Hepatic adenomatosis – a rare double complication of multiple adenoma rupture and malignant transformation

Nitin Arvind, Duraimurugan D, J S Rajkumar
Department of Surgical Gastroenterology, Lifeline Rigid Hospitals, Kilpauk, Chennai

Hepatic adenomatosis (HA) is a rare disorder that is susceptible to hemorrhagic complications and, rarely, to malignant transformation. We report a 24-year-old woman who was found to have HA with >10 tumors; she presented with simultaneous rupture of two of the adenomas, along with malignant change in one. Hematoma evacuation and caudate lobe resection were done, and she has been doing well 6 months later. [Indian J Gastroenterol 2006;25:209-210]

Hepatic adenomatosis is a condition with presence of multiple adenomas in the liver, variously defined as more than 4 adenomas or more than 10 adenomas. In contrast to solitary adenomas, adenomatosis shows no association with the use of oral contraceptives, anabolic steroids and type I glycogen storage disease. Increase in size and propensity to rupture during pregnancy is also not known with hepatic adenomatosis. The potential for spontaneous bleeding and malignant transformation are, however, common to both.

A 24-year-old woman was referred with history of spontaneous, sudden-onset abdominal pain of 12 days' duration. Her abdomen was soft, with palpable liver and epigastric and right hypochondrial tenderness. Ultrasonogram showed fluid in the pouch of Douglas. Culdocentesis showed hemorrhagic fluid. Coupled with a history of delayed menstrual cycle, a provisional diagnosis of ruptured ectopic gestation was made. At laparotomy, approximately 900 mL of altered blood was found in the peritoneal cavity and the fallopian tubes were found to be normal. The surface of the liver was irregular and nodular and a large hematoma was seen over the posterior superior aspect of the right lobe. The clots in the peritoneal cavity were evacuated, a wash given, and the abdomen closed. She was then referred to us for management.

The patient gave no history of recent trauma or usage of oral contraceptives. She had been operated on for atrial septal defect 10 years previously and had had a caesarean section 2 years back. On examination, she had an enlarged, tender liver. Her vital signs were stable; red and white blood cell counts were normal and liver function tests were normal, except for raised alkaline phosphatase (250 IU/L; normal: 30-180) and gamma glutamyl transpeptidase (276 IU/L; normal: 5-80).

MRI showed a 14 cm × 12 cm lesion involving the posterior superior aspect of the right lobe of the liver, minimally hyperintense in T2-weighted images and hypointense in delayed images with inhomogenous early enhancement. There was evidence of fat within the lesion along with multiple areas of hemorrhage. There was another rounded lesion with similar signals in the caudate lobe with surrounding hemorrhage. About 8 smaller lesions with similar signals were seen in the right lobe, the largest measuring 4 cm. Another 5-cm lesion was seen in segment II of the left lobe.

Soon after admission, the patient again developed signs of peritonism. At laparotomy, the liver was found to be studded with rounded, nodular swellings. A large subcapsular hematoma was seen over the region of segment V with a rent in the capsule over it, and another subcapsular hematoma over the caudate lobe. The hematoma was evacuated and non-anatomical resection of the caudate lobe adenoma along with surrounding liver parenchyma was done along with segmentectomy V (Fig). The patient responded well to surgery. On follow up she has not shown complications or increase in size of the remaining adenomas.

Histology suggested changes consistent with adenoma in the caudate lobe specimen. The larger specimen showed moderately differentiated hepatocellular carcinoma (grade II/III of Edmondson and Steiner) whereas the smaller adenoma that had ruptured showed no such change.

The conditions that predispose to hepatic adenomatosis are poorly understood. One speculation has to do with congenital or acquired abnormalities...
of the hepatic vasculature.3 Adenomas are hypervascular tumors containing multiple sinusoids of capillaries with thin walls that are perfused exclusively by the high-pressure hepatic arterial flow. Due to poor connective tissue support, there is a predisposition to bleeding. Malignant change, although rare, is also known to occur.

They are usually asymptomatic and are diagnosed incidentally, but occasionally present with abdominal pain, hepatomegaly or altered liver function tests. They can present with catastrophic intra-peritoneal bleed. Ultrasonography, CECT or MRI can be utilized in the diagnosis, with MRI proving to be the best bet. On unenhanced CT, they are hypodense, but lesions with hemorrhage appear hyperdense or heterogeneous. These lesions show significant enhancement on arterial phase images as they are supplied by the hepatic artery. The presence of a capsule and demonstration of intralesional fat on out-of-phase T1-weighted images are helpful in diagnosis on MRI. Fine needle biopsy has the dual drawback of precipitating bleeding, and seeding of the needle track if the mass turns out to be malignant.

Hepatic resection is indicated in the presence of a major complication such as intraperitoneal bleed, but its usefulness in the uncomplicated patient has been debated. The consensus is that larger and more vulnerable tumors (>5 cm, subcapsular, exophytic and hemorrhagic) need to be resected,4 while smaller asymptomatic tumors can be monitored. Orthotopic liver transplantation may be used in patients who have progressive symptoms after partial resection, or in whom carcinoma is suspected.3

References


Correspondence to: Dr. Arvind, E47/2, 3rd Street, Annanagar East, Chennai 600 102. E-mail: drnitinarvind@gmail.com

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