Case 10779

Eurorad

Primary malignant fibrous histiocytoma of the liver presenting in a young woman

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Section: Abdominal imaging
Area of Interest: Liver
Procedure: Contrast agent-intravenous
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 23 years, female

Clinical History:

A 23-year-old woman was admitted due to abdominal fullness, right upper quadrant pain and fever of one month's duration. Physical examination revealed hepatomegaly and liver tenderness. Laboratory tests showed leukocytosis and elevated serum transaminases. AFP and CEA were within normal limits and HBsAg was negative.
Imaging Findings:

MDCT of the abdomen revealed severe hepatomegaly. Unenhanced scanning demonstrated a large, ill demarcated heterogeneously hypodense mass in the right lobe of the liver (Fig. 1). Contrast enhanced CT at arterial phase revealed rapid, early, marked enhancement in the periphery of the mass and an abundance of large, tortuous blood vessels surrounding the tumour (Fig. 2). At portal and delayed phases mild contrast enhancement of the solid components of the mass was noted (Fig. 3, 4). Large hypodense parts within the tumour corresponded to areas of necrosis on histopathology. A small amount of ascites was present. No evidence of portal vein invasion, bile duct obstruction or regional lymph node metastasis was detected.
Clinical history and CT imaging features of a large necrotic mass were strongly suggestive of a hepatic sarcoma. Histology following an extended right hepatectomy reported the presence of primary malignant fibrous histiocytoma of the liver, of the inflammatory variety.
Discussion:

Background
Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma. It is considered to originate from undifferentiated primitive mesenchymal cells and has five different histological subtypes: storiform pleomorphic, myxoid, giant cells, inflammatory, and angiomatoid. It is a relatively rare tumour, however, it is the commonest sarcoma arising in adult population (6th-7th decades) with a slight male predominance. It may occur in any part of the body and most commonly in the extremities (lower extremity 49%, upper extremity 19%), followed by the retroperitoneum (16%) and peritoneal cavity (5-10%) [1-6].
Liver is an exceptional site of involvement, with no predilection for one lobe. The prognosis is poor, with a 2-year survival rate of approximately 60% [4-6]. This is a case of primary MFH of the liver presenting in a young woman.
Imaging perspective
The CT findings of primary MFH of the liver are closely correlated with the histopathologic characteristics and the

size of the lesion. At unenhanced CT, it usually appears as an ill-defined or a well circumscribed large or multinodular mass, heterogeneously hypodense, owing to the presence of necrosis. Occasionally, intratumoural calcifications have been reported [1-3].

Following contrast material injection, various enhancement patterns may be noted. A strongly and heterogeneously enhancing mass with several necrotic areas is usually seen. An abundance of blood vessels surrounding the tumour has also been reported and this was seen in our patient. This pattern is mainly correlated with the inflammatory variety of MFH [1, 2]. CT findings also include the presence of a contrast-enhancing pseudocapsule surrounding the lesion or a multilocular cystic mass with fibrous septa enhancement. Solid components and fibrous septa may better enhance on delayed scanning, depending on the amount of fibrosis. Small lesions may appear solid and homogeneously enhancing. Cong et al. reported the CT findings in five patients with MFH of the liver as a 'fast rushing in and washing out' enhancement pattern, probably due to the presence of less fibrous tissue in these tumours [2].

MFH may involve the liver capsule and adjacent organs, but without evidence of portal vein invasion, bile duct obstruction, or regional lymph node metastases.

Although there are no specific imaging findings of liver MFH it should be included in the differential diagnosis in a patient with a large necrotic mass with rapid growth combined with an aggressive clinical course, as in this case [1-6].

Differential Diagnosis List: Primary malignant fibrous histiocytoma of the liver, Hepatic adenoma, Haemangioma, Hepatocellular carcinoma (HCC), Fibrolamellar HCC, Cholangiocarcinoma, Hepatic epithelioid haemangioendothelioma, Undifferentiated embryonal sarcoma

Final Diagnosis: Primary malignant fibrous histiocytoma of the liver

References:

Karki B, Xu Y-K, Wu Y-K, et al (2012) Primary malignant fibrous histiocytoma of the abdominal cavity: CT findings and pathological correlation. World J Radiol April 28; 4(4):151-158 (PMID: 22590669)

Cong Z, Gong J, et al (2011) Primary malignant fibrous histiocytoma of the liver: CT findings in five histopathological proven patients. Abdom Imaging 36:552–556 (PMID: 21287171)

Yu R-S, Chen Y, Jiang B, et al (2008) Primary hepatic sarcomas: CT findings. Eur Radiol 18: 2196–2205 (PMID: 18463872)

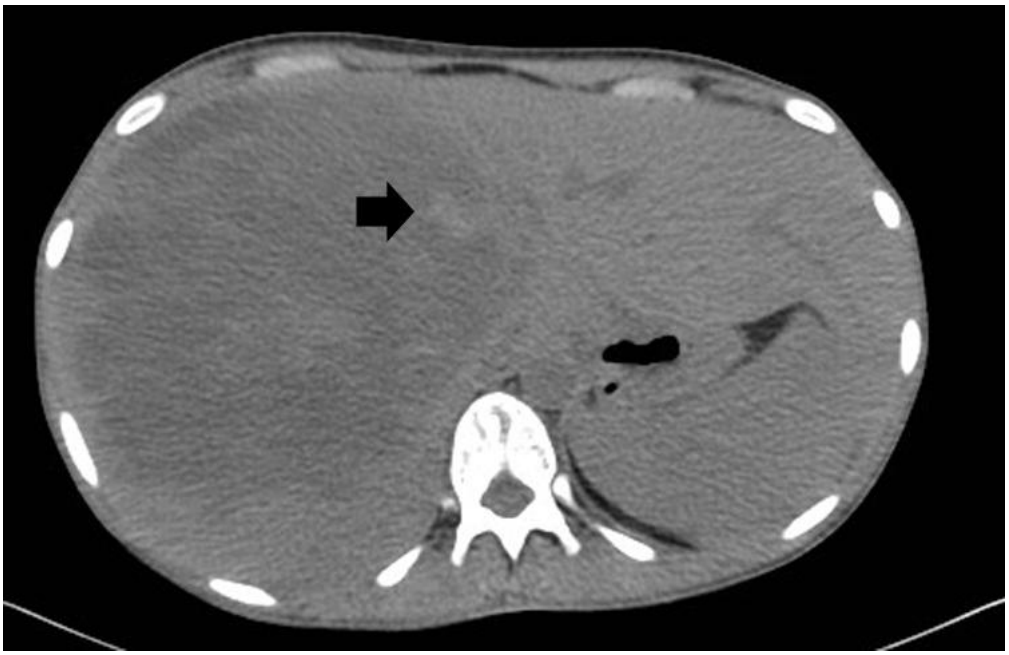
Ferrozzi F, Bova D (1998) Hepatic malignant fibrous histiocytoma: CT findings. Clin Radiol 53:699–701 (PMID: 9766725)

Reed JG, Goodman P, Soloway RD (1993) Primary malignant fibrous histiocytoma of the liver MRI findings. Magn Reson Imaging 11:139–143 (PMID: 8380881)

Yu JS, Kim KW, Kim CS, et al. (1999) Primary malignant fibrous histiocytoma of the liver: imaging feature of five surgically confirmed cases. Abdom Imaging 24:386–391 (PMID: 10390563)

Figure 1

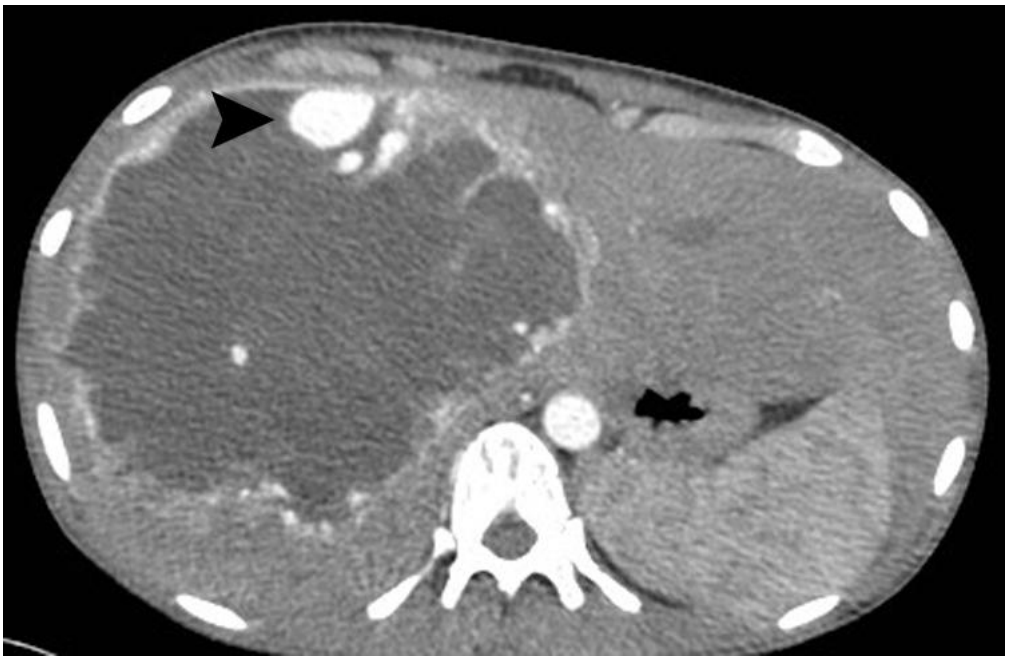
a



Description: Unenhanced transverse CT image depicts a large, poorly demarcated mass involving the right lobe of the liver, of heterogeneous low density. Hyperdense parts (arrow) within the lesion corresponded to areas of haemorrhage on histology. **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece

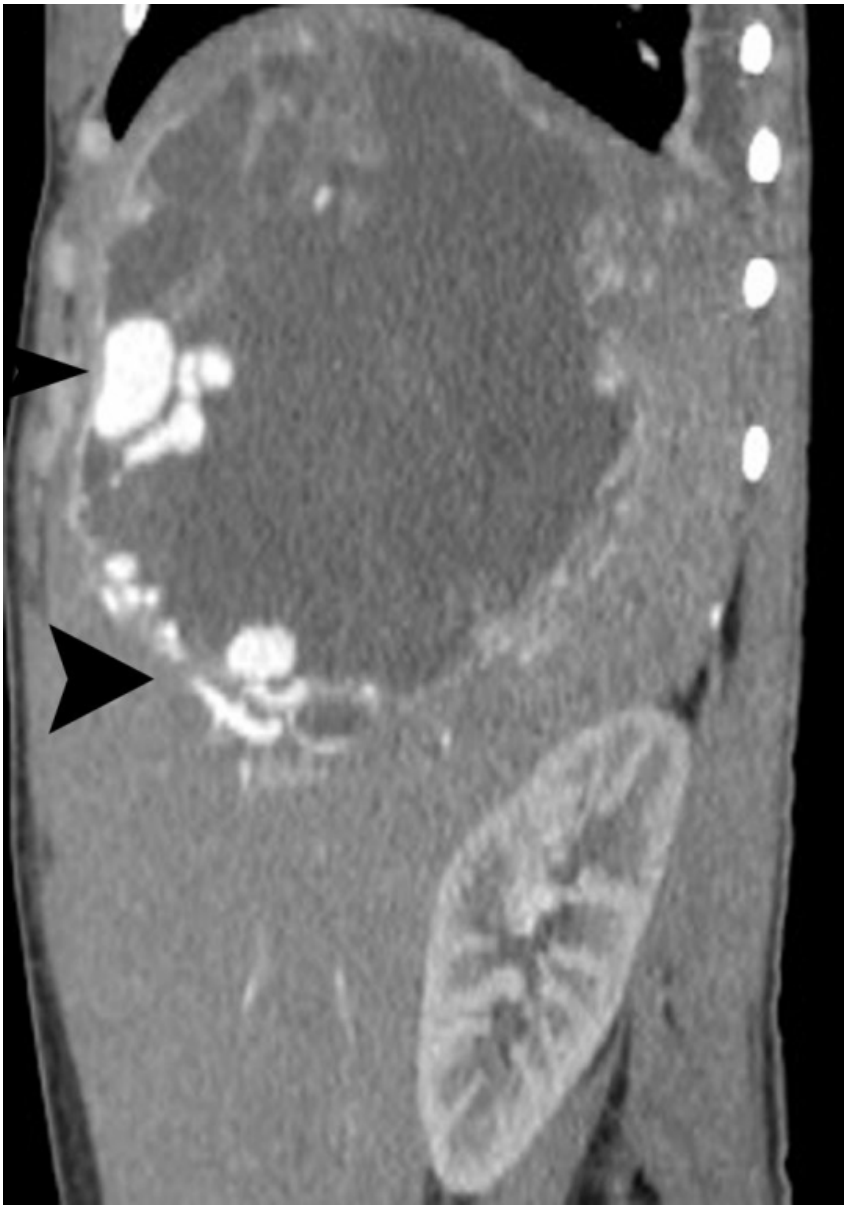
Figure 2

a



Description: Transverse contrast-enhanced image (arterial phase) shows marked enhancement in the periphery of the lesion and abundant large and tortuous blood vessels surrounding mass (arrowhead).
Origin: Ntorkou A, Department of Radiology, Ioannina, Greece

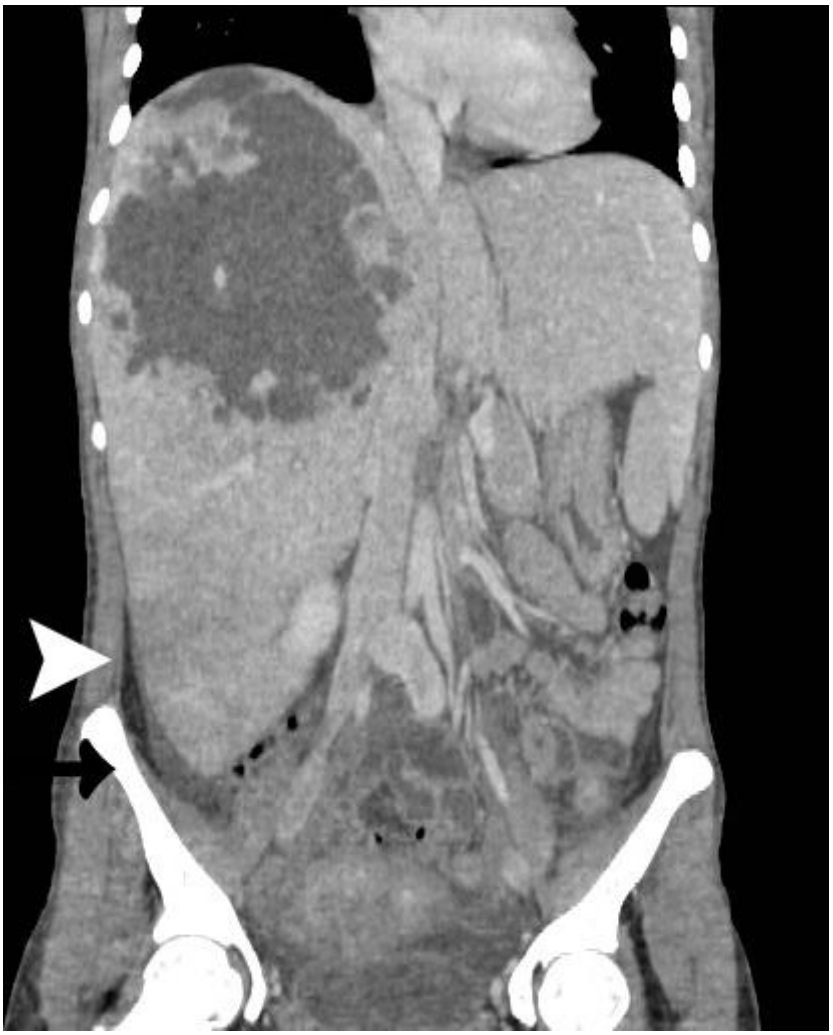
b



Description: Sagittal contrast-enhanced reformation (arterial phase) depicts extreme hepatomegaly and tumour's supply from multiple dilated, tortuous surrounding vessels (arrowheads). **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece

Figure 3

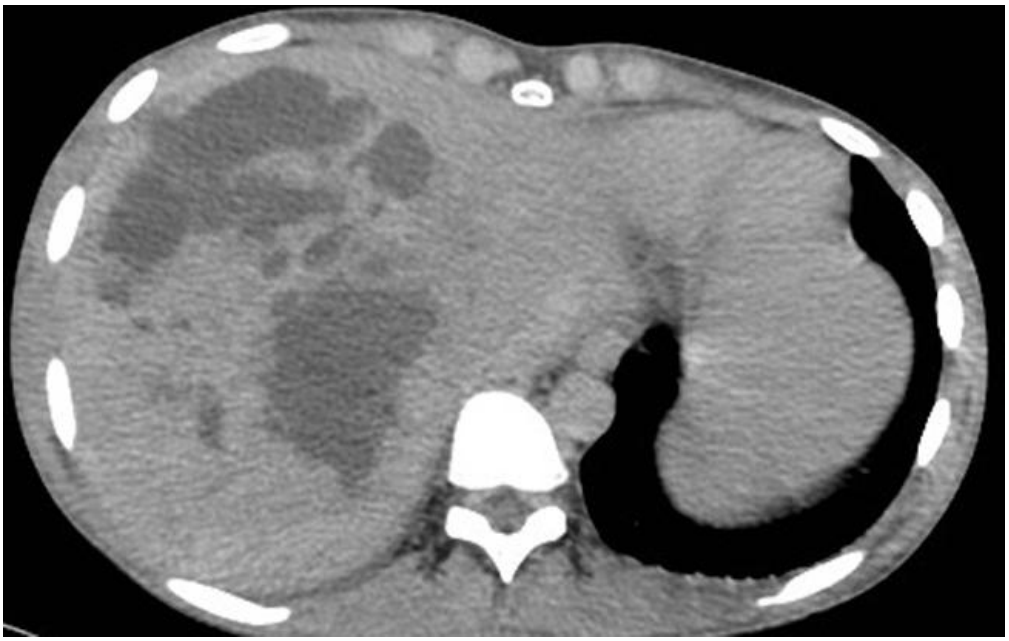
a



Description: Coronal contrast-enhanced reformation (portal phase) shows mild enhancement of the solid components of the tumour. Transient perfusion disorders are seen in segment VI (arrowhead). A small amount of ascites is also present (arrow). **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece

Figure 4

a



Description: Transverse contrast-enhanced image (delayed phase) depicts centripetal enhancement of the solid components of the mass. Hypodense areas within the tumour corresponded to areas of necrosis on pathology. **Origin:** Ntorkou A, Department of Radiology, Ioannina, Greece