

RARE FORM OF PRESENTATION IN A HISTIOCYTOMA CASE

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ABSTRACT

Malignant fibrous histiocytoma is also known as malignant fibrous xanthoma and is a form of neoplasm with uncertain histological origin. This tumour is rich in histiocytes and fibroblasts. MFH represent one of the most common type of soft tissue sarcoma but it has a low incidence in general population. We report the case of a 60 years old male patient, presented at the University Emergency Hospital Bucharest for a tumoral mass on the left hemithorax, in evolution for about 1 year, with an accelerated grow in the last 6 months. This case is typically because the patient is a male and he is 60 years old (range 50-70 years). In the same time, there are some particularities that make the case special and determine us to draw some conclusions about the management of this pathology. Histiocytoma represent a rare tumoral pathology and it is difficult to do a differential diagnostic preoperatively. Surgery is the cornerstone of treatment for all soft tissues. The goal of surgery is to eradicate disease in the affected area. All excisions of tumoral mass must be with resection margins for oncological security to avoid the possible complications.

Keywords: histiocytoma fibrous malignant, xanthoma, thorax

Note: All authors have contributed significantly and have been involved in the writing of the manuscript at draft and any revision stages, and have read and approved the final version.

INTRODUCTION

Malignant Fibrous Histiocytoma is also known as Malignant Fibrous Xanthoma and it is a malignant neoplasm with uncertain origin. In 1964, this tumor was first described like a specific soft tissue sarcoma type by O'Brien and Stout (1).

The frequency of soft tissue sarcomas in general population is down to 1% of all malignancies and the most common type of these is MFH. (2) It is identified at approximately 20-30% of cases. (3)

MFH is one of the most common type of soft tissue sarcoma. It occurs in adults (range 50-70 years) with a slight male predilection and with a M : F ratio of 1.2 :1 (4).

Although it can affect almost anywhere in the body, it has a predilection for extremities (up to 70% of patients) (5).

MFH manifests a broad range of histologic appearances with 5 sub-types described (6):

- Storiform-pleomorphic – is the most common type of sarcoma in patients older than

40 years, accounting for up to 70% of most cases;

- Myxoid – is the second most common, accounting for approximately 20% of cases;
- Giant cell – is a rare form;
- Inflammatory – tends to occur in retroperitoneum;
- Angiomatoid – usually occurs in young adults/adolescents.

The most common clinical presentation is as an enlarging painless soft-tissue mass, typically 5-10 cm diameter. Symptoms such as weight loss and fatigue are not typical but can be present at patients with advanced disease. Frequently, the biological status of patient is not affected. For paraclinical diagnostic, we can use ultrasonography of soft tissues, CT, IRM (7). An incisional biopsy or an excisional biopsy with a histopathologic evaluation on the tissue, is the method that provides the certain diagnostic.

Macroscopically, the tumor is typically large, well circumscribed but unencapsulated with a grey

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firm heterogeneous cut surface, sometimes with areas of hemorrhage and necrosis.

Microscopically, it is a heterogeneous fibroblastic tumor made up of poorly differentiated fibroblasts, myofibroblasts, histiocyte-like cells with storiform architecture and also demonstrate bizarre multi-nucleated giant cells with atypically mitosis.

Immunohistochemistry has a little value in the diagnosis of the MFH because the lesion hasn't any specific marker (6,8).

The prognosis depends on the following factors: tumour size, location (superficial is better), histological grade and the presence/absence of metastases.

The survival at 10 years is 90% in noncomplicated forms and in aggressive forms of MFH is only 20%. Regional lymph node involvement occurs in 0-15% of cases and distant metastases are found in 25-35% of cases. Local recurrence is present in 20-41% of cases (5,9,10).

CASE PRESENTATION

We present the case of a 60-year old male patient who came to the doctor because of a lump that had appeared on his left hemithorax about 1 year before. During the last 6 months prior to the presentation the lump started growing in an accelerated manner. Previous medical history and family history did not bring any relevant information regarding the incriminated pathology.

Clinical examination reveals a tumor-like formation on the left hemithorax with a diameter of about 10 cm, soft, motionless in relation to the underlying tissues, painless when palpated and having an intense erythematous area on the skin overlying the tumor (Fig. 1,2,3).



FIGURE 1. Tumor-like formation – lateral view



FIGURE 2. Tumor-like formation – posterior view



FIGURE 3. Tumor-like formation – anterior view

The complete biological exam shows a minimal inflammatory syndrome – VSH = 13 mm/1h (N: 3-10 mm), Fibrinogen = 460 mg/dL (N: 200-400 mg/dL) without any other pathological changes regarding the blood count, biochemistry and coagulogram.

Imaging examination – ultrasound revealed a subcutaneous soft tissue formation with a hypoechogenic ultrasound signal, with an inhomogeneous structure and measuring 14/11/9 cm. The

Doppler exam revealed a present intratumoral signal, which indicates the presence of vascularization inside the tumor.

The surgical intervention was then decided and performed with the purpose of tumor excision under general anesthesia with endotracheal intubation. The dissection of the tumor with an oncological safety border was performed intraoperative, along with a rigorous hemostasis. The resection piece was sent to be examined histopathologically. Samples of adjacent tissue were also excised in order to confirm the lack of invasion. In their case the histopathological exam did not identify any tumor cells (Fig. 4).



FIGURE 4. Resection piece – macroscopic aspect

The histopathological exam showed an encapsulated tumor-like formation, having 15/11/8 cm in dimensions, with a polycyclic contour and a smooth outer surface. When sectioned the surface presented a relatively homogenous aspect, white/grey/pink, with a multinodular character and high in consistency. The final diagnostic was: pleomorphic malign fibrous histiocytoma (Fig. 5, 6).

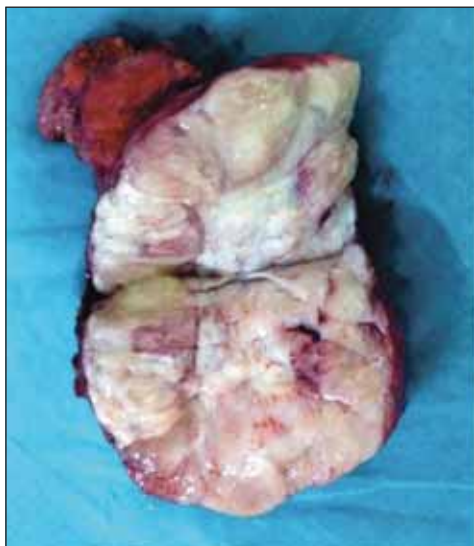


FIGURE 5. Tumor-like formation – macroscopic aspect – sagittal section

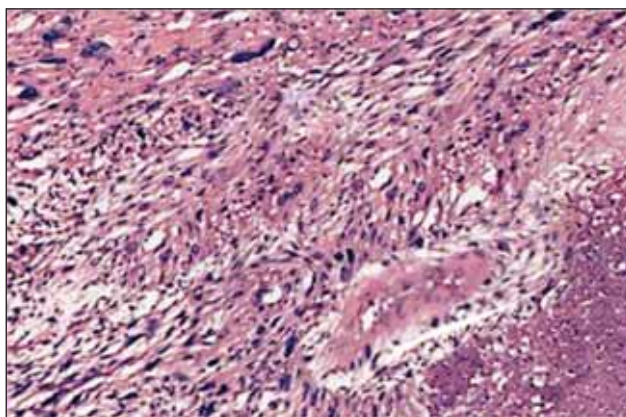


FIGURE 6. Resection piece – Microscopic aspect – tumor proliferation which consists of cells with an intense nuclear pleomorphism with a storiform and fascicular pattern

The postoperative evolution was favorable, the patient being released as surgically cured. The next follow-ups after 6 months, 1 year and 3 years did not detect any local relapses or other complications.

DISCUSSIONS

This case is typically because the patient is a male and he is 60 years old (range 50-70 years). In the same time, there are some particularities that make the case special and determine us to draw some conclusions about the management of this pathology.

Most cases of malignant fibrous histiocytoma under literature cited present a slow growth of the tumor mass for more than 2 years and they rarely arrived at up to 10 cm diameter. This patient presents a fast growth of the mass for about 1 year, with an accelerated growth in the last 6 months. These aspects did the presentation in our service to be late, the tumor size was 15 cm.

Frequent, this histological type-pleomorphic has a predilection for extremities -up to 70% of cases.

The location of this malignant fibrous histiocytoma on the thorax is extremely rare and it is very difficult to do a differential diagnostic preoperatively. In many situations this tumor location is a local complication of radiotherapy for other types of cancer. Our patient has no history of other disease.

Location and big dimensions of the tumor mass were the reasons which determine the surgeon to excision with resection margins for oncological security without an anterior needle biopsy. The differential diagnostic of this pathology includes benign tumors, we must know that there is a big probability to be a malignant neoplasm.

The excision of any mass tumor for which we do not have a certain diagnostic we should think of a malignant tumor. The goal surgery is to remove all tumor with negative margins.

In literature, up to 50% of MFH present an aggressive biological behavior and poor prognosis with local recurrence of the tumor and metastases in the next 2 years postop. In our patient situation the evolution was good without any complication for 3 years postop.

CONCLUSIONS

Histiocytoma represent a rare pathology for a general surgeon.

The location of this malignant fibrous histiocytoma on the thorax is extremely rare and it is very difficult to do a differential diagnostic preoperatively.

The goal of surgery is to eradicate disease in the affected area. All excisions of tumoral mass must be with resection margins for oncological security to avoid the possible complications.

Immunohistochemistry has a little value in the diagnosis of malignant fibrous histiocytoma because the lesion has not any specific marker.

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