Case Snippets

Conservative management of giant hepatic mesenchymal hamartoma

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We report a six-month-old male infant with mesenchymal hamartoma of the liver which was left untreated. Repeat CT scan 1 year later showed regression and calcification of the tumor. The child is well 3 years later. [Indian J Gastroenterol 2004;23:26]

Key words: Liver tumor

Mesenchymal hamartoma is an uncommon benign liver tumor of childhood.1-5 Though the standard treatment is surgery, a few reports suggest that expectant management is likely to result in spontaneous regression of this tumor.1,2,5

A six-month-old male infant was brought with history of abdominal fullness and frequent regurgitation of feeds. On examination, the baby was pale. He had a large mass occupying the right and left hypochondrium and epigastric regions.

Investigations: Hemoglobin 4.8 g/dL. Serum bilirubin was 1.4 mg/dL (conjugated 0.7); serum proteins and transaminases were normal. Serum alkaline phosphatase was 24 KA units (normal 3-13). Ultrasonography and CT scan suggested a large (9 cm x 10 cm) space-occupying lesion in the liver (Fig 1a). After correction of anemia, laparotomy was done. The tumor was not excised as it occupied most segments of the liver; liver biopsy was done. Histology revealed a tumor with loose, vascular connective tissue, interspersed with strands of epithelial cells, focally cuboidal or flattened. Immunohistochemistry revealed strong smooth muscle actin positivity whereas factor VIII-related antigen was negative, favoring a diagnosis of mesenchymal hamartoma.

The child was managed expectantly. Repeat CT one year later showed marked regression of the lesion (3.5 cm x 4 cm) and calcification (Fig 1b). The child is well 3 years later; he has no clinical hepatomegaly.

The usual presentation of mesenchymal hamartoma is as an asymptomatic mass, though occasionally decreased appetite, respiratory distress, high-output cardiac failure, and death due to bleeding into the tumor have been reported.2,3 Serum alpha-fetoprotein may be elevated. Since most of the tumors were resected at the time of discovery, the natural history of these tumors remained unknown. Some that were missed in childhood are being picked up incidentally in adults on CT scan. Pathologically, mesenchymal hamartomas are considered tumor-like lesions, and studies suggest that the changes may be due to anomalous blood supply leading to ischemic and cystic changes in the liver.4

Several methods of surgical removal of these tumors have been described. There are a few deaths reported with aggressive surgery. No tumor recurrences have been reported with any treatment modality. Recently, recognizing the benign nature of these tumors, a more conservative surgical approach has been advised.1,2,5 Expectant management was reported first by Leary and Barnhart.5 They concluded that asymptomatic lesions can regress spontaneously and interval expansion does not preclude subsequent regression. However, they advised open biopsy before an expectant line of management is undertaken.

We feel that a majority of these tumors may be managed conservatively after confirming the diagnosis. Surgery may be reserved for life-threatening situations, like cardiac failure or uncontrolled bleeding.

References

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Transcatheter hepatic artery embolization for spontaneous rupture of amyloid liver

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Spontaneous rupture of amyloid liver is a fatal complication. A 48-year-old man with systemic amyloid-
sis secondary to multiple myeloma presented with acute hemoperitoneum. Emergency angiogram showed extravasation of contrast from the liver into the subhepatic space, which was successfully stopped by embo-

lization of the right hepatic artery. [Indian J Gastroenterol 2004;23:26-27]

Key words: Transcatheter embolization

Spontaneous rupture of liver in patients with systemic amyloidosis is rare. Survival after such a rupture is even rarer, with only four of ten reported cases in literature surviving the event.1,4 We report a patient with amyloid liver whose bleeding could be controlled with transcatheter hepatic artery embolization.

A 48-year-old man presented with pedal edema, facial puffiness and abdominal distension, and progressive loss of appetite and weight since two years. He did not give history of diarrhea or urinary abnormalities. On examination, he was pale, with bilateral pitting pedal edema and facial puffiness. Abdominal examination revealed hepatomegaly 6 cm below the costal margin, and free fluid. The spleen was not palpable. The rest of the clinical examination was unremarkable.

Investigations: hemoglobin 11.8 g/dL, platelets 439,000/ cumm, prothrombin time 14.4 s/11.3 s, serum protein 5.3 g/dL, albumin 2.5 g/dL, ALT 26 U/L, AST 32 U/L, alkaline phosphatase 2019 U/L, and gamma GT 457 U/L. Twenty-four-hour urinary protein was 2.5 g. Ultrasonography showed hepatomegaly with altered echotexture and mild splenomegaly. There was no evidence of portal hypertension. There was free fluid in the abdom-

enal and the kidneys were of normal size. Ascitic fluid was low in protein.

In view of the hepatosplenomegaly, nephritic-range proteinuria, and infiltrative picture on liver biochemistry, the possi-

bility of systemic amyloidosis was considered. Duodenal bi-

opsy was consistent with the diagnosis. Bone marrow aspiration cytology showed plasmacytosis consistent with a diagno-

sis of myeloma. Serum electrophoresis showed the classical paraprotein M band. Skull and pelvic X-rays were normal.

During his hospital stay, he suddenly collapsed in the bath. On reinstating him in the bed he was found to be in shock. An appreciable increase in abdominal girth was noted; ascitic tap from two sites yielded bloody fluid. The ascitic fluid PCV was 9%, with concomitant blood PCV having dropped to 13%. Hemodynamic resuscitation was done. On revival, the patient complained of intense abdominal discomfort. The possibility of hepatic rupture was considered; emergency CT showed an area of hepatic injury with perihpatic hematomata.

Emergency abdominal angiography was done through the right femoral approach. Flush aortogram and hepatic angiogram showed extravasation of contrast from the inferior aspect of the liver into the peritoneal cavity. The hepatic artery was catheterized with a 4 F Cobra catheter (Tempo: Cordis). As no single branch of the hepatic artery was seen leading to the site of extravasation, two pushable fibrous platinum coils 5 x 30 (Boston Scientific) were placed in the right hepatic artery. Post 

embolization angiography showed no extravasation. Splenic, renal and the other aortic branches were normal. The patient was subsequently monitored in the ICU and his hemoglobin remained stable. At discharge, he was hemodynamically stable and asymptomatic. Chemotherapy with prednisolone and melphalan was planned.

Spontaneous rupture of liver is an ominous complica-

tion of amyloid liver. Various causes for this predis-

position are massive hepatomegaly, rigidity of the hep-

atic parenchyma, and vascular fragility due to vascular 

involvement.2 Factor X deficiency has also been docu-

mented in amyloidosis and is a possible cause for bleed-

ing.4 This bleeding tendency is also manifested in the skin as easy bruisability, spontaneous ecchymoses and pinch purpura. Hemorrhage in hepatic amyloidosis is also known to occur after liver biopsy.

Treatment options tried have been mainly surgical, 

where either hemostasis or resection of affected seg-

ments has been attempted.2,4 Surgical hepatic artery 

ligation was tried in one patient with no success. Sur-

gical attempts are often offset by underlying dissemi-

nated intravascular coagulopathy, which can aggravate the bleeding. Emergency liver transplantation has been done in one case of spontaneous rupture.3

We believe this is the first report of control of bleeding in amyloid liver by transcatheter embolization.

References

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Pancreatic pseudocyst presenting as
odynphagia

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A 40-year-old man presented with abdominal pain and
dynphagia. CT scan revealed a pseudocyst in the posteri-

or mediastinum and pleural effusion complicating pancreatitis. He was managed with translumbar pancreatic duct stenting along with other supportive