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Case Report

Leiomyosarcoma of the Inferior Vena Cava with Cardiac Extension; A Rare Cause of Acute Liver Failure

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ABSTRACT

Leiomyosarcoma of vascular origin is a rare malignant tumor. Inferior vena cava (IVC) is the most common site of vascular leiomyosarcomas. Leiomyosarcoma of IVC is predominantly seen in middle-aged women presenting with non-specific symptoms and clinical findings. Herein, we report a case of leiomyosarcoma of IVC in a 44-year-old woman presented with progressive abdominal discomfort and postprandial vomiting. Laboratory results were in favor of acute liver injury. Imagings revealed a retroperitoneal mass and enhancement in almost the total course of inferior vena cava with extension into both renal veins and right atrium. Histopathological examination showed a malignant spindle cell tumor and immunohistochemistry confirmed smooth muscle origin of the tumor.

Keywords: Leiomyosarcoma, Inferior vena cava, Acute liver failure

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INTRODUCTION

Leiomyosarcoma (LMS) of inferior vena cava (IVC) is a rare tumor originating from smooth muscle cells of caval wall (1). Primary LMS of vascular origin accounts for 2% of all LMSs, with the most common site being IVC (2). Most of these tumors are seen in women in their 6th decade of life. The most common presenting symptoms are abdominal discomfort or pain due to an abdominal mass. About a third of patients with dominant intraluminal involvement might present with swelling of the legs due to caval obstruction (3).

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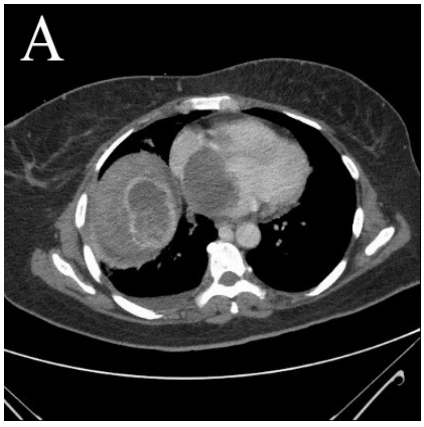
CASE REPORT

The patient was a 44-year-old woman who presented with epigastric discomfort. She was hypertensive for 2 years and on antihypertensive drugs and otherwise healthy. She complained of non-specific abdominal pain in the past two years before the diagnosis. The pain was constant and mostly localized to the epigastrium. She reported an increase in the frequency and severity of pain during the previous 2 months. Furthermore, she occasionally had recurrent and postprandial vomiting. In the physical examination, the abdomen was fatty and distended with mild tenderness in deep palpation.

In laboratory studies, the patient showed an acute increase in liver enzymes, elevated bilirubin, and prolongation in prothrombin time and INR (International



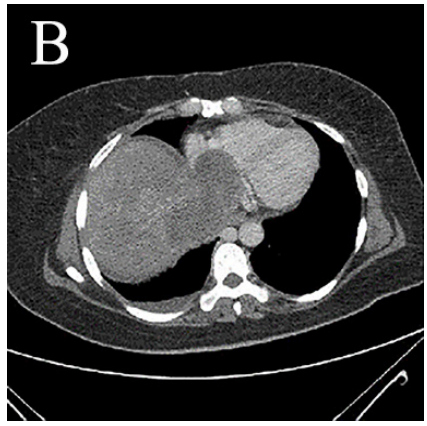


Fig.1, A and B: Contrast-enhanced tomogram of the abdomen and pelvis. A heterogeneously enhancing mass lesion with peripheral irregular enhancement and central necrosis is seen in the most superior part of the right lobe of the liver in favor of metastases.

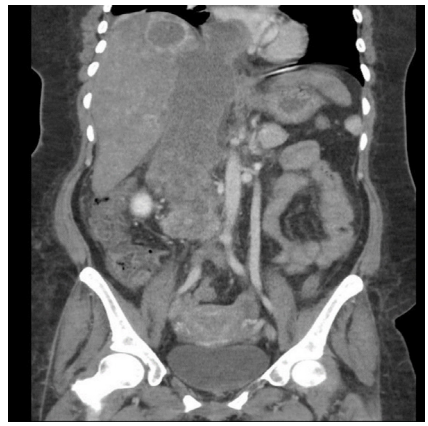


Fig.2: Contrast-enhanced tomogram of the abdomen and pelvis revealed a large retroperitoneal mass lesion with heterogeneous enhancement in almost the total course of inferior vena cava with extension into both renal veins and right atrium.

Table 1: Initial laboratory findings of the patient

Test items	First test	Following test
AST (U/L)	42	200
ALT (U/L)	674	457
ALP (U/L)	244	268
Total bilirubin (mg/dl)	2.3	1.7
Direct bilirubin (mg/dl)	1.2	0.7
Albumin (g/dl)	3.2	2.9
PT (seconds)	17.7	21
INR	1.72	2.1
PTT (seconds)	37.7	45

AST: aspartate aminotransferase, ALT: alanine aminotransferase, ALP: alkaline phosphatase, PT: prothrombin time, INR: international normalized ratio, PTT: partial thromboplastin time.

Normalized Ratio) as depicted in table 1.

Abdominal and pelvic ultrasonography revealed a 140 × 70 × 50 mm mass with mixed echotexture in the epigastric area. Spiral computed tomography (CT) of the abdomen and pelvis with intravenous (IV) contrast was done, which revealed a large retroperitoneal mass lesion with heterogeneous enhancement in almost the total course of inferior vena cava with extension into both renal veins and right atrium (figures 1, and 2). IVC was dilated and enhancing vessels were detected within the mass. Azygos vein was dilated and multiple varicose veins were seen in the pelvic cavity and subcutaneous tissue of anterior abdominal wall secondary to IVC obliteration. A heterogeneously enhancing mass lesion with peripheral irregular enhancement and central necrosis was seen in the most superior part of the right lobe of the liver measuring about 6 × 4 cm in favor of metastases. Inhomogeneous mottled enhancement of liver parenchyma was seen, which was suggestive of Budd-Chiari syndrome secondary to the obliteration of intrahepatic portion of IVC.

Transesophageal echocardiography was performed exhibiting a large mass filled approximately 90% of the right atrial cavity with attachment to the interatrial septum. This sizable mass resulted in the mild obstruction of tricuspid valve inflow. CT guided biopsy of the mass lesion was performed. Histological analyses revealed a spindle cell tumor with smooth muscle origin (figure 3).

Immunohistochemical study findings confirmed the diagnosis by the positive results for smooth muscle actin (SMA), desmin, and vimentin in favor of LMS (figure 4).

Because of the advanced stage and extension of the tumor from right atrium up to renal veins and concomitant liver metastatic lesion, surgery was not considered as a suitable option in this case. The patient underwent palliative chemotherapy with a combination of doxorubicin and ifosfamide as the last resort. The patient developed hepatic encephalopathy and liver failure and succumbed to death 6 weeks after admission.

DISCUSSION

Malignant tumors originate from soft tissues that compose less than 1% of all malignant tumors

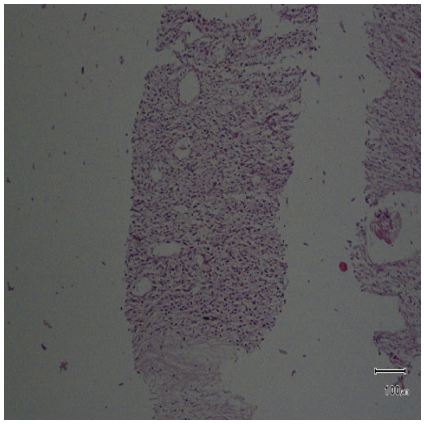


Fig.3: Tissue biopsy shows a hypercellular spindle cell neoplasm with smooth muscle cell origin, Hematoxylin and eosin (H&E) (Magnification × 100).

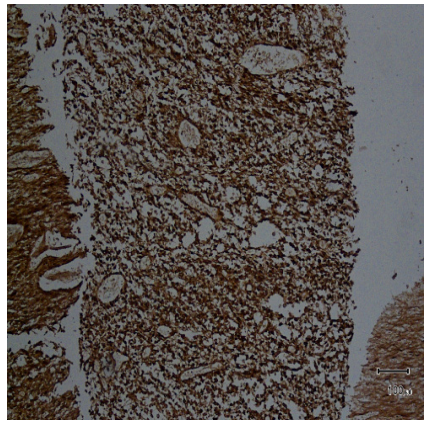


Fig.4: Immunohistochemistry shows positive staining for vimentin, desmin, and smooth-muscle α-actin. The diagnosis of leiomyosarcoma was confirmed.

in adults. Of those, LMSs account for less than 5%. Meanwhile, LMSs with vascular origin are rarer encompassing less than 1% of all soft tissue tumors. About 2% of LMSs are originated from IVC, which is the most commonly involved vascular structure (4). These tumors are slow-growing; as a result, LMSs of IVC would remain asymptomatic for a long time before definite diagnosis. The lag between disease onset and diagnosis usually leads to poor prognosis and decreased survival rate. LMS of IVC grows extraluminally in more than 95% of cases. So, these lesions may be mistaken for masses of adjacent organs. Therefore, the differential diagnoses should include primary tumors of the other organs (5).

According to the location of the tumor, IVC can be divided into three parts: the upper part from the hepatic vein confluence up to the right atrium, the middle part between the hepatic vein confluence to the renal veins, and the lower part below the renal veins (6). An increased risk of death, lower limb edema, Budd-Chiari syndrome, intraluminal tumor growth, and IVC occlusion are mainly associated with the tumors involving the upper IVC segment. Tumors occur in the lower and middle parts of IVC in 44.2% and 50.8% of cases, respectively. Whereas, only a small number of tumors occur in the upper third or suprahepatic region (4.2%) (1). Patients with upper segment tumors usually present as having Budd-Chiari syndrome with hepatomegaly, jaundice, and massive ascites (7). Metastasis has been reported in fewer than 50% of cases at the time of diagnosis, which is commonly seen in the liver and lung, suggesting hematogenous or lymphatic spread (8).

The findings of CT and magnetic resonance imaging (MRI) are likely to be most helpful in distinguishing this tumor from other retroperitoneal tumors. Although imaging studies may strongly suggest a primary tumor of the IVC, a biopsy taking is required and is best performed using ultrasonography (9). Complete resection of primary LMS of IVC is feasible and associated with improved survival. The IVC can be managed by primary repair or ligation. In a prospective follow up of postoperative patients, local recurrence occurred in 33% of patients and distant recurrence occurred in 48% of the patients. Patients undergoing complete resection had 3-year and 5-year disease-specific survival rates of 76% and 33%, respectively. There were no 3-year survivors among patients with incomplete resections (10).

Doxorubicin is the key drug for the treatment of advanced soft tissue sarcoma. Combination therapy showed better response and time to treatment failure than monotherapy but did not show better overall survival (11).

CONFLICT OF INTEREST

The authors declare no conflict of interests related to this work.

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