Title:

Myxoinflammatory fibroblastic sarcoma of the thumb: a case report at an unusual site with review of the literature.

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Conflict(s) of interest:

The authors declare there is no conflict of interest.

Background:

Myxoinflammatory fibroblastic sarcoma (MIFS) is a low-grade mesenchymal neoplasm, characterized by pleomorphic fibroblastic cells in a myxohyaline background with prominent inflammatory cell infiltrates.¹⁻⁴ MIFS typically presents as a painless subcutaneous mass in the distal extremities of patients in the fourth through sixth decades of life. MIFS of the thumb has rarely been reported in the literature and the etiology of it remains unknown.³ Here, we present a case of MIFS in the thumb of a middle-aged gentleman, highlighting the diagnostic features and reviewing the surgical management of this rare tumor.

Case:

A 50-year-old male presented with four weeks of progressive left thumb pain that worsened with grasp and pinching. The patient was otherwise healthy, denied trauma or previous surgery in the area, and family history was noncontributory. On physical examination, a palpable, subcutaneous mass was present at the volar flexor crease on the left thumb. Magnetic resonance imaging of the thumb demonstrated a loculated mass abutting the flexor tendon. The patient opted for surgical excision and was subsequently taken to the operating room. The mass was meticulously dissected while releasing and preserving the ulnar neurovascular bundle. Excisional biopsy revealed low-grade MIFS. At one year follow-up, our patient remained neurovascularly intact with good function. Clinical exam and postoperative imaging did not reveal any evidence of recurrence.

Discussion:

This case demonstrates that while MIFS commonly presents in the distal extremities, it is important for orthopedic surgeons to include MIFS in their differential diagnosis when presented with a painless mass in an atypical location like the thumb. When MIFS is suspected, we recommend a meticulous, margin-negative excision, while maintaining hemostasis and preserving surrounding neurovascular bundles. Finally, the use of adjuvant radiation therapy may be beneficial in patients with positive surgical margins to prevent local recurrence and distant metastasis.⁵

Images:



Figure 1. Physical examination showed a palpable, slightly indurated, subcutaneous mass at the volar flexor crease within the interphalangeal region of the left thumb.



Figure 2. Plain radiographs demonstrated soft tissue mass without bone erosion or reaction (top). Sagittal and axial magnetic resonance images of the left thumb demonstrated a loculated mass abutting the flexor tendon (bottom).



Figure 3. The surgical incision exposes a subcutaneous mass with overlying the flexor tendon sheath and enveloping the ulnar neurovascular bundle (**A**). A tan, fibrotic tumor mass was identified, measuring $2.5 \times 1.5 \text{ cm}$ (**B**).



Figure 4 (**A-F**). Hematoxylin and eosin stained sections demonstrate the histopathological features of MIFS characterized by mixed cellular features including 3 morphological zones: myxoid, inflammatory and hyalinized (**A** at low magnification 1.25x). Variable views of MIFS with heterogeneous cell component: Inflammation zone including extensive infiltration by neutrophils, lymphocytes, plasma cells and eosinophils (**B** at magnification 20x); myxoid zone with focal hyalinized stroma (**C** at magnification 20x); tumor cells show histiocytoid and epithelioid appearance (**D** at magnification 40x); admixed feature including plump tumor cells with focal marked nuclear atypia and smudgy heterochromatin (arrows) in an inflammatory background of rich in neutrophils (**E** at magnification 40x); a higher magnification view of some tumor cells with macronucleoli reminiscent of Hodgkin cells (arrows in **F** at 65x).

References:

- Gaetke-Udager K, Yablon CM, Lucas DR, Morag Y. 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. PMID: 26563559.
- Willems SM, Schrage YM, Baelde JJ, Briaire-de Bruijn I, Mohseny A, Sciot R, Bovée JV, Hogendoorn PC. Myxoid tumours of soft tissue: the so-called myxoid extracellular matrix is heterogeneous in composition. Histopathology. 2008 Mar;52(4):465-74. doi: 10.1111/j.1365-2559.2008.02967.x. PMID: 18315599.
- Montgomery EA, Devaney KO, Giordano TJ, Weiss SW. Inflammatory myxohyaline tumor of distal extremities with virocyte or Reed-Sternberg-like cells: a distinctive lesion with features simulating inflammatory conditions, Hodgkin's disease, and various sarcomas. Mod Pathol. 1998 Apr;11(4):384-91. PMID: 9578090.
- Nishio J, Iwasaki H, Nabeshima K, Naito M. Cytogenetics and molecular genetics of myxoid soft-tissue sarcomas. Genet Res Int. 2011;2011:497148. doi: 10.4061/2011/497148. Epub 2011 Jul 28. PMID: 22567356; PMCID: PMC3335514.
- Tejwani A, Kobayashi W, Chen YL, Rosenberg AE, Yoon S, Raskin KA, Rosenthal DI, Nielsen GP, Hornicek FJ, Delaney TF. Management of acral myxoinflammatory fibroblastic sarcoma. Cancer. 2010 Dec 15;116(24):5733-9. doi: 10.1002/cncr.25567. Epub 2010 Aug 24. PMID: 20737559.