Primary Hepatic Undifferentiated Pleomorphic Sarcoma: CT and angiographic findings in two cases

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ABSTRACT

Primary hepatic undifferentiated pleomorphic sarcoma (UPS) is an extremely rare disease entity whose imaging characteristics are not well documented in literature. We present two cases of primary hepatic UPS with contrasting disease courses, and their CT and angiographic findings in this report. Imaging characteristics include inhomogeneous appearance and peripheral enhancement pattern, with necrosis and delayed gradual fill-in enhancement more evident in the larger tumor. Although imaging findings are not specific, being familiar with the common manifestations of primary hepatic UPS will allow radiologists to include it in their differential diagnosis list for suspected sarcomatous or mesenchymal tumors of liver.

Undifferentiated pleomorphic sarcoma (UPS), previously known as malignant fibrous histiocytoma (MFH), is the most common soft tissue sarcoma of late adult life [1]. UPS primarily occurs in the extremities and less commonly involves the retroperitoneal spaces and other sites [2]. Primary hepatic UPS is very rare and its imaging characteristics have not been well documented in literature. Here we present two cases of primary hepatic UPS with contrasting presentations, their CT and angiographic findings, and their outcomes.

CASE REPORTS

The first case was a 72-year-old man who came to the hospital for further evaluation of an incidental hepatic tumor found on sonography in a health exam. Physical exam and laboratory data were unremarkable. Serum alpha fetoprotein (AFP) level was within normal limit. Subsequent CT exam showed a solitary hypodense nodule with ill-defined margin measuring 2.5 cm in segment 6 of liver, with a peripheral enhancement pattern (Fig. 1a-1d). The underlying liver showed fatty metamorphosis and mild cirrhotic change. Diagnostic angiography showed an irregular hypervascular stain in right hepatic lobe (Fig. 1e, 1f). He underwent curative resection under the clinical impression of hepatocellular carcinoma (HCC). Histopathology revealed UPS. He had a smooth postoperative course without local recurrence, but developed pleural and vertebral metastases ten months after surgery. He declined systemic chemotherapy but received palliative radiotherapy to the thorax and lumbar spine. He was lost to follow-up at one-year-and-two-months after surgery.

The second case was a 64-year-old man who presented with a two-week history of abdominal fullness and pain. He denied having any past medical history, but serology tests revealed anemia (hemoglobin = 7.3 g/dL), and hepatitis B and C. Serum carbohydrate antigen 19-9 (CA 19-9) level was elevated (64.4 U), while AFP and carcinoembryonic antigen (CEA) levels were within normal limits. CT at emergency department showed a lobulated mass measuring 14 cm occupying segments 4, 5 and 8 of liver, with an initial peripheral and gradual fill-in enhancement pattern, and central areas of necrosis (Fig. 2a-2d). The tumor had ruptured at its lateral margin, with blood clot in the right perihepatic space. Diagnostic angiography showed a huge
Figure 1. 72-year-old man with primary hepatic undifferentiated pleomorphic sarcoma. Pre-contrast a. and post-contrast CT at arterial b., portal venous c. and equilibrium d. phases showed a solitary small hypodense nodule (arrow) with ill-defined margin in segment 6 of liver, with a peripheral enhancement pattern. Diagnostic angiography (e. and f.) showed an irregular hypervascular stain (arrow) in right hepatic lobe.
Figure 2. 64-year-old man with primary hepatic undifferentiated pleomorphic sarcoma. Pre-contrast a. and post-contrast CT at arterial b., portal venous c. and equilibrium d. phases showed a lobulated hypodense mass (arrow) occupying segments 4, 5 and 8 of liver, with an initial peripheral and gradual fill-in enhancement pattern, and large central areas of necrosis. The tumor had ruptured at its lateral margin, with blood clot in right perihepatic space. Diagnostic angiography (e. and f.) showed a huge hypervascular stain with peripheral pattern of enhancement in right side of liver.
hypervascular stain with peripheral pattern of enhancement in right side of liver, without evidence of active bleeding (Fig. 2e, 2f). Both the reporting radiologist and surgeon were under the impression of a ruptured HCC. He received right lobectomy, followed by systemic chemotherapy after histopathology revealed UPS. Tumors recurred in liver within two months along with multiple lung metastases. He died of fulminant disease at two months after surgery.

DISCUSSION

O-Brien and Stout first described the malignant fibrous xanthoma in 1964 [3], which later became known as MFH. It was reported to be the most common sarcoma in late adult life with a peak incidence in the fifth to sixth decades [1]. MFH was originally described as a heterogeneous group of tumors that were classified into the storiform-pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid subtypes based on morphology [4]. The current World Health Organization classification of MFH had been narrowed down to a pleomorphic sarcoma without definable lines of differentiation [5], and hence the name UPS.

UPS mainly affects the extremities, but may also occur in the retroperitoneal spaces, abdominal cavity, and other uncommon sites [2]. Primary UPS that arises from the liver is extremely rare. Since its first description in 1985, less than 60 of such cases have been reported in English literature [6]. Its clinical presentation is nonspecific, which may include abdominal pain, fever, body weight loss, fatigue, anorexia, and jaundice. Abdominal pain and fullness usually become apparent when the tumor reaches a certain size or has invaded the capsule, as in our second case. Some patients may be asymptomatic, as in our first case.

The radiologic features of primary hepatic UPS can be nonspecific. On plain CT, UPS usually appears as a large, poorly-demarcated or encapsulated, inhomogeneous and hypodense mass with central necrosis [7-9]. The extent of necrosis or cystic change increases with tumor size [10]. Intratumoral calcification has been reported [7]. After contrast administration, UPS usually shows heterogeneous enhancement, but small tumors can enhance homogeneously with early washout [10]. Delayed enhancement can be observed and the intensity increases with increasing fibrosis [9]. The inhomogeneous nature and peripheral enhancement pattern in both our cases are comparable to those described in literature. In retrospect, the delayed gradual fill-in enhancement of the larger tumor in the second case should have hinted at a mesenchymal or sarcomatous tumor, or even a cholangiocarcinoma, other than the ruptured HCC as reported.

Angiography is a useful adjunct to cross-sectional imaging, and especially helpful in the case of a large intraabdominal tumor where origin is uncertain. There have been very few reports with angiographic findings of primary hepatic UPS. Marginal tumor staining without central feeding arteries has been reported as a characteristic feature [11]. This recurring peripheral enhancement pattern is consistent with the angiographic findings in both our cases, but less apparent in the first case. We believe this is due to smaller size and less fibrous content in the tumor, where tumor stain can appear more homogeneous, making it more difficult to differentiate from HCC on angiography.

MR imaging is another useful modality for hepatic tumors, especially when used in conjunction with hepatocyte-specific gadolinium chelates, but not performed in either of our cases. On MR imaging, primary hepatic UPS has been reported as a large encapsulated mass of heterogeneous signal intensity on both T1- and T2-weighted images [12]. The varying signal intensity may reflect the amount of intratumoral myxoid content, fibrosis, hemorrhage and necrosis. After contrast enhancement, inhomogeneous enhancement of solid components of tumor can be observed [7].

Sarcomatoid HCC and intrahepatic cholangiocarcinoma are two primary epithelial neoplasms of the liver that must be excluded first before a diagnosis of primary hepatic UPS can be made [6]. The former may occur in patients with underlying viral hepatitis or cirrhosis, and with areas showing signs of more conventional HCC, such as strong arterial enhancement, early washout on portal venous phase, and portal venous thrombosis. Intrahepatic cholangiocarcinoma shares the same peripheral and delayed enhancement pattern as UPS, but has other characteristics such as capsular retraction and intrahepatic bile duct dilatation to aid in differential diagnosis. Other primary mesenchymal neoplasms of the liver such as angiosarcoma, hepatic epithelioid hemangioendothelioma, dedifferentiated liposarcoma, leiomyosarcoma, and fibrosarcoma should also be considered in the differential diagnosis [6], but definite diagnosis of these tumors are reliant on immunohistochemical staining. When primary hepatic UPS assumes a cystic appearance, the differential diagnosis should also include biliary cystadenocarcinoma and hepatic abscess.

The prognosis for primary hepatic UPS is very poor. About two-thirds of patients die within one year of diagnosis [6]. Poor prognosis is associated with large tumor size, late clinical stage, and a high ezrin expression score on histopathology [6]. Surgical resection with negative resection margins is the mainstay of treatment for primary UPS of the liver. Unfortunately, distant metastasis can occur in spite of a complete curative resection, as seen in the first case. Late clinical stage at presentation is the main reason for the poor outcome in our second case.

In conclusion, primary hepatic UPS is a very rare malignant mesenchymal tumor with nonspecific clinical manifestation and variable radiologic features, making preoperative diagnosis difficult. Key imaging characteristics include inhomogeneous appearance and peripheral enhancement pattern, with necrosis, cystic change and
Primary hepatic undifferentiated pleomorphic sarcoma

delayed gradual fill-in enhancement more evident in larger
tumors. Although imaging findings are not specific, being
familiar with the common manifestations of primary hepatic
UPS will allow radiologists to include it in their differential
diagnosis list for suspected sarcomatous or mesenchymal
tumors of liver.

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