Lelomyosarcoma of Inferior vein cava presenting as acute Budd-Chiari syndrome

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We report a 49-year-old lady who presented with acute Budd-Chiari syndrome. Spiral CT scan showed inferior vein cava (IVC) tumor and ischemia of the right liver secondary to hepatic vein blockage. These were confirmed by MRI scan and IVC gram, at which time tissue diagnosis was obtained. At surgery, the tumor was seen to originate from the infrahepatic IVC and extended to the level of the diaphragm, blocking the hepatic vein outflow. The tumor was excised completely. Histology confirmed it to be lelomyosarcoma of the IVC. The patient is well, without recurrence of symptoms or tumor, 10 months later. [Indian J Gastroenterol 2001;20:33-34]

Key words: Hepatic venous outflow obstruction

Lelomyosarcoma is a rare tumor of the inferior vena cava (IVC) and can rarely cause Budd-Chiari syndrome. We report a patient with this tumor who presented with features of acute Budd-Chiari syndrome, and was treated successfully by resection of the tumor.

A 49-year-old woman presented with acute pain in the right hypochondrium radiating to the back, with difficulty in breathing and generalized distension of the abdomen of 10 days’ duration. On inquiry, she gave history of intermittent abdominal distension since 3-4 years, relieved with homoeopathic treatment. She also had history of edema of the feet on prolonged standing since 15 years. She was diagnosed to have deep vein thrombosis and varicose veins 15 years ago following child delivery. She was a diabetic on diet control.

Examination revealed mild tachypnea, no edema of the feet, and bilateral varicosities of the long saphenous veins. There was evidence of ascites and 4 cm nontender, firm hepatomegaly. Respiratory system was normal.

Investigations: normal hemogram and renal function tests. Liver profile showed elevated levels of AST (128 IU/L), ALT (471 IU/L) and alkaline phosphate (347 IU/L); serum bilirubin was normal. Ultrasonography with color Doppler showed evidence of thrombus in the IVC from the level of the renal vein up to the diaphragm, with patent hepatic veins and portal vein: ascites was present. A provisional diagnosis of acute IVC thrombosis leading to pulmonary embolism was made. However, lung perfusion scan did not reveal evidence of pulmonary embolism. Spiral CT angiography showed a heterogeneous, enhancing, soft-tissue mass arising from the infrahepatic IVC and extending up to the suprarehepatic IVC, suggestive of a tumor in the IVC. This was blocking the opening of all the hepatic veins. The right liver lobe had a mottled appearance suggestive of hypoperfusion. There was ascites and bilateral pleural effusion. MRI scan with gadolinium contrast confirmed the find-
ings and also showed that the right atrium was free of tumor. It also revealed a non-tumor thrombus in the right hepatic vein. IVC graft showed complete blockage of the infrahepatic IVC and multiple retroperitoneal collaterals. Biopsy from the tumor during the IVC graft was reported as showing atypical cellular leiomyoma. There was no evidence of metastasis and ascitic fluid was negative for malignant cells.

At laparotomy, the liver was congested. There was a tumor arising from the posterior wall of the IVC at the level of the renal veins and extending like a pedunculated intraluminal mass up to the level of the diaphragm. The area of origin of the tumor was 8 cm long and 1.5 cm wide. The supradiaphragmatic IVC and renal vein openings were free of tumor. Vascular control of the IVC below the level of the renal veins and above the diaphragm (transpericardial) was obtained. Right and left renal veins were clamped. The anterior wall of the IVC was incised, tumor was excised, and the anterior and posterior walls were primarily closed. During excision, vascular inflow to the liver was occluded for 25 min. The excised tumor was 17 cm long, 5 cm wide, grayish white in color, with smooth surface and firm consistency (Fig). The tumor weighed 80 grams. Cut surface showed whorled gray-white appearance with small foci of congestion. Microscopic examination showed interlacing fascicles of oval to spindle cells, blunt plump spindle nuclei, moderate hyperchromasia, brisk mitotic activity (3-5/hp) and scattered bizarre giant nuclear forms. It was reported as high-grade leiomyosarcoma. Biopsy from both lobes of liver revealed centrilobular necrosis without evidence of cirrhosis.

Postoperative recovery was uneventful. Color Doppler studies showed patent hepatic veins and retrohepatic IVC. However, there was a thrombus in the infrarenal IVC at the site of sutures; the rest of the IVC was patent. The patient was put on low-molecular-weight heparin from the third postoperative day and was later switched to oral anticoagulation. Follow-up color Doppler study after 3 months showed patent IVC and hepatic veins without residual tumor.

Leiomyosarcoma of the IVC is a rare malignant tumor originating in the smooth muscle of the media. More than 50% of vascular leiomyosarcomas arise from the IVC. Most patients are women (sex ratio 6:1) in their sixth decade. Most tumors involve the middle or upper segment of the IVC, while a small number involve the entire IVC. Invasion by the tumor of the hepatic veins can lead to acute or chronic forms of the Budd-Chiari syndrome. Rarely, this can lead to death from acute hepatic failure. The present case presented with features of acute Budd-Chiari syndrome.

On sonography, a lobulated tumor in the IVC with dilatation of the IVC suggest a leiomyosarcoma. CT scan is more definitive in identifying intravascular leiomyoma. The typical appearance is dilatation of the IVC by an intermediate-attenuation mass that shows irregular enhancement and total or almost complete obstruction of the IVC. Intravascular tumors are differentiated from thrombus by presence of IVC dilatation in the former. MRI with contrast has been found to define accurately the upper extent of malignancies invading or growing in the vena cava. Vena cavaography delineates the extent of the tumor, its relationship with renal and hepatic veins, and the degree of collateral circulation. During the procedure, biopsy can be taken from the tumor.

Surgical resection is recommended as the treatment of choice, even in patients with metastasis; although it may not cure the patient, it offers effective palliation. When partial resection of the IVC is performed, direct suture is sometimes feasible. If not, patch angioplasty using a polytetrafluoroethylene graft is done. When adequate venous collateral pathways have developed, the IVC can be ligated without resulting in significant edema of the lower extremities. Concomitant partial or complete vena caval resection and replacement may be necessary. In most case, however, the mass can be removed from within the lumen by blunt dissection.

The tumors are poorly radiosensitive and the proximity of digestive structures limits the amount of radiation that can be delivered safely. Radiotherapy can be used in the management of metastases or in combination with chemotherapy to make surgery possible. Chemotherapy is recommended in patients with or without metastasis. Distant metastases are present at the time of diagnosis in approximately 40% of cases.

The main prognostic factor is the location of the tumor, particularly its highest level of extension; upper-segment tumors have the poorest prognosis. Long-term prognosis is poor. Most patients die of recurrence with invasion of neighboring viscera or distant metastasis.

References
Mesothelial cyst at porta hepatis

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Mesothelial cysts are fluid-filled sacs lined by mesothelial cells. They are rare lesions that have been known to occur at various sites, but have not been reported at the porta hepatis. We report a 45-year-old woman with mesothelial cyst at the porta hepatis that was detected incidentally during open cholecystectomy. [Indiana J Gastroenterol/2001;20:35-36]

Key words: Benign cyst

A variety of benign lesions have been documented to compress or obstruct the extrahepatic biliary tree.1 Mesothelial cysts have been known to occur at various intra-abdominal sites but have not so far been reported at the porta hepatis.

A 45-year-old woman was admitted with complaints of pain in the right upper quadrant of the abdomen for three years. Abdominal examination was unremarkable. Liver profile revealed normal serum bilirubin, AST and ALT levels. Alkaline phosphatase level was markedly raised (845 U/L, normal up to 305 in adults). Ultrasonography detected multiple gall bladder calculi and normal wall thickness. The common bile duct and intrahepatic biliary radii were not dilated.

In view of the raised alkaline phosphatase level, we planned open cholecystectomy and peroperative cholangiogram. At exploration, the gall bladder contained multiple calculi. There was a tensely cystic, white swelling, measuring 3 cm x 3 cm x 2 cm, at the porta hepatis. The swelling was displacing the hepatic artery to the right and causing extrinsic compression of the confluence of the hepatic ducts.

Peroperative cholangiogram through the cystic duct failed to clearly delineate the left hepatic duct and the confluence. No filling defects suggestive of calculi were seen in the biliary tree. The cyst was aspirated and yielded turbid white fluid. It was then carefully dissected out and excised. No feeder ves-

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