

Malignant Fibrous Histiocytoma of the Liver:

A Case Report and Review of the Literature

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Abstract

Malignant fibrous histiocytoma (MFH) is a common soft tissue sarcoma, usually occurring in the extremities. MFH of the liver is an extremely rare neoplasm, with only 28 cases reported in the international literature since 1985. We present a case of MFH of the liver in an 87-year-old woman. The tumor was located in the right lobe of the liver and measured 12 × 8 cm. It consisted of spindle-shaped, pleomorphic, malignant cells in a storiform pattern associated with histiocyte-like cells and giant cells. Most of the tumor cells and giant cells were vimentin and α_1 -antichymotrypsin positive. Histopathological findings were consistent with an MFH of the storiform / pleomorphic subtype. The literature is briefly reviewed.

Key Words: Histiocytoma, liver, sarcoma.

Introduction

MALIGNANT FIBROUS HISTIOCYTOMA (MFH) represents the most common soft tissue sarcoma in adults (1). It can occur almost everywhere, owing to its mesenchymal origin (2, 3). However, its origination in the liver is extremely rare, with only a few cases reported in the literature. A case of an MFH of the liver is presented and the literature is briefly reviewed.

Case Report

An 87-year-old woman was admitted to our hospital for evaluation of progressive weight loss of 15 kg (33 lbs) in 6 months, right upper quadrant pain, and low-grade fever. The patient denied any history of abdominal trauma, blood transfusions, or alcohol intake. Physical examination revealed a non-icteric

woman with a temperature of 37.7°C. Abdominal examination revealed tenderness in the right upper quadrant of the abdomen, while the liver was palpable 7 cm below the right costal margin. Chest X-ray showed an elevated right hemidiaphragm. Erythrocyte sedimentation rate was 112 mm/h, hematocrit 34.2%, white blood cell count 13,700 cells/mm³, and platelet count 617,000/mm³. Total serum protein, electrolytes, creatinine, glucose, transaminases, alkaline phosphatase and bilirubin were within normal limits. Fibrinogen was elevated (478 mg/dL), as was lactate dehydrogenase (1652 IU/L). Test results for carcinoembryonic antigen (CEA), α -fetoprotein (AFP), 19-9 carbohydrate antigenic determinant (CA 19-9), and hepatic viral markers were all negative.

Abdominal ultrasound revealed a large mass localized in the right lobe of the liver. Abdominal computerized tomography (CT), after bolus intravenous injection of contrast medium, confirmed the presence of a large, hypodense intrahepatic mass (12 × 8 cm) in the right hepatic lobe (Fig. 1). There was no evidence of adenopathy or retroperitoneal masses, and the pancreas was normal.

CT-guided biopsy of a solid portion of the tumor yielded spindle-shaped, pleomorphic, malignant cells arranged in sheets and fascicles with a prominent storiform pattern (Fig. 2). Numerous histiocyte-like

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Fig. 1. Abdominal CT scan showing a large hypodense mass involving almost the entire portion of the right liver lobe.

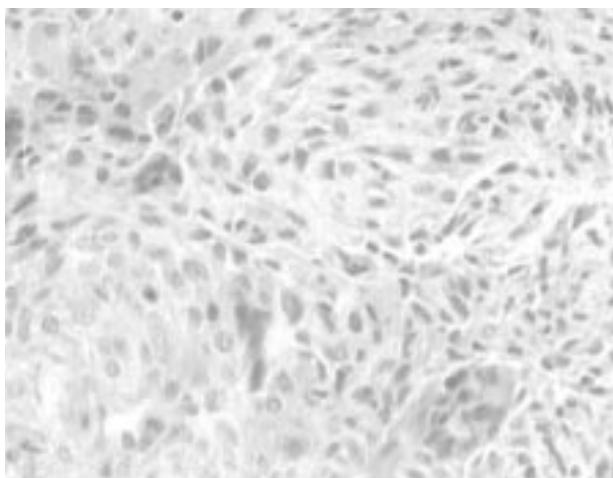


Fig. 2. Histology revealing the presence of pleomorphic spindle cells arranged in sheets and fascicles with a prominent storiform pattern (H&E, $\times 400$).

cells and giant cells were observed. Immunohistochemistry showed that the tumor cells and most of the giant cells and histiocyte-like cells were vimentin-positive (Fig. 3). Most of the tumor cells, including giant cells, were also cytoplasm positive for α_1 -antichymotrypsin. The histopathologic findings were consistent with an MFH of the storiform-pleomorphic type. Because of the large size of the tumor, the advanced age of the patient, and the presence of significant comorbidity, the tumor was judged to be inoperable. The patient died six months after diagnosis.

Discussion

MFH is a common soft tissue sarcoma. It usually occurs in the extremities, presenting as a painless mass, and less commonly in the retroperitoneal

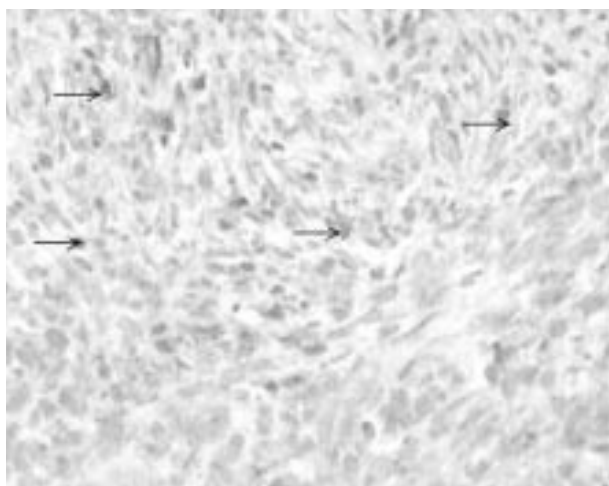


Fig. 3. Cytoplasmic immunoreactivity for vimentin was present in most tumor cells (see arrows, showing some of these cells) (immunoperoxidase stain, $\times 100$).

space, presenting with weight loss and increased intra-abdominal pressure (1). Since its initial description in 1963 (2), MFH has remained a rare neoplasm of uncertain histogenesis. It is thought to originate from undifferentiated mesenchymal cells, which are capable of multidirectional differentiation; this may explain why it has the potential to be found in all organs (3). Five histologic subtypes of MFH have been described: pleomorphic storiform (65% of cases), myxoid (15%), giant cell (10%), inflammatory (8%), and angiomatoid (2%). The first two subtypes tend to be high-grade neoplasms, while the others are usually low-grade sarcomas (1). The differential diagnosis of MFH should include pleomorphic liposarcoma and rhabdomyosarcoma. The former lacks the storiform pattern and shows evidence of cellular differentiation, while the latter shows cross striations on histologic examination. Weiss and Einzinger analyzed 200 cases of MFH (1) and showed that the 2-year survival rate of patients with pleomorphic / storiform type of MFH was 60% and the rate of metastases was 42%. Tumors larger than 10 cm had a rate of metastases of 57%, while those with a diameter from 5–10 cm, 34%. Surgery is the only effective method of treatment of MFH. Chemotherapy and radiotherapy have been used, but without success.

Primary MFH of the liver is very rare. It was first reported in 1985 (4), and since then only 28 other cases have been reported in the international literature (4–19) (16 men, 12 women, age range: 27–79 years, mean age: 51 years). Mean diameter of the tumor was 12 cm, with no predilection for one lobe or the other as the origin site. In 11 cases there was direct invasion of the adjacent organs; in only 2

cases were there distant metastases in the lungs. Most of the MFHs were pleomorphic; only two myxoid and one inflammatory type have been reported. In our patient, the MFH was the storiform / pleomorphic type, and immunohistochemically most tumor cells expressed vimentin and α_1 -antichymotrypsin. There was no evidence of metastatic disease at the time of diagnosis.

Most of the patients reported in the literature were treated surgically, but prognosis was poor. With a mean follow-up of 48 months, only 7 patients survived following surgery without any evidence of local recurrence or distant metastasis.

Modern imaging modalities (ultrasonography, CT) typically reveal a well-defined, large hepatic mass, with variable hemorrhagic necrosis, and invasion of the liver capsule or even tumoral spread to adjacent organs, without intratumoral calcification or regional lymph node metastasis. There is no specific imaging difference between hepatic MFH and MFH arising in other organs, but dystrophic calcification and metaplastic ossification—which is frequently reported in retroperitoneal and musculoskeletal MFH—has not been described in any case of hepatic MFH.

In conclusion, MFH of the liver is an extremely rare neoplasm with an aggressive biological behavior; it should be considered in the differential diagnosis of large liver lesions.

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