**Case Report**

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

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Received July 21, 2016; Accepted September 21, 2016; Epub November 1, 2016; Published November 15, 2016

**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

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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 negative for S-100, Desmin, CD34, EMA, Caldesmon, ALK, MyoD1, Myoglobin; meanwhile proliferative index Ki-67 was approximately 10%.

Discussion

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

local recurrence rate about 67% [4], rare distan
t 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 sam-
pies, namely, acra myxoinflammatory fibroblas-
tic sarcoma (AMIFS) [6]. As Jurcic [7] and others
reported that occurs in the proximal limb soft
tissue MIFS cases, a growing number of stud-
ies found that MIFS not only occurs in limb
extremities, but also can occur in the upper
arms, thighs and other parts. According to sta-
tistics, 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 occur in the chest wall
and popliteal fossa are much more rare.

Clinically, it usually shows a painless subcuta-
neous tumor with slowly growing, occasionally
accompanying a pain or itching feeling. Given
the location, it is often misdiagnosed as effu-
sion or tenosynovitis, tendon sheath cyst, etc.
Because of the misdiagnosis, it’s easy to cause
the in-situ recurrence after an incomplete surgi-
cal 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 infl-
ammatory exudation region. Fibrous connec-
tive 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 lympho-
cytes and sometimes lymphoid follicles can
be seen. Visible hemosiderin deposition and
even pigmented villonodular synovitis (PVS)
were identified. The morphology of our cases
share the similar pathologic characteristics,
however, one of case developing in the popli-
teal fossa demonstrate a cartilaginous and
osseous metaplasia.

Immunohistochemistry, the vast majority of
tumor cells showed Vimentin positive, histio-
cytic 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 labora-
tory screening, which showed both the micro-
bial 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 ultrastruc-
ture of abnormal tumor cell containing abun-
dant rough endoplasmic reticulum and inter-
mediate 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 exclud-
ed. 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 differ-
ential diagnosis with inflammatory myofibro-
blastic tumor which mainly involve abdominal
cavity, rarely occurring in Limbs acra. Some-
times, when fake adipocytes are scattered in
the prominent myxoid matrix, myxoid liposar-
coma 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 devel-
ops 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.

Acknowledgements

This work was financially supported by the First
Affiliated Hospital of Zhengzhou University.
Disclosure of conflict of interest

None.

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References


