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Case Report

Myxoinflammatory fibroblastic sarcoma in a rare location: breast and popliteal fossa

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Abstract: Myxoinflammatory fibroblastic sarcoma is an uncommon soft tissue tumor of low malignant potential. Two cases were discussed there. One is from a 31 years old male with painless solid mass and the other one is from a 38 years old female with painful lump. Tumors in these two cases occurred in chest wall and popliteal fossa respectively, which were measured from 8.5 to 9 cm in diameter progressively expanding occasionally accompanied by acid feeling. During the follow-up period (60 to 72 months), one patient had in situ recurrence. Histological examination in the tumor has lobulated structure, with myxoid change area, transparent sample area and the inflammatory area. Cells in the tumor are similar to R-S cells, virus samples with inclusions of different cells, there are fake adipocytes in some of the Myxoid areas. Immunohistochemical markers such as vimentin (2/2), SMA (1/2), CD68 (1/2), can also be found in those areas; but S100 (0/2), desmin (0/2), CD34 (0/2), EMA (0/2), caldesmon (0/2), ALK (0/2), myoD1 (0/2), myogenin (0/2) usually can not; and Ki-67 is both about 10%. These two cases demonstrate that MIFS is a rare soft tissue neoplasm, which can grow in the breast and popliteal fossa is much more scant; various misleading morphological features should been taken into consideration in the diagnosis.

Keywords: Myxoinflammatory fibroblastic sarcoma, chest wall and popliteal fossa, soft tissue neoplasm

Introduction

Myxoinflammatory Fibroblastic sarcoma, first described by Meis Kindblom, is a rare Mesenchymal neoplasm of low malignant potential which is seldom recurrenced or metastasis [1] in 1998. Then Montgomery [2] and Michal [3] described a group of similar lesions, named respectively infalmmatory myxohyaline tumor of distal extremities with virocyte or Reed-Sternberg-Like cells and inflammatory myxoid tumor of the soft parts with bizarre giant cells. After that, it was proved that the three kinds of descriptions belong to a same entity. The tumor mainly occurs in young people (aged 40-50), no significant gender differences, with the predilection of locations such as the hand, wrist or ankle. The tumor can also involve non-distal extremities including the Proximal limb, such as upper arm andthigh or other locations. Therefore the WHO classification deleted "acra" two words [4]. Here, we added two cases of MIFS with uncommon ocation to discuss the clinicopathologic, immunohistochemical, and differential diagnosis.

Materials and methods

Two cases were derived from department of pathology of the first affiliated hospital of Zhengzhou University. The surgical specimen were fixed in 10% buffered formalin, embedded routinely in paraffin and then stained with hematoxylin and eosin. Immunohistochemical use ready-to-use anti-bodies. The antibodies included Vimentin, SMA, CD68, S-100, Desmin, CD34, EMA, Caldesmon, ALK and Ki-67. Both the used antibodies and immunohistochemical staining kits were purchased from Roche co., LTD, and a control was regularly set up.

Clinical history

One of the two cases is a man who is 31 years old finding lumps for three years in right breast, which gives a prominent bulge beneath the affected skin, and imaging studies also suggest that the right side of the chest of subcutaneous displaying a 9×5×3 cm soft tissue lesion with an uniform density and relatively fine demarcation Patients undergone a stable condition after

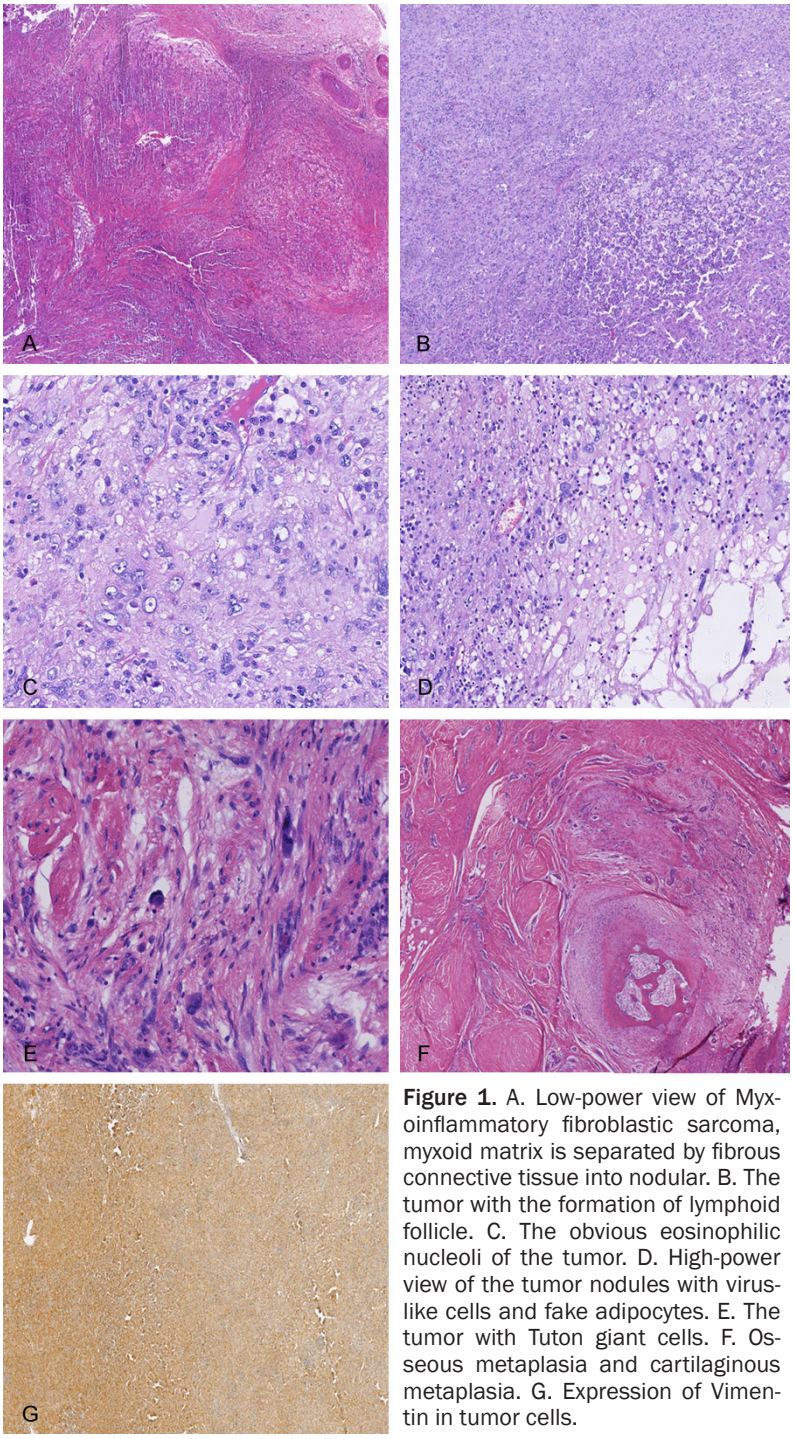

Figure 1. A. Low-power view of Myxoinflammatory fibroblastic sarcoma, myxoid matrix is separated by fibrous connective tissue into nodular. B. The tumor with the formation of lymphoid follicle. C. The obvious eosinophilic nucleoli of the tumor. D. High-power view of the tumor nodules with virus-like cells and fake adipocytes. E. The tumor with Tuton giant cells. F. Osseous metaplasia and cartilaginous metaplasia. G. Expression of Vimentin in tumor cells.

a complete resection surgery with no recurrence or metastasis for 72 months. Another is a female of 38 years old, who found the popliteal fossa mass seven years and encountered a recurrence with increasing size of the tumor feeling significant swelling pain within a year after a surgery. But after a second completely expanded resection before 60 months, she is in stable condition now, with no evidence of recurrence or metastasis.

Results

Grossly, tumors in the two cases talked about measuring 9*5*2 cm and 8.5*6.5*3.8 cm respectively, with a clear margin, and a texture of tenacity with some loose areas. Histologically, fibrous connective tissue and myxoid areas alternate with a lobulated structure, together with a large number of lymphocytes and a small amount of neutrophils, eosinophils infiltrating (**Figure 1A**), and the formation of lymph-oid follicle is appreciated in some areas (**Figure 1B**). The proliferated fibroblasts is ovoid, polygonal, or fat spindle with abundant cytoplasm and the obvious eosinophilic nucleoli (**Figure 1C**). A large number of virus samples or ganglion cells tumor giant cells and fake adipocytes can be seen (**Figure 1D**) as well as some tuton giant cells in some areas (**Figure 1E**). The mitotic activity of tumor cells was low (<1/HPF) and no necrosis was seen. But one of two samples in female presented with the blood vessels of cystic expansion with a small amount of protein fluid exudation and osseous nd cartilaginous metaplasia (**Figure 1F**). The tumor cells were positive for Vimentin (**Figure 1G**), SMA, CD68; but were negetive for S-100, Desmin, CD34, EMA, Caldesmon, ALK, MyoD1, Myogenin; meanwhile proliferative index Ki-67 was approximately 10%.

Discussion

Myxoinflammatory Fibroblastic sarcoma is a low-grade malignant soft tissue tumors with

local recurrence rate about 67% [4], rare distant metastasis. There are no age and gender differences, and about 10% of the reported cases occurred in children under the age of 12, or the elderly over the age of 75 [5]. In the beginning, MIFS was considered to only occur in the extremities, called a accompanied by virus cells and R-S sample cell tumors of the extremities of inflammatory mucous glass samples, namely, acra myxoinflammatory fibroblastic sarcoma (AMIFS) [6]. As Jurcic [7] and others reported that occurs in the proximal limb soft tissue MIFS cases, a growing number of studies found that MIFS not only occurs in limb extremities, but also can occur in the upper arms, thighs and other parts. According to statistics, about 60% involves the hands and wrists; about 30% involves the foot and ankle; occasionally the locations such as elbow, hip and knee can also be affected [1, 2, 7-10]. The two cases we providing occurr in the chest wall and popliteal fossa are much more rare.

Clinically, it usually shows a painless subcutaneous tumor with slowly growing, occasionally accompaning a pain or itching feeling. Given the location, it is often misdiagnosed as effusion or tenosynovitis, tendon sheath cyst, etc. Because of the misdiagnosis, it's easy to cause the in-situ recurrence after a incomplete surgical resection. Therefore, correct diagnosis for the effect of the treatment and prognosis is of great help. In general, the tumor often occurred in subcutaneous fascia, sometimes involving bone or joint, skin is generally not affected. Surgery should be expanded resection, and ensure the cut edge is negative. Histologically, the tumor mainly has three characteristics: myxoid change area, hyaline stroma and inflammatory exudation region. Fibrous connective tissue staggered with myxoid areas or hyaline areas forming nodular or lobulated pattern. Dispersed distinctive pleomorphic neoplastic cells can mimic lipoblastoma cells Hodgkin cells, or ganglion cells Inflammatory infiltrates are mainly composed of lymphocytes and sometimes lymphoid follicles can be seen. Visible hemosiderin deposition and even pigmented villondular synovitis (PVS) were identified. The morphology of our cases share the similar pathologic characteristics, however, one of case developing in the popliteal fossa demonstrate a cartilaginous and osseous metaplasia.

Immunohistochemistry, the vast majority of tumor cells showed Vimentin positive, histiocytic stain for CD68 and the fibroblastic cells is positive for SMA, but absence of S-100, Desmin, CD34, EMA, Caldesmon, Myo-D1, Myogenin and ALK, expression of KI-67 about 10%. The vast majority of the tumor cells show a consistent immunophenotype, but it's also not reliable, because of the lack of specific chromosomal hallmarks. Because in our cases can see many virus-sample cells, so we have to perform an EB virus and fungus related laboratory screening, which showed both the microbial special dyeing and EBER are negative so that the EB virus infection, as well as other infectious disease was ruled out [11]. Some relevant researches displayed the ultrastructure of abnormal tumor cell containing abundant rough endoplasmic reticulum and intermediate filaments, suggesting the nature of fibroblasts [1, 7].

When the significant hemosiderin deposition and multinucleated tumor giant cells appear, tenosynovial giant cell tumor should be excluded. However, The multinucleated giant cells of tenosynovial giant cell tumor are positive for CD68 for its synovial tissue origin. While the giant cells of MIFS derived from fibroblasts. Additionally, when a large number of fibroblast proliferations, it's necessary to have a differential diagnosis with inflammatory myofibroblastic tumor which mainly involve abdominal cavity, rarely occurring in Limbs acra. Sometimes, when fake adipocytes are scattered in the prominent myxoid matrix, myxoid liposarcoma needs to come into consideration. The obvious proliferated branched vasculature and typical lipoblasts with *CHOP* breakup make the differential diagnosis straightforward.

Anyway, MIFS is a rare and fibroblast-s and myofibroblast-derived malignant tumor with a relatively indolent course, which mainly develops in the distal extremities, but also in other uncommon locations such the chest wall and popliteal fossa as we report. Extensive surgical resection is the main treatment method. Recognizing this rare entity would be good help for the correct diagnosis and treatment of the patients in the clinical practices.

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Disclosure of conflict of interest

None.

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