Malignant fibrous histiocytoma of the liver

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INTRODUCTION
Malignant fibrous histiocytoma (MFH) also known as malignant fibrous xanthoma or fibrousxanthoma is a common sarcoma occurring in the soft tissues, which usually originates in the extremities and less commonly in the retroperitoneal space\(^1\)-\(^5\). This lesion is a pleomorphic spindle cell neoplasm that was first described by O'Brien and Stout in 1964\(^4\). A primary lesion of the liver can rarely occur and only less than 50 cases have been reported so far in the literature \(^5\), so conclusions about forms of presentation and management can hardly be done. We are reporting a new case of primary MFH occurring in the liver. A review of the literature is done.

CASE REPORT
A 62-year old female patient was referred to us after a laparoscopic biopsy. During laparoscopy bleeding occurs and hemostasis could not be achieved. A laparotomy was performed and perihepatic packing was done. The patient was referred to our hospital for definite management. At admission, the patient looked very ill but stable. She presented abdominal pain and persisting bleeding from the abdominal drainage. Laboratory studies showed hemoglobin of 8.2 mg/dl, hematocrite 24\%, white cell count 20,000/ mm\(^3\). The liver function test were normal as well as the carcinoembrionic antigen, CA 19-9, and alpha fetoprotein. Abdominal ultrasound and computed tomography (CT) scan were performed that showed a 10 x 12 cm well circumscribed left side liver mass replacing the left lateral segment (fig. 1). No other intra-abdominal lesions were observed. She began with her symptoms two months before admission when presented abdominal pain, weight loss and an abdominal mass. A second laparotomy was performed. The liver mass was ill defined due to the previous operation and the perihepatic packing. Metastatic lymph nodes as well as other lesions were not observed. She began with her symptoms two months before admission when presented abdominal pain, weight loss and an abdominal mass. A second laparotomy was performed. The liver mass was ill defined due to the previous operation and the perihepatic packing. Metastatic lymph nodes as well as other lesions were not observed. The non-tumoral liver was not cirrhotic. A left lateral segmentectomy was performed. The evolution was uneventful and was discharged at the seventh postoperative day. Her conditions improved but adjuvant therapy was not considered.

Macroscopic and histological findings
The tumor measured 12 x 10 cm in size. The cut surface was yellowish and had a firm consistency. The tumoral area consisted of fusiform and atypical cells, with elongated pleomorphism and hyperchromatic
nuclei (fig. 2). Beside this neoplasm showed numerous atypical mitotic figures. Occasionally multinucleated cells with phagocytosis were found (fig. 3). Necrosis was present associated with hemorrhage. Immunocytochemical study revealed positivity in neoplastic cells to vimentin and alpha-1 antichimiotripsin, desmin and actine. Keratin and epithelial membrane antigen were negative (fig. 4).

The patient remained well but 6 months after heptectomy she presented abdominal pain. Abdominal CT scan showed a recurrent peritoneal lesion (fig. 5). The patient died of recurrent disease 12 months after operation.

**DISCUSSION**

Sarcomas of the liver are exceedingly rare neoplasms representing less than 1% of the primary malignant liver lesions\(^6,7\). Angiosarcoma is the most common of all mesenchymal tumors representing 56% followed by leimyosarcoma and fibrosarcoma that represent 12% and 7% respectively\(^8\). MFH is not included in the classifications of hepatic mesenchymal tumors, be-

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Fig. 1. Computed tomography scan of the liver that shows a left side liver focal lesion. Note that air is observed within its capsule due to a previous operation.

Fig. 2. Malignant fibrous histiocytoma showing tumoral area and necrosis (150 x, HE).

Fig. 3. Neoplastic cells are pleomorphic with occasionally giant cell and mitotic figures (300 x HE).

Fig. 4. Alpha-1-antichimiotripsin strongly positive (300 x, avidin-biotin peroxidase).

Fig. 5. Abdominal computed tomography scan that demonstrate peritoneal recurrent disease.
nantley in men at a median age of 55 years (range 35 to 78 years) so MFH appears to be a tumor of middle to late adult life\textsuperscript{2,13}. The lesion did not present predilection for one lobe as the site of origin. The size of the lesions recorded range from 6 to 23.5 cm (mean 11 cm). None of the patients had history of chronic liver disease or cirrhosis. Commonly, these patients come to treatment with an advanced disease presenting abdominal pain, sensation of an abdominal mass, fullness, discomfort, weight loss, dyspnea, anorexia, malaise and fever. Episodic hypoglycemia, hyperinsulinemia and unusual manifestation like fever with leukocytosis have also been attributed to MFH\textsuperscript{3,19,20}. Laboratory examinations as well as tumor markers did not help to make the diagnosis.

In general, MFH manifests a broad range of histological patterns and is divided into 5 histological subtypes: pleomorphic/storiform (60%-70% of cases), myxoid (10%-20%), giant cell (5%-15%), inflammatory (5%-10%) and angiomatoid (1%-5%)\textsuperscript{3,19,20}. The majority of the lesions arising in liver had a pleomorphic/storiform type as we can see in the table 1. Some authors reported immunocytochemical and ultrastructural similarities between embryonal sarcoma and the pleomorphic type of MFH\textsuperscript{10}. Nevertheless embryonal sarcoma of the liver occurs predominantly in children and in the soft tissue, these lesions occur exclusively in adults. MFH commonly occurs in adults with the exception of angiomatoid type of MFH that may occur in children\textsuperscript{3}. The combination of sarcoma and benign cystic structures has been observed in some hepatic sarcomas reported as “primary malignant mesenchymal tumors” or “undifferentiated (embryonal) sarcoma”\textsuperscript{3,10}. Although MFH is considered a high grade malignant neoplasm in the soft tissue, lesions with marked inflammatory reaction have a better prognosis that those with minimal inflammation. The presence of inflammation may be a better prognostic factor in hepatic sarcomas\textsuperscript{2,11,21}. Immunocytochemical study is made to differentiate the tumor from carcinoma and to investigate the differentiation of the sarcomas. The antisera used are keratin, desmin, S100 protein, and various proteins found in MFH, including alpha 1 antitrypsin, alpha 1 antichymotrypsin, ferritin and lysozyme\textsuperscript{11}. The present case was positive to alpha 1 antichymotrypsin, desmin and actine that confirmed our diagnosis\textsuperscript{11,12,14,15}. Imaging features of MFH were discussed by Yu et al\textsuperscript{5} using ultrasonography (US), CT, magnetic resonance imaging (MRI) and hepatic angiography. The lesion could vary on echogenicity in ultrasound, on CT scan most of the lesions shows a low density area in the zone of the lesion and there were no evidence of calcifications in any of the patients. Angiography shows that some times MFH presented avascular lesions and sometimes they present fine tumor vessels\textsuperscript{5,15}. Depending on the radiologic manifestations, there were non specific findings that could differentiate primary hepatic MFH from other hepatic lesions\textsuperscript{3}. Liver resection has been the most common form of treatment when it can be achieved, because is offered the best results. Chemotherapy and radiation therapy have been used without success. Liver transplantation have been used as a treatment of MFH in one case but systemic recurrence occur\textsuperscript{15}. Indications for liver transplantation for hepatic malignancies including sarcomas is controversial due to the poor prognosis\textsuperscript{22-24}. MFH is an aggressive sarcoma that has a 2 years survival rate of 60%, with a recurrence rate of 44% and a rate of metastases of 42%. The size of the tumor correlated with metastatic potential. Tumors from 5 to 10 cm had a metastatic rate 54%, and those greater than 10 cm, 57%\textsuperscript{2,14}.

**Key words:** malignant fibrous histiocytoma, sarcomas, immunocytochemical studies, embryonal sarcoma, liver tumors.

### References