Cancer Association of South Africa (CANSA)

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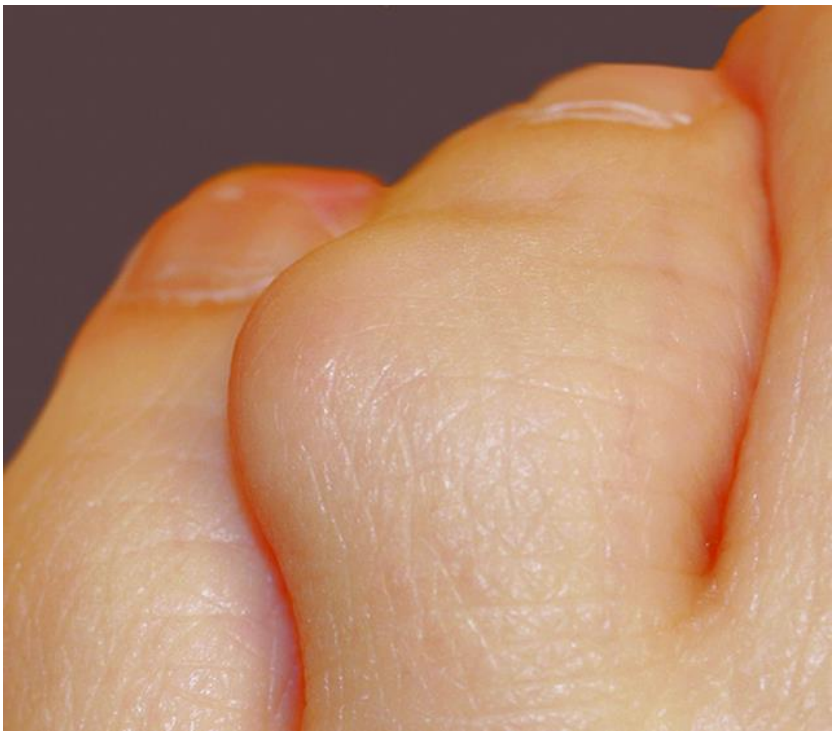
Fact Sheet
on
Myxoinflammatory Fibroblastic Sarcoma

Introduction

Myxoinflammatory Fibroblastic Sarcoma (MFS) is a soft tissue sarcoma. Like other soft tissue sarcomas, it arises in connective tissue - specifically, the connective tissue that surrounds muscles and separates muscles from each other and from skin. An MFS may lie just below the skin or deeper in muscle tissue.



[**Picture Credit**: Myxoinflammatory Fibroblastic Sarcoma of the toe]

MFS is not as well-known or understood as many other types of cancer, partly because the features distinguishing it from another soft tissue sarcoma came into focus only recently. Also, an MFS is often mistaken for a benign tumour and treated as such, by removing only the visible growth. This delays accurate diagnosis and complicates further treatment.

There is generally an equal sex distribution, with some small series showing a very slight male predilection

Myxoinflammatory Fibroblastic Sarcoma

Myxoinflammatory fibroblastic sarcoma is a low grade sarcoma that is composed of a mixed inflammatory infiltrate along with spindled, epithelioid and bizarre appearing cells in a background of hyaline and myxoid zones. Seen affecting the distal extremities commonly, with an equal sex predilection, these tumors are rare and require an extensive immunohistochemical work up for proper diagnosis. They have a tendency to recur.

Researched and Authored by Prof Michael C Herbst
[D Litt et Phil (Health Studies); D N Ed; M Art et Scien; B A Cur; Dip Occupational Health; Dip Genetic Counselling; Dip Audiometry and Noise Measurement; Diagnostic Radiographer; Medical Ethicist]
Approved by Ms Elize Joubert, Chief Executive Officer [BA Social Work (cum laude); MA Social Work]

Synonyms may include:

- Atypical Myxoinflammatory Fibroblastic Tumour
- Inflammatory Myxohyaline Tumour of the Distal Extremities with Virocyte or Reed-Sternberg-Like Cells
- Inflammatory Myxoid Tumour of the Soft Parts with Bizarre Giant Cells

Myxoinflammatory Fibroblastic Sarcoma (MIFS) is a rare, malignant, soft tissue tumour that is found to be locally aggressive. In a majority of cases, Myxoinflammatory Fibroblastic Sarcoma is found in middle-aged adults. MIFS is known to grow very slowly over several years that it is often mistakenly considered as a benign tumour. Commonly the tumour involves the hands and feet (mostly the fingers). The joints, muscles, and bones may also be affected resulting in associated signs and symptoms. Tumour recurrence is commonly noted. The prognosis of Myxoinflammatory Fibroblastic Sarcoma depends upon several factors including the size and stage of the tumour. Small-sized tumours that are detected early and can be completely removed have a good prognosis

It mainly affects middle-aged persons, and is usually located in the extremities (fingers and hands, for 75% of cases). Clinically, the lesion is infiltrating, pain-free and of slow evolution.

Differential diagnosis:

- Epithelioid Sarcoma
- Hodgkin Lymphoma
- Myxofibrosarcoma
- Pleomoarphic Liposarcoma
- Rosai-Dorfman Disease
- Tenosynovitis

Lucas, D.R. 2017.
"Myxoinflammatory fibroblastic sarcoma is a rare soft tissue tumor with most occurring in the distal extremities of adult patients. It has a high rate of local recurrence and a low rate of metastasis. Because it may appear benign on clinical examination, and because the microscopic features are generally underrecognized, it is often inadequately treated and misdiagnosed. In this review, based upon experience and that of the literature, the intent is to highlight salient clinicopathologic features, detail the broad microscopic spectrum including high-grade aggressive variants, review the molecular features, and discuss its relation to hemosiderotic fibrolipomatous tumor."

Srivastava, P., Husain, N., Nevaz, A. & Gupta, V. 2018.
"Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare soft tissue sarcoma which was initially observed in acral sites and characterised by spindle cells, pleomorphic bizarre cells and distinctive large Reed-Sternberg-like cells admixed with an intense inflammatory cell infiltrates. MIFS manifests as a slow growing often superficial lesion which can be mistaken as infectious or chronic inflammatory process or benign tumours such as nodular fasciitis, giant cell tumour of tendon sheath or synovial pseudocyst. We report a rare presentation of a MIFS in a 38-year-old man with extensive local spread from subcutaneous tissue to the ankle joint and bones as well as multiple synchronous metastases to lung, sixth rib and vertebra. Our case is peculiar for its aggressive clinical behaviour with short duration, fast growth and extensive metastases, a feature infrequent in MIFS."

Incidence of Myxoinflammatory Fibroblastic Sarcoma

The National Cancer Registry does not provide any information on the incidence of Myxoinflammatory Fibroblastic Sarcoma in South Africa.

Signs and Symptoms of Myxoinflammatory Fibroblastic Sarcoma

Signs and symptoms may include:

- Painless swelling of affected area
- Swollen area may be nodular in appearance
- Usually cutaneous lesions appearing a plaques

Diagnosis of Myxoinflammatory Fibroblastic Sarcoma

Myxoinflammatory Fibroblastic Sarcomas are low-grade sarcomas with a protracted clinical course, a high rate of local recurrence and a low rate of metastasis. They are rare, painless, low-grade neoplasm which commonly occurs in the extremities. It has a distinctive morphology and can be a diagnostic challenge, simulating inflammatory conditions as well as neoplastic conditions.

An MRI scan may be helpful in reaching a diagnosis.

Ozaki, S., Kasashima, S., Kawashima, A. & Ooi, A. 2018.

"Myxoinflammatory fibroblastic sarcoma (MIFS) is rare low-grade soft-tissue tumor that occurs in extremities. Clinically it is difficult to differentiate from benign lesions, such as nodular fasciitis, and malignant tumors, such as liposarcoma. We report a case of MIFS in the forearm of a 34-year-old man. The resected tumor measured 5.3 × 2.7 × 2.5 cm^3, had a lobular structure with indistinct boundary, and consisted of a large amount of translucent and yellow mucous-like substrate. Cytological examination of a preoperative puncture aspiration specimen showed histiocyte- and fibroblast-like tumor cells in a mucous-like matrix together with scattered lipoblast- and ganglion-like cells. Nuclear chromatin of the tumor cells showed a fine granular appearance but no notable hyperchromasia. There were no clear malignant findings. On Giemsa staining of the tumor cells, there were fine granular and hyaline intracytoplasmic substances that showed purple to reddish-purple metachromaticity. Prominent inflammatory cells were not observed in the specimens. MIFS should be considered in the differential diagnosis for a myxoid tumor in the extremities."

Kumar, R., Lefkowitz, R.A. & Neto, A.D. 2017.

OBJECTIVE: This retrospective study was undertaken to highlight clinical and magnetic resonance imaging features of myxoinflammatory fibroblastic sarcoma.

METHODS: The clinical records of 29 patients (14 men and 15 women) were analyzed.

RESULTS: The soft tissue tumors involved hand, wrist, foot and ankle in 21 patients, more proximal extremities in 7 patients, and neck in 1 patient. The tumors were subcutaneous in 11 patients, and intramuscular in 15 patients. On magnetic resonance imaging, these tumors had nonspecific features suggestive of benign and malignant lesions. An unusual tumor in hand indicated infection, whereas an intraarticular knee tumor mimicked pigmented villonodular synovitis. All tumors were surgically resected.

CONCLUSIONS: Myxoinflammatory fibroblastic sarcoma, a rare low-grade subcutaneous soft tissue tumor of distal extremities with high local recurrence after resection can mimic several benign and malignant lesions on histopathology and imaging. Rarely, it can arise in muscles and tendons, occur in nonacral sites, be

aggressive, and even metastasize. In most cases, surgical resection with wide margins can be curative with low local recurrence.

Gaetke-Udager, K., Yablon, C.M., Lucas, D.R. & Morag, Y. 2016. Myxoinflammatory fibroblastic sarcoma: spectrum of disease and imaging presentation. *Skeletal Radiol*. 2016 Mar;45(3):347-56. doi: 10.1007/s00256-015-2286-2. Epub 2015 Nov 12.

OBJECTIVES: To describe the imaging findings of a series of myxoinflammatory fibroblastic sarcomas (MFSs) from our institution, including a case of dedifferentiated MFS and two cases with areas of high-grade tumor, in addition to typical cases of low-grade tumor. To correlate the imaging findings with the pathologic features of these tumors.

SUBJECTS AND METHODS: IRB approval was obtained. Retrospective search of the pathology database at our institution from 2000 to 2015 identified seven cases of MFS with available imaging. Imaging, pathology, and clinical data were reviewed.

RESULTS: Unlike the majority of well-differentiated tumors in our series (four cases), one tumor showed dedifferentiation and two cases had areas of high-grade tumor. The dedifferentiated tumor showed peripheral post-contrast enhancement. One case with a substantial high-grade component showed osseous destruction and peripheral enhancement in the high-grade area, while the low-grade component enhanced diffusely. The second case had a small high-grade area and showed diffuse enhancement. All three of these cases had non-acral locations and lacked association with a tendon. The four cases of low-grade MFS demonstrated diffuse enhancement, were located in the distal extremities, and were associated with a tendon.

CONCLUSION: The imaging findings of dedifferentiated and high-grade MFS differ from the more typical low-grade tumors in that they have nonenhancing areas, a non-acral location, lack association with a tendon, and may involve bone. The radiologist should be aware that MFS represents a spectrum that includes low-grade tumors, tumors with high-grade areas, and tumors with dedifferentiation and that this spectrum presents with differing imaging features.

Kato,M., Tanaka, T. & Ohno, T. 2015. Myxoinflammatory fibroblastic sarcoma: a radiographical, pathological, and immunohistochemical report of rare malignancy. *Case Rep Orthop*. 2015;2015:620923. doi: 10.1155/2015/620923. Epub 2015 May 18.

"Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare, painless, and intermediate (rarely metastasizing) fibroblastic tumor, which commonly occurs in the extremities, with an equal sex predilection. This sarcoma is composed of a mixed inflammatory infiltrate along with spindled, epithelioid, and bizarre tumor cells in a background of hyaline and myxoid areas. In spite of such a distinctive morphology, the tumor can be a diagnostic challenge, simulating inflammatory conditions as well as neoplastic nature. For accurate diagnosis, the tumor requires extensive clinical, radiological, and pathological investigations. We present a case of MIFS in a 19-year-old female who presented with a mass in the left ankle. After appropriate excision and postoperative radiation therapy, she is free of disease, including recurrence and metastasis, at 12 years postoperatively."

Treatment of Myxoinflammatory Fibroblastic Sarcoma

Therapy may include:

- Excision of lesion
- Amputation
- Limited use of radiotherapy

- Limited use of chemotherapy.

Jagadesh, N., Miller, D.H., Schenk, W., Attia, S., Sherman, C.E., Cortese, C., May, B.C. & Miller, R.C. 2017. "Myxoinflammatory fibroblastic sarcoma is a rare sarcoma which typically presents in the extremities and is treated by definitive surgery. In recurrent disease, the reported utilization of radiotherapy is increasing, and more modern techniques such as intensity-modulated radiotherapy may be improving long-term outcomes."

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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