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Letters to the Editor

Malignant fibrous histiocytoma – a rare hepatic tumor

Key words: Hepatic malignant fibrous histicytoma. Hepatic soft tissue sarcoma.

Dear Editor,

Malignant fibrous histiocytoma (MFH) represents the most common soft tissue sarcoma in adults. Visceral locations are uncommon and primary hepatic MFH is exceedingly rare.

Clinical case

We present a case of a 63-yr-old-man was admitted to our hospital because of abdominal distension causing discomfort, lower extremities edema and weight loss. Past history includes abusive chronic alcool ingestion and arterial hypertension. No fever, jaundice or pain. Abdomen was soft but volumous with positive ascitic wave, tenderness in the right hypocondrium but no distinct hepatomegaly was found.

No remarkable abnormalities in laboratory findings. α-feto-protein, CEA, CA19-9, AgHBs or Ac-HVC were negative.

Abdominal TC (Fig. 1) showed a large, heterogeneous mass in the right lobe of the liver, with multiple hipodensity nodules. There was presumptive evidence of peritoneum invasion. No other masses or evidence of lymphadenopathy were present in the abdomen, pelvis and retroperitoneum.

Ultrassound-guided needle biopsy of a solid portion of the tumor was performed.

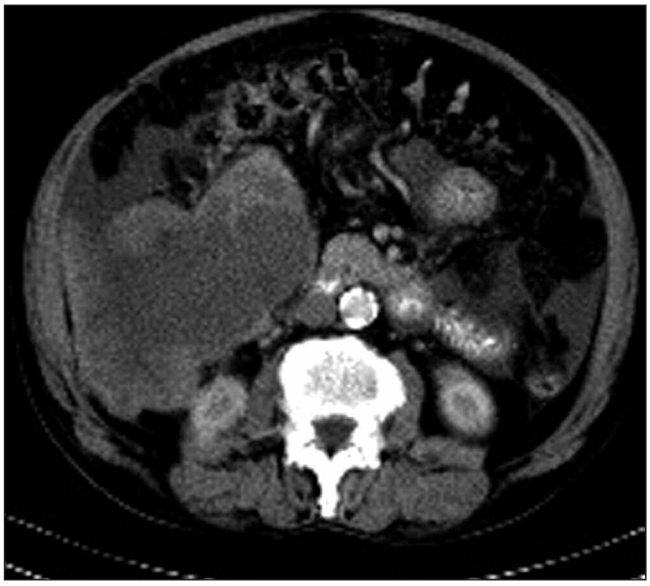

Fig. 1. Abdominal TC showing a large, heterogeneous mass in the right lobe of the liver, with multiple hipodensity nodules.

The histophatological findings were considered with a malignant fibrous histiocytoma, pleomorphic subtype (Fig. 2 A-H).

Patient died 45 days after admission.

Discussion

Since the first report of MFH in 1964 by O'Brien and Stout, it has been well recognized as the most common malignant soft tissue tumor. It usually occurs in the deep planes of proximal extremities, in the retroperitoneum and in the trunk, although it can occur almost anywhere owing to its mesenchymal origin, including bone (1).

To the best of our knowledge less than 35 cases of hepatic MFH have been reported over the last 24 years, since 1985 when it was first described (2).

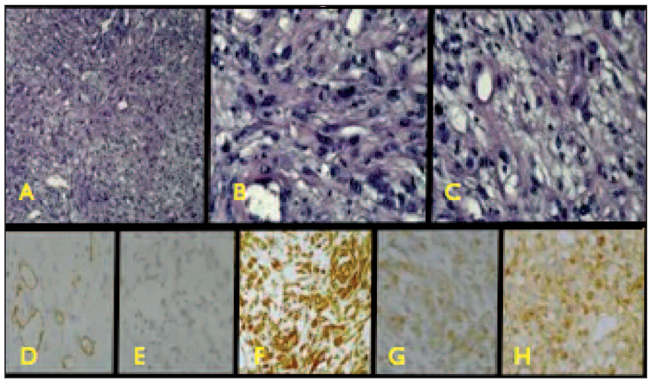

Fig. 2. The tumor was composed of interlacing bundles of spindle cells in a storiform disposition with intermingled histiocyte-like cells, exhibiting a pleomorphic pattern. A. H/E x100 magnification; B. and C. H/E x400 magnification. Immunohistological staining demonstrating negative reaction in the tumor cells for CD 31 (D) and S100 protein (E). Cytoplasmic immunoreactivity for vimentin (F), actin (G) and lysosim (H) is present in most tumor cells.

Primary MFH of the liver appears to be a tumor of late adulthood with a mean age of 58, where 23 of 32 cases noted are older than 50 years (age range: 27-87 years). No sex predisposition has been recognized (18 men; 14 women). Clinically symptoms, similar to the present case, are usually non-specific, and includes: weight loss, anorexia, fever, jaundice, malaise, right upper quadrant pain and palpable abdominal mass. Laboratory tests are usually unremarkable. No predilection for either lobe appears to be the rule. Mean diameter was 12,2 cm. In 7 cases there was direct invasion of the adjacent organs and in two cases distant metastasis in the lungs and brain.

Surgical treatment alone or combined with chemotherapy has been the mainstay of management for primary MFH of the liver, but prognosis was poor. Hepatic arterial embolization was performed in two cases with no sucess. Overall, the prognosis is grim where 16 out of the 32 reported patients were dead (and in other 8 cases it is not available if the patient died in the course of the disease) between 6 days and 34 months (median survival: 6 months), 11 of them were operated and died, in median, by the 6,8th month after surgery.

In conclusion, although of rare occurence, MFH should be considered in the differential diagnosis of large liver lesions. Tumor size, adjacent organ invasion or metastases at diagnosis remain the most important prognostic factor. The biopsy is determinant to the diagnosis, often delayed, being prognosis very poor.

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