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Case report

Malignant mesenchymatous tumor of the abdominal cavity

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SUMMARY

Malignant fibrous histiocytoma (MFH) represents a soft tissue sarcoma originating from fibroblast cells, and is characterized by a high rate of metastasis or recurrence. The development of this tumor in the gastrointestinal tract is a very rare entity. No more than 30 cases are described in the litrature.

We report on a case of such tumor of the abdominal cavity in a 45 year-old woman, presented with epigastric pain, anorexia and weight loss. The CT-scanning revealed the presence of multiple solid tumors in the peritoneal cavity. On the exploratory laparotomy at least 15 solid whitish tumors were found, attached to the wall of the small intestine, as well as to the parietal peritoneum. In addition, there were three metastases revealed in the liver. All tumors were excised; most of them were about 10 cm in diameter. The histopathologic study indicated the presence of a stromal tumor consisting of spindle cells. The consequent immuno-histo-chemical examination of the resected specimens established the definite diagnosis of a pleomorphic malignant fibrous histiocytoma.

The postoperative course of the patient was uneventful. She received adjuvant chemotherapy and is still alive and in good health two years later.

Key words: malignant fibrous histiocytoma, soft tissue tumors, stromal tumors, sarcomas, storiform-pleomorphic lesions.

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INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a pleomorfic sarcoma; its malignant potential has been recognized only recently. O'Brien and Stout first described it in 1964. The presence of this lesion in the GI could be considered as a very rare entity, since no more than 30 case reports on the subject are to be found in the literature.[1]

MFH's histologic features include spindle and round cells arranged in a storiform pattern with a number of pleomorfic, inflammatory and giant cells. It may be distinguished from other soft-tissue tumors by immuno-cytochemical staining.[2] This malignant tumor commonly arises from the deep soft tissues of the extremities of adults followed by the trunk and retro-peritoneum in decreasing order of frequency. Primary gastrointestinal MFH is an extremely rare lesion, as are intestinal secondary sites, which have also occasionally been described.[3]

In this paper, a case of a multi-centric malignant fibrous histiocytoma of the small intestine and the peritoneum, with liver implantations in an adult female patient is described, because of its rareness.

CASE REPORT

A 45-year-old woman was admitted to our Surgical Department with a history of persistent epigastric abdominal pain, anorexia and significant weight loss, reaching 10 Kg in the three months prior to admission.

The patient undergone open cholecystectomy for cholelithiasis and total hysterectomy for fibromyomatosis eight years and six years, respectively. After that, symphysiolysis for adhesive acute intestinal obstruction had been performed five years before admission. She had a long history of vague gastrointestinal tract complaints.

The routine laboratory investigation including biochemical parameters, blood count and erythrocyte sedi-

mentation rate, as well as the chest radiograph were normal. The CT-scanning of the abdomen demonstrated multiple solid tumors with rich vascularity (Figure 1). The latter was indicated by contrast medium concentration within the masses. After oncologist's consultation, it was decided to proceed with exploratory laparotomy.

On operation, at least, 15 solid whitish tumors were found in the peritoneal cavity, attached to the parietal peritoneum, as well as to the wall of the small intestine. In addition, three metastases in the liver were noted. All lesions were excised by debulking surgery; most of them being around 10 cm in diameter.

The histo-pathologic study of the resected specimens indicated the presence of a mesenchymatous (stromal) tumor consisting of splindle cells. The consequent immuno-histo-chemical examination of multiple sections of paraffin embedded tissue revealed a mixture of pleomorphic fibroblast-like spindle cells arranged in fascicles and a storiform pattern, with irregular, prominent, round-edged nuclei. A lot of cells stained positively for vimentin and CD 34 antigen. Stains for desmin, actin of smooth muscular fibers, S-100 protein and for surface membrane antigen, were negative. Thus, the above results established the definite diagnosis of malignant fibrous histiocytoma.

The patient was discharged after an uneventful postoperative course. She received adjuvant chemotherapy for six months. The patient remains in a good health two years after surgery, without evidence of metastasis or recurrence.

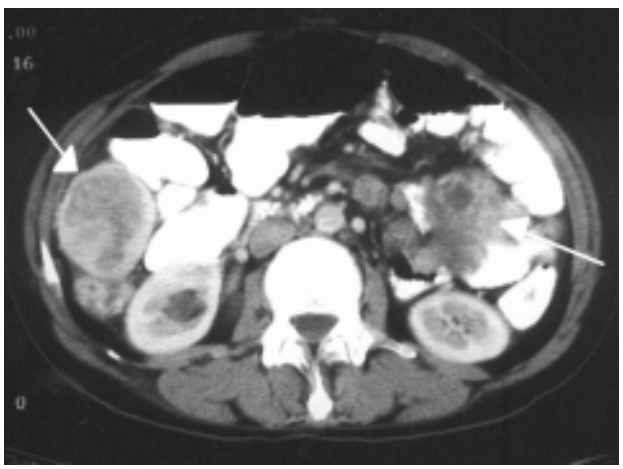

Figure 1. CT-scanning of the abdomen demonstrating multiple solid tumors with rich vascularity.

DISCUSSION

The incidence of malignant fibrous histiocytoma is more common in elderly patients in the seventh decade of life. Men are the most frequently affected. The lesion usually arises from the extremities (68%), or retro-peritoneum (16%) and often involves skeletal muscle (59%). This kind of tumors is usually locally invasive, but distant metastasis may the occur via the blood (30%) or lymphatics (12%). The risk of local recurrence and distant metastasis correlates best with the depth and the size of the primary tumor. The overall two-year survival rate is 60% and another 20% of patients will die or develop local recurrence subsequently.[3-5]

The location of this malignant tumor in the alimentary tract is extremely rare. Multi-focal malignant fibrous histiocytomas occur most commonly within bone, and only occasionally in other sites,[6] as in our patient. The two major clinical manifestations of gastrointestinal involvement are bleeding from ulcerated tumors and intussusception caused by polypoid tumors.[7] Our patient did not present any of the above-mentioned emergencies. However, although aggressive in behavior, its manifestations were of mild intensity. It is also important to consider that the occurrence of a metastasis from a MFH before discovery of the primary tumor, as in our patient, is quite rare[4]. In our case, it was impossible to distinguish a probable primary site of the lesion within the abdomen. Likewise, there was no primary location in the extremities or elsewhere, outside the peritoneal cavity.

Since MFH's first definition in 1964, many attempts have been made to describe its pathology and histogenesis. Based on its histopathologic features, MFH has been categorized into storiform-pleomorphic, myxoid, inflammatory, giant cell and angiomatoid variants. Among these types, the storiform-pleomorphic is the most common, comprising two thirds of the cases. It is composed of spindle-shaped cells arranged in short fascicles in a cartwheel or storiform pattern, along with plump histiocyte cells, mitotic features and multinucleated giant cells[8]. In our case, the histologic pleomorphism of the resected tumors, the degree of vimentin staining, the absence of the smooth muscle marker desmin, and in general the macroscopic and microscopic findings, favoured the diagnosis of malignant fibrous histiocytoma.

Surgical resection is the treatment of choice but depends upon the stage of the disease and the size of depth invasion by the tumor. Radiotherapy, chemotherapy and immuno-therapy are other therapeutic modalities.[8] Long-term follow-up with repeated chest X-ray and

CT-scanning of the abdomen for detection of local lymph node involvement is recommended.[9]

In this case we considered some form of adjuvant chemotherapy necessary, as the disease was extensive, with liver metastasis involved, and the resection was not curative.

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