CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 927–931

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Cardiac metastases and tumor embolization: A rare sequelae of primary undifferentiated liver sarcoma

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ARTICLE INFO

Article history:
Received 25 July 2014
Received in revised form
21 September 2014
Accepted 6 October 2014
Available online 16 October 2014

Keywords:
Liver sarcoma
Hepatectomy
Cardiac metastases
Tumor emboli

ABSTRACT

INTRODUCTION: Primary hepatic sarcomas are uncommon malignant neoplasms; prognostic features, natural history, and optimal management of these tumors are not well characterized.
PRESENTATION OF CASE: This report describes the management of a 51-year-old patient that underwent a right trisectionectomy for a large hepatic mass found to be a liver sarcoma on pathology. He subsequently developed tumor emboli to his lungs and was discovered to have cardiac intracavitary metastases from his primary tumor. The patient underwent cardiopulmonary bypass and resection of the right-sided heart metastases to prevent further pulmonary sequela of tumor embolization.
DISCUSSION: The lack of distinguishing symptoms or imaging characteristics that clearly define hepatic sarcomas makes it challenging to achieve a diagnosis prior to pathologic examination. Metastatic spread is frequently to the lung or pleura, but very rarely seen within the heart. Failure to recognize cardiac metastatic disease will ultimately lead to progressive tumor embolization and cardiac failure if left untreated.
CONCLUSION: The most effective therapy for primary liver sarcomas is surgery; radical resection should be performed if possible given the aggressive nature of these tumors to progress and metastasize.
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1. Introduction

Primary hepatic sarcomas are extremely rare tumors that comprise a heterogeneous group of histological subtypes including angiosarcoma, leiomyosarcoma, undifferentiated (embryonal) sarcoma (UES), epithelioid hemangioendothelioma, and fibrosarcoma. In the adult population, these sarcomas, as a group, represent less than 1% of liver malignancies[1,2]; the various histological compositions, in addition to vague symptoms, lack of common tumor markers, and inconsistent imaging findings often make diagnosis difficult with delays in subsequent treatment.

2. Presentation of case

A 51-year-old male patient developed progressive right upper quadrant abdominal pain over several weeks. He presented to a local hospital with acute pain; computed tomography (CT) imaging demonstrated a 28 cm complex cystic tumor with septal enhancement, nodularity, and inferior vena cava (IVC) compression (Fig. 1). Aspiration of the tumor was noted to be initially hemorrhagic; this was followed by percutaneous drainage and removal of 4L serosanguinous fluid for symptom relief. The fluid quickly re-accumulated and he was transferred to our hepatobiliary surgical service for further management.

Due to mass effect of the large tumor and resultant anatomic stretch of the bile ducts, the patient was mildly jaundiced with an elevated bilirubin level of 3.7 mg/dL and alkaline phosphatase level of 509U/L. A percutaneous transhepatic biliary drain was placed into the left lateral segment of the liver for preoperative drainage and his jaundice improved. The tumor had imaging features concerning for malignancy, but minimal overall solid component; therefore, the differential diagnosis included biliary cystadenoma, cystadenocarcinoma, or less likely an intrahepatic sarcoma. There

Abbreviations: CT, computed tomography; IVC, inferior vena cava; PE, pulmonary emboli; UES, undifferentiated (embryonal) sarcoma; TVE, total vascular exclusion.

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http://dx.doi.org/10.1016/j.ijscr.2014.10.004
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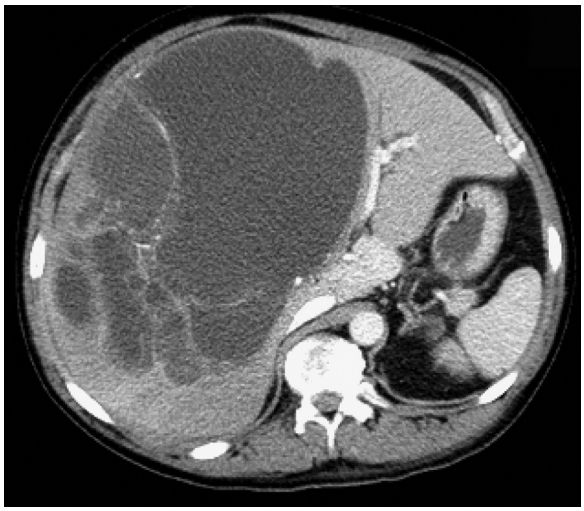

Fig. 1. CT scan of primary liver sarcoma. Axial CT scan slice of a 28 cm complex, multiloculated cystic mass with enhancing mural nodularity and septations predominantly in the right lobe of the liver and extending into the left medial section. There is mass effect from the liver displacing the IVC and left portal vein.

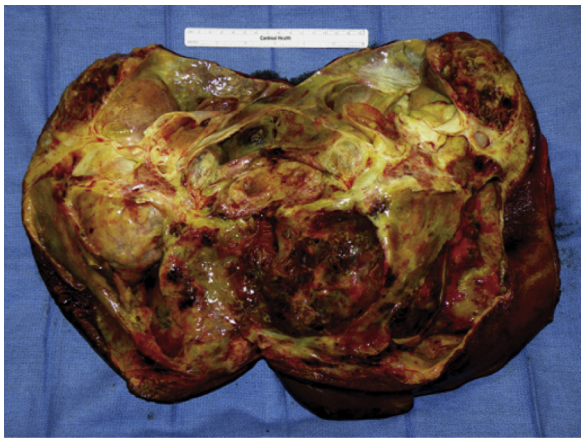

Fig. 3. Gross architecture of liver sarcoma. The specimen is bivalved to demonstrate a heterogeneous mass of solid and cystic components with areas of hemorrhage and necrosis.

was a strong probability that the tumor had bled both into itself and into the peritoneum adjacent to it (causing his acute onset of symptoms); however, the cytology from the aspiration done at the local hospital was negative for malignancy. He underwent a right trisectionectomy with caudate lobectomy via a thoracoabdominal approach (Fig. 2A) given the large tumor size (Fig. 2B). Despite division of the right liver inflow, the weight of the tumor against the IVC produced a very high physiologic central venous pressure. This did not permit an anterior approach to parenchymal division; we encountered brisk venous back-bleeding during the initial attempt at transection. The right liver was mobilized and the short hepatic veins and right hepatic vein were divided. Total hepatic vascular exclusion was obtained by placing one Rummel tourniquet around the left liver inflow (for Pringle maneuver) and another tourniquet around the common middle and left hepatic vein outflow. Two periods of vascular clamping were used to divide the liver parenchyma; total occlusion time was 33 min. During the operation, anesthesia had placed a transesophageal echocardiogram probe for routine continuous monitoring which suggested a vegetation on the tricuspid valve – though the significance of this finding was uncertain so further evaluation was planned for after surgery. The liver specimen weighed 4650 g and demonstrated a heterogeneous mass with areas of hemorrhage and necrosis (Fig. 3). Histological sections demonstrated atypical spindled cells with hyperchromatic nuclei and multiple areas of necrosis consistent with an undifferentiated liver sarcoma of intermediate-grade.

On post-operative day one, the patient developed hypoxia and a CT angiogram of the chest demonstrated small, right-sided segmental pulmonary emboli (PE). A transthoracic echocardiogram confirmed a mobile mass on the triscupid chordal apparatus. The patient was anticoagulated and subsequent blood cultures grew *staphylococcus*. The combination of events seemed odd, but he was presumptively diagnosed with endocarditis (believed to have been present preoperatively) and placed on a 6-week regimen of antibiotics. He recovered without further sequela was discharged 11 days after his initial operation. A repeat echocardiogram at 8 weeks postoperatively demonstrated a second mass originating from the right atrium and resultant pulmonary hypertension. Interval CT angiogram of the chest also showed progressive pulmonary emboli, despite being on therapeutic Lovenox. Finally at 10 weeks postoperatively, repeat imaging showed an increased size of the mass attached to the tricuspid chordal apparatus with extension into the main pulmonary artery (Fig. 4A) as well as a second mobile mass within the IVC originating from the left hepatic vein and extending into the right atrium (Fig. 4B). The bilateral pulmonary emboli had increased in size and number, suggestive of truly being tumor emboli from the cardiac mass.

Given such rapid progression and risk of further embolization, he was referred to cardiothoracic surgery for resection of the cardiac mass. A median sternotomy was performed and after initiation of cardiopulmonary bypass, umbilical tapes were placed around the superior and inferior vena cava. The right atrium was opened

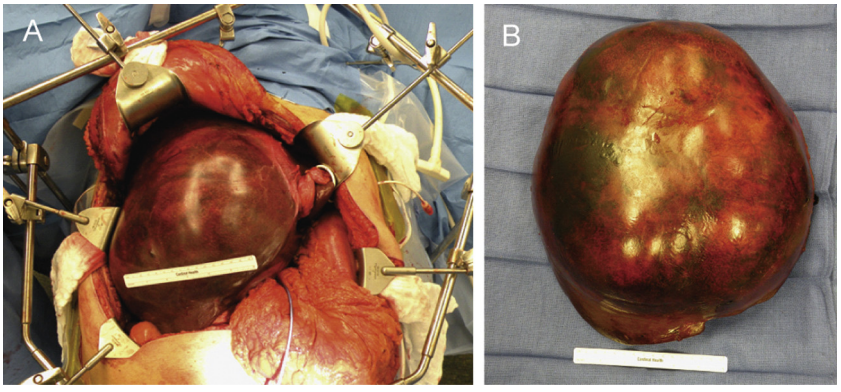

Fig. 2. Resection technique and specimen. Operative photographs illustrating (A) the thoracoabdominal extension used to split the costal margin and diaphragm for improved exposure of the massive tumor; and (B) the specimen after right trisectionectomy.

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