

Cytological diagnosis of myxoinflammatory fibroblastic sarcoma: A case report and discussion of differential diagnosis

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Abstract

Myxoinflammatory fibroblastic sarcoma previously known as inflammatory myxohyaline tumour of distal extremities with virocyte or Reed Sternberg like cells is a low grade tumour of the hands and feet. It is a distinctive lesion with features simulating inflammatory conditions, Hodgkin's disease and various sarcomas. It characteristically occurs in the distal extremities and has a propensity to recur locally. We describe cytological appearance of this condition with relevant differential diagnosis. A 51 year old male presented with a swelling over upper 1/3 of medial aspect of left tibia of 4 months duration. Fine needle aspiration smears showed myxoid material, epithelioid cells, spindle cells, vacuolated cells and giant cells along with scattered lymphocytes. Histopathological examination of the excision biopsy specimen was confirmatory.

Key words: Myxoinflammatory fibroblastic sarcoma, Sarcoma, Myxoid tumours, fineneedle aspiration cytology

Myxoinflammatory fibroblastic sarcoma is a distinct low-grade tumour of modified fibroblasts which characteristically occurs in the distal extremities and has a propensity for local recurrence⁽¹⁾. Most patients with this tumour are in the fourth and fifth decades of life. It affects males and females equally and patients present with a slowly growing painless, ill defined mass of the distal extremities. Fingers and hands are most commonly involved followed by toes, feet and lower leg². The tumour is multinodular and poorly circumscribed ranging from 1-8cm in size. They mostly arise in the subcutaneous tissue and may involve the dermis or skeletal muscle. Bone involvement is not known. Meis Kindblom et al¹ studied forty-four cases out of which twenty-four cases (67%) had at least one local recurrence. Only one patient had histologically proven lymph node metastasis. An overall good prognosis was seen and therefore it is essential to differentiate this from Hodgkins disease and other sarcomas. However it is also essential to differentiate this disease from inflammatory conditions. It is now known that this tumour can occur more proximally³ and the term acral has been dropped in the 2002 WHO classification⁴.

Case report

The patient was a 51 year old male soldier who presented with a complaint of swelling near the upper 1/3 of left tibia since 4 months. The patient was apparently well 4 months back. The swelling was small initially and increased in size presently to 5x3cms approximately. There was no pain or associated fever or cough. On examination his general condition was fair and he was afebrile.

A soft to cystic non-tender nodular growth was seen on the medial aspect of upper 1/3 of leg measuring 5x4 cms. Swelling was mobile and not attached to underlying bone. Skin appeared free. Other system examination was unremarkable. X- ray examination revealed a soft tissue swelling measuring 6x5cms. Tibia was normal. A fine needle aspiration was done.

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Cytologic findings

Smears were both alcohol fixed and air dried and were stained with Papanicolaou and May Grunwald-Giemsa stain. Smears were paucicellular and showed histiocyte like cells with vacuolated amphophilic cytoplasm with vesicular nuclei. A few vacuolated

lipoblast like cells, giant cells and cohesive cell clusters were also seen along with scattered lymphocytes. The background showed abundant myxohyaline material (fig1, fig 2). A diagnosis of myxoinflammatory fibroblastic sarcoma was made.

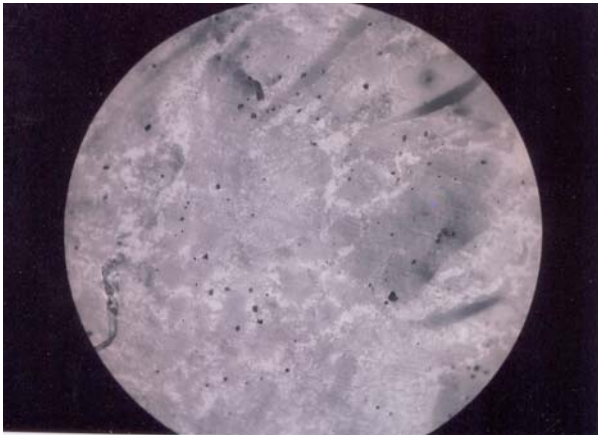


Fig 1: Cytology smear- Myxohyaline material and inflammatory cells MGG stain – 10 x magnification

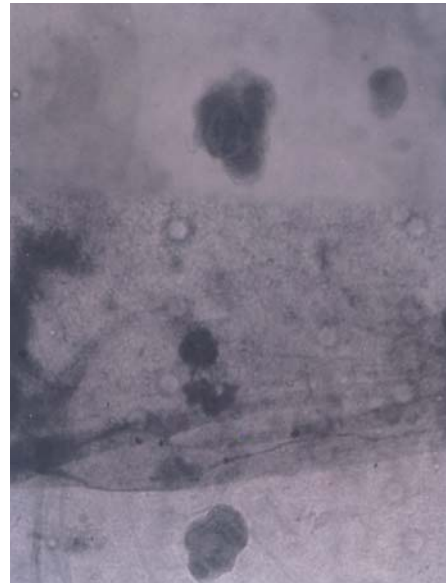


Fig 2: Cytology smear- Giant cells against hyalinised background MGG stain- 40x magnification

Discussion

Myxoinflammatory fibroblastic sarcoma is a recently documented low grade fibrosarcoma occurring mainly in digits, wrist and lower extremities. It arises in the subcutis and bone involvement is unknown. This tumour was first reported by Montgomery et al⁵ in 1997 where they noticed both inflammatory and neoplastic process of the tumour with its characteristic predilection for distal extremities. Therefore they coined the term “Inflammatory myxohyaline tumour of distal extremities with virocyte or Reed-Sternberg like cells.” They noticed local recurrence in one fourth of the patients but metastatic potential was unknown. Meis- Kindblom and Kindblom¹ subsequently reported a series with one patient having metastatic disease. They coined the term “ Acral myxoinflammatory fibroblastic sarcoma”. Later it was found that this tumour can occur more proximally and then the term “Acral” was

dropped in the WHO classification⁴. Myxoinflammatory fibroblastic sarcomas form infiltrative multinodular masses characterized by dense inflammation with myxoid to collagenous stroma.

Due to the wide range of appearances of this tumour the differential diagnosis depends on the cellularity of the tumour as well as the relative amount of myxoid and hyaline stroma. When there is a prominent inflammatory background with virocyte like nuclei and necrosis, an inflammatory process is often considered. However, special stains for microbial organisms are invariably negative as also immunohistochemical stains for cytomegalovirus².

Cellular zone shows atypical cells ranging from plump spindled cells to histiocytoid or epithelioid cells. Myxoid zones contain multivacuolated lipoblasts like fibroblasts. The inflammatory zones have scattered bizarre cells with vesicular nuclei and prominent nucleoli and abundant focally vacuolated cytoplasm, reminiscent of Reed Sternberg cells or virocytes. Some of these cells contain phagocytosed neutrophils. Mitotic figures are less than 2/50 HPF⁴. Multinucleated giant cells are occasionally seen. There is in addition round mononuclear cells with a bland nucleus and a small amount of amphophilic cytoplasm.

The common differential diagnosis includes giant cell tumour of the tendon sheath, inflammatory myofibroblastic tumour, inflammatory fibrosarcoma, other benign and malignant myxoid lesions and Hodgkin's disease. Presence of the prominent inflammatory component and Touton like giant cells leads to a misdiagnosis of giant cell tumour or tendon sheath whereas recognition of scattered bizarre large cells helps in distinguishing these lesions. Inflammatory myofibroblastic tumour and inflammatory fibrosarcoma commonly occur in the abdomen or thorax in contrast to acral myxoinflammatory fibroblastic sarcoma (AMIFS) which occurs on extremities. Further cells of AMIFS are more bizarre and lack immunohistochemical features of myofibroblasts.

Benign myxoid lesions lack large bizarre cells as found in AMIFS. Distinction from myxoid malignant fibrous histiocytoma is extremely difficult. However focal areas of high grade pleomorphic storiform malignant fibrous histiocytoma are often seen in myxoid malignant fibrous histiocytoma which is not found in AMIFS. Further there is alternating myxoid and hyalinized zones and striking inflammatory infiltrate in AMIFS. The possibilities of Hodgkin's disease can be considered only by the presence of Reed Sternberg like cells, however these cells are CD 15 & CD 30 negative.

Pohar Marinsek et al⁶ analysed cytomorphological characteristics of 3 fine needle aspiration biopsy (FNAB) samples of Acral myxoinflammatory fibroblastic sarcoma (AMIFS) as well as the features

of a number of other benign and malignant myxoid lesions. The analysis showed that FNAB samples from 2 cases of AMIFS had similar cytological features as described in surgical biopsies like myxoid material, spindle cells, epithelioid cells with globules or extra-cellular material and lipoblast like giant cells. Inflammatory cells were scarce. Samples from other myxoid tumours however lacked all the tumour components characteristic of AMIFS. They concluded that the cytological features of AMIFS may be characteristic enough to give a definitive diagnosis. Similarly in our case all the distinctive features of AMIFS was seen in the cytology smears and it was adequate enough to give a definitive diagnosis.

At present wide local excision appears to be adequate treatment for this tumour.

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