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Diffuse intra-abdominal low grade fibromyxoid sarcoma with hepatic metastases: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Low grade fibromyxoid sarcoma (LGFMS)[1] is a rare soft tissue tumor involving deep soft tissues of the extremities and trunk. Abdominal location is extremely uncommon in which the few cases published in the literature are characterized by slow tumoral progression and long recurrence-free intervals.
METHODS: We report the first case of an intra-abdominal LGFMS which was discovered incidentally in a 42-year-old woman presenting diffuse peritoneal nodules and hepatic metastasis on CT and MRI scans.
RESULTS: The patient was successfully treated through conservative measures and remained asymptomatic at the 48 month follow-up.
CONCLUSIONS: This is the first reported case of LGFMS with peritoneal and hepatic metastases. Despite the discovery of an advance disease at exploration, the patient who refused a major surgical operation presents an uneventful follow-up and long term survival.

1. Introduction

Low grade fibromyxoid sarcoma (LGFMS) is a rare entity, first described by Evans in 1987 as a malignant tumor having a deceptively benign appearance [1–3]. It is most commonly found in the trunk and proximal extremities but can occur almost anywhere in the body [1,2]. There are very few cases of intra-abdominal LGFM reported in the literature, even fewer with diffuse peritoneal nodules [4–8] and only one case describes a hepatic metastasis in a patient with LGFMS of the extremities [3].

We present the first case of a 42-year-old woman with an incidental discovery of an intra-abdominal LGFMS with peritoneal and hepatic metastases. Diagnosis management and the current follow-up are subsequently discussed.

2. Presentation of case

Following an abdominoplasty in a 42-year-old female patient, resection of a fibro-adipose tissue protruding through a hernial defect revealed a LGFMS with positive resection margins.

The patient was completely asymptomatic and had no relevant medical or surgical history. The physical examination was positive for palpable mobile intra-abdominal masses and nodules in the Douglas pouch.

An abdominal computed tomography (CT), showed a suspicious lesion of 49 mm in segment VIII of the liver and multiple disseminated peritoneal nodules suggesting peritoneal carcinomatosis (PC) (Fig. 1). An abdominal MRI showed non-specific post-operative fatty stranding in the abdominal wall and confirmed the CT scan findings. A whole body 18F-FDG-PET/CT showed only a weak FDG uptake within some peritoneal nodules (Fig. 2).

A biopsy of the liver lesion was positive for liver metastasis of a LGFMS. Initially, the diagnosis has been made exclusively on morphologic criteria including an alternance of collagenized and myxoid areas (Fig. 3). Negative immunostaining for CD117 and KIT ruled out a gastro-intestinal stromal tumor. The FISH test analysis for the FUS gene rearrangement was non-contributory. Afterwards, immunostaining with specific markers including MUC4 and epithelial membrane antigen (EMA) have been performed. EMA was negative but MUC4 was strongly and diffusely expressed (Fig. 4). Serum CA-125 level was normal and serum IgE antibodies for

Abbreviations: LGFMS, low grade fibromyxoid sarcoma; CT, computed tomography; MRI, magnetic resonance imaging; PC, peritoneal carcinomatosis; EMA, epithelial membrane antigen; FISH, fluorescence in situ hybridization; PCI, peritoneal carcinomatosis index; HSCTGR, hyalinizing spindle cell tumor with giant rosettes; RT-PCR, reverse transcription polymerase chain reaction.

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[1] LGFMS low grade fibromyxoid sarcoma.

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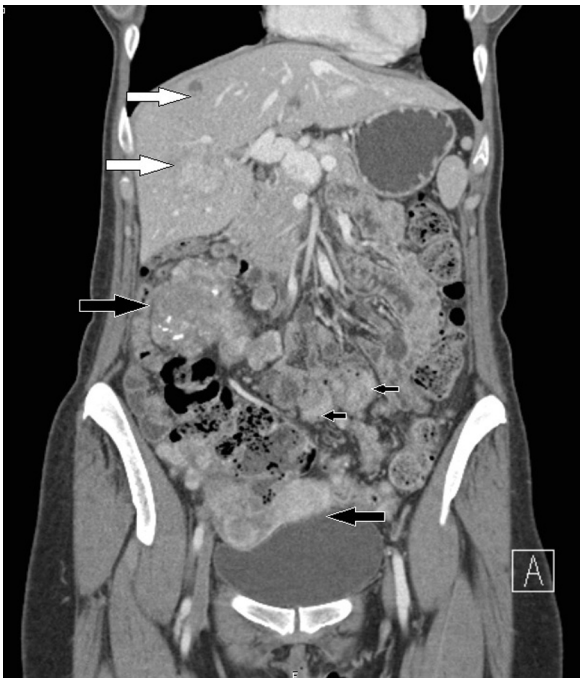

Fig. 1. Coronal contrast abdominal CT shows the hepatic metastatic lesions (white arrows) and multiple disseminated peritoneal nodules (black arrows).

Entameaba histolytica, *Entameaba granulosus* and *Schistosoma mansoni* were all negative.

We opted for an exploratory laparotomy in view of a debulking surgery and central partial hepatectomy. Peri-operatively, the peritoneal carcinomatosis index (PCI) was 25/39, indicating a more advanced peritoneal disease than had been shown on imagery. This would require a very aggressive surgery including: a large small bowel resection, a splenectomy, an omentectomy, a low anterior rectal resection, a partial cystectomy with a partial ureterectomy, a hysterectomy, a douglasectomy, as well as a partial hepatectomy and a protective ileostomy. Despite the fact that complete cytoreductive surgery could be achieved and that PCI score was not a contra-indication for surgery in presence of a low grade disease, we decided to abort surgery since patient agreed to surgery only under the condition that we avoid major imposition on her quality of life. Partial omentectomy was performed for histological examination. Intra-operative histopathological frozen examination of the omentum was positive for the LGFMS, confirmed by final examination.

Postoperatively, the patient confirmed her refusal of such an extensive surgery and also refused any adjuvant palliative treatment proposed after a multi-disciplinary discussion. The patient undergoes regular clinical and radiological follow-ups at 3 month intervals. At 48 months of diagnosis, she is still asymptomatic and in very good general health and the recent abdominal CT scan showed very discrete progression of the peritoneal carcinomatosis with no change at the hepatic and thoracic level.

3. Discussion

LGFMS is a rare type of soft-tissue sarcoma described for the first time in 1987 by Evans, characterized by bland histological features and, paradoxically, a high mitotic rate [1–3]. During the last 20 years the clinical and histopathological features have been delineated by several series in order to facilitate the diagnosis which was previously often misinterpreted [9].

Cytogenetically, LGFMS is characterized by a *t*(7;16)(q33; p11) or more rarely *t*(11;16)(p11; p11) translocation, resulting in FUS-

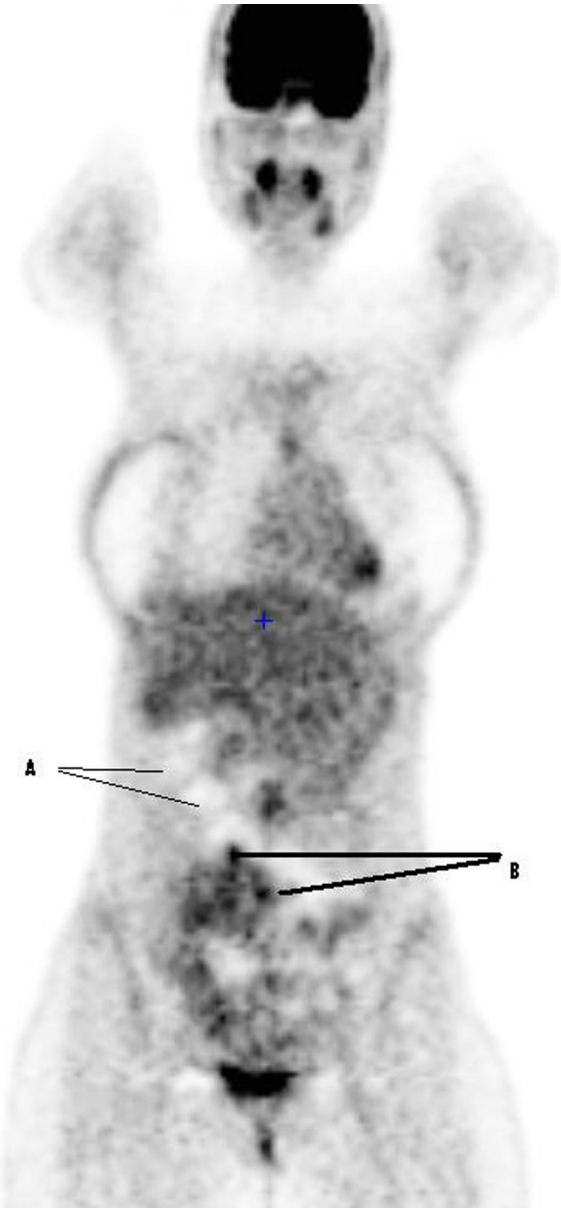

Fig. 2. Coronal Pet/CT image. A: nodules without metabolic activity; B: nodules with a weak hypermetabolic activity.

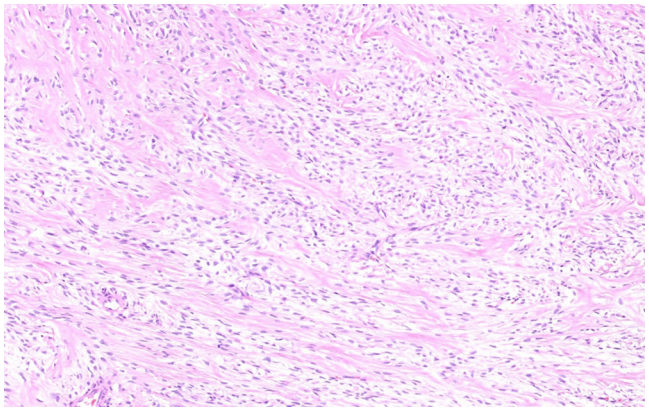

Fig. 3. Histopathological image of LGFMS characterized by fibrous and myxoid areas and swirling whorled growth patterns with low to moderate cellularity and bland cells with minimal nuclear pleomorphism.