The postulated mechanisms of thrombosis in this metabolic disorder include endothelial dysfunction, proliferation of vascular smooth muscle cells, lipid peroxidation and oxidation of LDL, which are presumably mediated by reactive oxidant stress. Various causes for hyperhomocysteinemia have been described, with a genetic inborn error of metabolism being often found in those with very high serum levels. The markedly elevated homocysteine level in our patient and his sibling had normalized (12.02 µmol/L; normal <15). Ectopia lentis, skeletal deformities and mental retardation were absent.

He was started on oral folic acid (5 mg/day) to which he had good clinical response, with progressive recovery of the digital gangrene. No further vascular events occurred. Serum homocysteine was normal (8.65 µmol/L) after 6 months' therapy with folic acid. At one-year follow up, he is asymptomatic.

His brother, who was asymptomatic, was also found to have elevated serum homocysteine (88.8 µmol/L). He too was administered folic acid and after 6 months his homocysteine levels also had normalized (12.02 µmol/L).

Hyperhomocysteinemia is a well-recognized cause of macrovascular thrombosis and associated sequelae. The most common site of involvement appears to be the coronary arteries. The cerebrovascular arteries and retinal vasculature are some of the other commonly affected sites. To the best of our knowledge, there has been only one earlier report of superior mesenteric artery thrombosis extending from the proximal jejunum to the mid transverse colon. The proximal 90 cm of jejunum appeared relatively viable. The gangrenous intestine was resected and end-to-end anastomosis was performed.

After CT scan of abdomen, he was taken up for emergency laparotomy. This showed a 2-cm thrombus in the trunk of the superior mesenteric artery, with extensive intestinal gangrene extending from the proximal jejunum to the mid transverse colon. The proximal 90 cm of jejunum appeared relatively viable. The gangrenous intestine was resected and end-to-end anastomosis was performed.

On the second postoperative day, he developed painless, blackish discoloration of the hallux and second toe of the right foot, and was started on low-molecular-weight heparin. By the seventh postoperative day he developed a similar superficial dry gangrene of the left hallux. He was evaluated for vasculitis and hypercoagulable states (ANA, RA factor, anticardiolipin antibodies, protein S, protein C, and homocysteine). All of these were normal, except serum homocysteine which was grossly elevated (178.30 µmol/L; normal <15). Ectopia lentis, skeletal deformities and mental retardation were absent.

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tating segment IV of the liver. No significant lymphadenopathy was present. An extended cholecystectomy with 2-cm wedge resection of the liver beyond the infiltration and hepatoduodenal lymphadenectomy was performed.

The resected specimen had an irregular surface, with a firm grey-white nodule in the gall bladder fossa. The nodule was around 4 cm x 3.5 cm in diameter, near the gall bladder wall but not involving it. On histologic examination, the tumor was composed of fascicles of plump spindle cells intermixed with numerous plasma cells and variable number of lymphocytes and histiocytes (Fig). The spindle cells were arranged in fascicles with a moderate amount of intercellular collagen demonstrable with Masson trichrome stain. There were no abnormal mitoses. The veins entrapped within the lesion showed features of endophlebitis. The adjacent liver parenchyma showed moderate portal fibrosis with mild chronic inflammation in the portal tract. The bile ducts showed concentric fibrosis. Immunohistochemistry revealed strong positivity for smooth muscle antigen (SMA) in the spindle cells, indicating their myofibroblastic origin. CD68 immunostaining revealed occasional presence of histiocytic cells. Immunostaining for Epstein-Barr virus was negative. The postoperative period was uneventful and the patient is doing well 6 months later.

Inflammatory pseudotumors of the liver are relatively rare, but more than 200 cases have been reported in the literature. Because of the many reports of spontaneous regression of hepatic pseudotumors, most patients are treated with simple observation or conservative therapy. In contrast to peripheral hepatic pseudotumors, those involving the porta hepatis require treatment for obstructive jaundice. Fourteen cases have been reported at the hepatic hilum till 2001,5 apart from a recently reported case.6 None of the cases occurred at the gall bladder fossa.

The exact etiology of this lesion is unknown, but it is generally regarded as a benign reactive inflammatory condition. Evidence of Epstein-Barr virus infection has recently been documented.1 The presence of obliterating phlebitis has been reported,3 as in the present case. It was hypothesized that micro-organisms gaining access to the hepatic parenchyma through the portal vein subsequently elicit an inflammatory reaction that results in obliterating phlebitis.7 Portal hypertension has been reported to be an associated feature.7

The prognosis of inflammatory pseudotumor of the liver is generally considered good. Most of the patients recover after resection.

References

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Severe esophagitis in a child with Henoch-Schönlein purpura presenting as protein-losing enteropathy

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A 4-year-old boy was seen for vomiting, diarrhea and peripheral edema. He had no evidence of nephrosis or liver dysfunction. Upper gastrointestinal endoscopy was performed for investigating the etiology of protein-losing enteropathy. It showed severe esophagitis and multiple ulcers in the descending duodenum. The symptoms and endoscopic mucosal abnormalities subsided after three weeks of hospitalization without specific therapy. Ten days after being discharged he was seen again with characteristic rash of Henoch-Schönlein purpura and arthritis without gastrointestinal symptoms. Biopsy of the skin rash revealed leukocytoclastic vasculitis. [Indian J Gastroenterol