown a parietal wall abscess and few dilated bowel loops locally.

Physical examination revealed tachycardia and pallor. Local examination showed an immobile lump, 6 cm x 4 cm, in the right iliac fossa, which was warm, tender, tense and cystic. It was diagnosed as parietal wall abscess and about 100 mL of pus was drained through an incision over 4 days. He was admitted again after 10 days with complaint of fecal discharge from the wound site. He was treated conservatively; the discharge decreased initially but again increased.

After 6 weeks on nutritional support, the patient was taken for exploratory laparotomy, which revealed 50 mL of fecal material in the abdominal cavity. A lump consisting of about 15 cm of ileum, cecum and ascending colon was densely adherent to the parietal wall. The local bowel wall was thickened and mesenteric lymph nodes were enlarged. Omentum was adherent to the lump. The lump was resected, with ileo-transverse colon anastomosis. Postoperative recovery was uneventful. Histology of the resected colon showed transmural chronic inflammatory cell infiltrate with noncaseating granulomas, suggestive of Crohn's disease. The patient is doing well on follow-up.

This case suggests that in patients with spontaneous fecal discharge (enterocutaneous fistula) with lump in right iliac fossa, a differential diagnosis of Crohn's disease should be kept in mind even in India.

D K Jain, V D Upadhya,
Arvind K Shukla, P Lubana
Department of Surgery, M G M Medical College and M Y Hospital, Indore 452 001

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Cruehlhier-Baumgarten syndrome associated with non cirrhotic portal fibrosis

Cruehlhier-Baumgarten (C-B) syndrome is a rare clinical entity characterized by presence of unusually prominent paraumbilical veins with evidence of portal hypertension and venous hum over the paraumbilical vein. It is commonly described in patients with cirrhosis of the liver. We report a patient with non cirrhotic portal fibrosis (NCPF) who had this syndrome.

A 35-year-old woman presented with gradually increasing splenomegaly for 12 years, with history of hematemesis 4 years and 1 month back, which was managed conservatively. There was no history suggestive of jaundice, ascites or encephalopathy during this period. On examination the patient was anemic, with no signs of chronic liver disease. Large dilated veins were seen in the paraumbilical area, forming a bunch of vessels. A continuous venous hum was audible over it. Spleen was enlarged up to the umbilicus (12 cm below costal margin) and was firm in consistency.

Investigations: hemoglobin 9.8 g/dL, WBC 8000/mm². Liver profile normal; serological markers for hepatitis B and C virus were negative. Ultrasound examination revealed normal liver and marked splenomegaly. Dilated and patent portal and splenic veins along with patent, large umbilical vein and collaterals around the splenic hilum were seen. Upper GI endoscopy showed three grade III esophageal varices and congestive gastropathy. Liver biopsy revealed normal lobular architecture, with minimal fibrosis of portal tract, sclerosis and obliteration of small-sized portal vein radicals; these features were suggestive of NCPF.

The patient was subjected to endoscopic variceal ligation, and there was no bleeding over the next 12 months.

In C-B syndrome, primary liver disease, usually cirrhosis, and portal hypertension are responsible for extensive dilatation of paraumbilical vein and recanalization of umbilical vein; C-B disease is due to congenital patency of umbilical vein occurring in the absence of liver disease.1,2

Only a few reports,3,4 including the original one by Cruehlhier,5 described prominent paraumbilical vein and splenomegaly with widely patent umbilical vein but without evidence of cirrhosis of the liver. We believe that at least some of these patients may have NCPF, a common cause of portal hypertension in India.

It is unlikely that presence of C-B syndrome alters the management of portal hypertension or primary liver disease; however it increases the prevalence of hepatic encephalopathy and surgical intervention may become difficult due to extensive collaterals.

Rupesh Kumar Pokharna, Sunil Kumar,* Kuldeep Saini, D K Kochhar*
Departments of Gastroenterology and *Medicine, S P Medical College, Bikaner, Rajasthan

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