

Juxta-articular myxoma of the wrist

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This case report describes a juxta-articular myxoma (JAM) in an uncommon location, demonstrating an unusually aggressive local growth pattern.

Introduction

Myxomas are benign mesenchymal tumours characterised by the production of mucinous material in a scant spindle cell matrix.¹ Juxta-articular myxoma (JAM) is a rare form of myxoma in the vicinity of a joint.² JAM has an association with trauma or joint arthrosis. It may be locally aggressive and commonly recurs after surgical excision.³

Case presentation

A 78-year-old man, with a history of colon carcinoma and gout, presented with a large mass in the thenar eminence and bilateral carpal tunnel syndrome. On clinical examination, the mass was fixed and had no inflammatory or infective features. Magnetic resonance imaging (MRI) was requested to assess the nature of the mass and exclude a neoplastic lesion.

Imaging findings

An extensive, multiseptated cystic mass was noted in the thenar eminence with infiltration around the flexor pollicis longis (FPL) and abductor pollicis brevis (APB) tendons. The mass had uniform fluid signal on proton density (PD) fat-saturated images (Figure 1 and Figure 2). Post-contrast images demonstrated enhancement of the septations with central fluid signal. Similar cystic infiltration was noted in and deep to the pronator quadratus (Figures 3b, 4, 5a and 5b).

Multiple osseous erosions involving the distal radius, the carpal bones and the metacarpal bases were observed. Prominent osseous erosion of the capitate and scaphoid was present with almost total destruction of the normal trabecular marrow. The erosions were PD hyperintense (Figure 1 and Figure 2), T1W hypointense (Figures 3a and Figure 6) and enhanced peripherally with almost uniform central fluid signal (Figures 3b, 4, 5a and 5b). The erosions were sharply margined and corticated and there was no associated bone marrow oedema.

Tenosynovitis was noted, involving all of the extensor and flexor compartments. The median nerve, compressed in the carpal tunnel, was oedematous and swollen proximal to the carpal tunnel (Figures 3b, 4, 5a and 5b). Prominent radiocarpal and intercarpal synovitis was present. Moderate arthrosis of the radiocarpal, first carpometocarpal (CMC) and scaphotrapeziotrapezoid (STT) joints. Periarticular fluid collections, synovitis and enhancing erosions were noted in relation to the metacarpophalangeal (MCP) joints.

A diagnosis of probable gout was made, with a differential diagnosis of polyarticular inflammatory arthritis. Metastatic disease was thought unlikely.

Surgical findings

At surgery, a complex, infiltrating, locally aggressive cystic mass was confirmed (Figure 7). The thenar mass was debulked, the carpal tunnel was released and a biopsy of the synovium of the radiocarpal joint was done.

Histopathology

Histology and immunohistological staining revealed a mucin-producing mass with scant spindle cells expressing smooth muscle actin and vimentin. No features of autoimmune synovitis were present and no crystals were found. S100 and desmin staining was negative. A pathological diagnosis of JAM was made.

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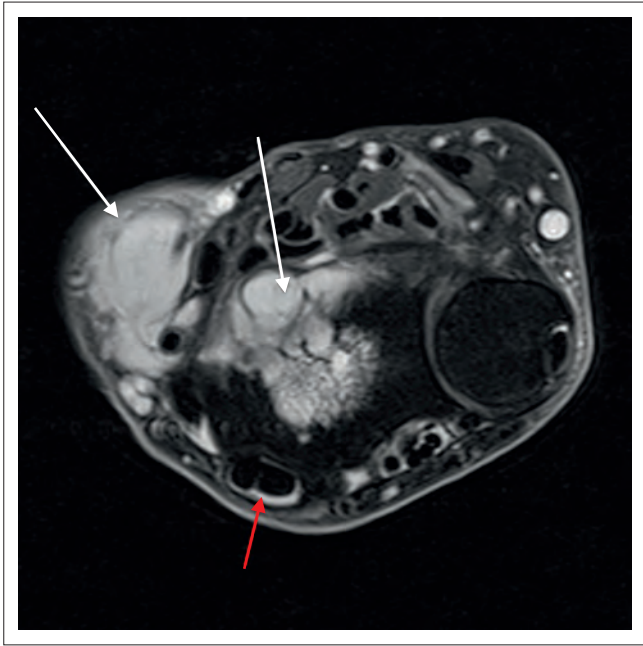


FIGURE 1: Axial PD fat-saturated image shows a cystic mass in the thenar eminence extending into the pronator quadratus and the radial epiphysis (white arrows). There is widespread flexor and extensor tenosynovitis (red arrow).

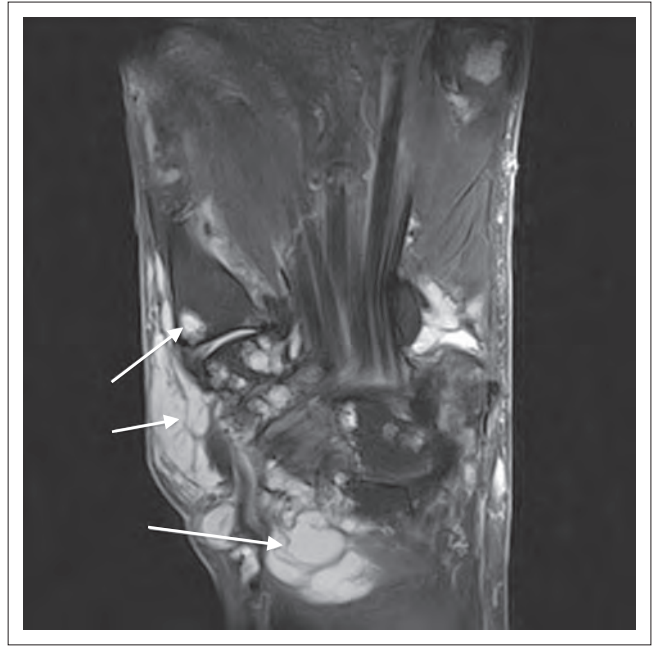


FIGURE 2: Coronal PD-weighted fat-saturated image shows a large, well-circumscribed cystic mass in the thenar eminence with extensive infiltration into the adjacent carpal bones, distal radius and pronator musculature (white arrows).



FIGURE 3: Sagittal T1 fast spin echo (FSE)-weighted images precontrast (a) and post contrast (b) demonstrate the pronator quadratus component and the intraosseous erosions in the radius, carpal bones and bases of the metacarpal bones. There is peripheral enhancement of the pronator mass, and intense solid and peripheral enhancement of the osseous erosions (white arrows).

Clinical course

The postoperative course was initially unremarkable; however, the patient returned four months later complaining of an increase in the size of the lesion,

with decreasing left hand function. A repeat MRI was performed prior to further surgery, as dramatic clinical deterioration suggested a possible infectious or malignant diagnosis.

Follow-up MRI findings

The MRI showed recurrence of the thenar eminence JAM, marked progression of the erosive intraosseous components, prominent flexor and extensor tenosynovitis, and radiocarpal and intercarpal synovitis (Figure 8).

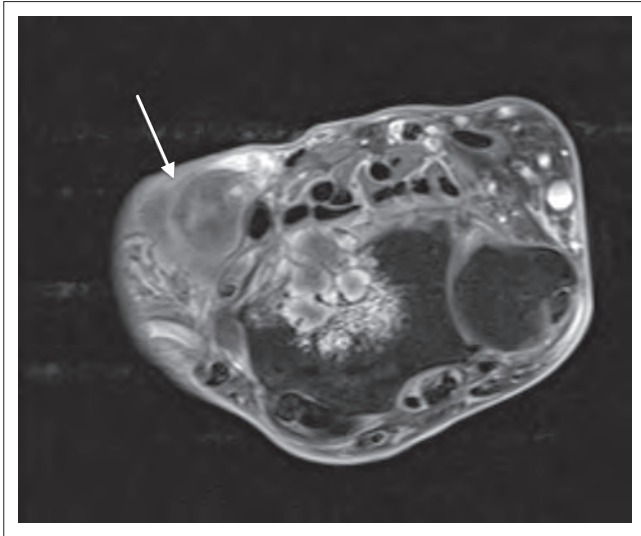


FIGURE 4: Axial T1-weighted fat-saturated image post intravenous gadolinium shows intense, predominantly peripheral, enhancement with central low T1 signal in the thenar mass (white arrow) and the radial erosion.

Discussion

JAM is most commonly associated with large joints, with a male preponderance. It is mostly found in the lower limbs, and is very unusual in the hand and wrist.⁵ JAM involves periarticular tissues including subcutaneous fat, joint capsule, tendons and rarely skeletal muscle.

The aetiology of the lesion is uncertain; however, there is an association with previous trauma and osteoarthritis of the underlying joint.^{1,6} The lesion itself is usually painless, but it may cause local mechanical symptoms. JAMs do not metastasise; however, as in this case, they can be very locally aggressive. Postoperative recurrence is seen in approximately 30% of JAMs within five years of excision.^{3,4} Other types of myxoma are intramuscular, cutaneous aggressive angiomyxoma, and myxoma of the nerve sheath (neurothekeoma).³

Intramuscular myxoma is the most commonly encountered myxoma in clinical practice, occurring in the thigh, upper arm and buttock.⁵ The lesion is characteristically a well-circumscribed oval mass that is cystic in appearance. Intramuscular myxomas are usually solitary but may be multiple and, in this scenario, are usually associated with monostotic or polyostotic fibrous dysplasia, which is known as Mazabraud's syndrome.⁵

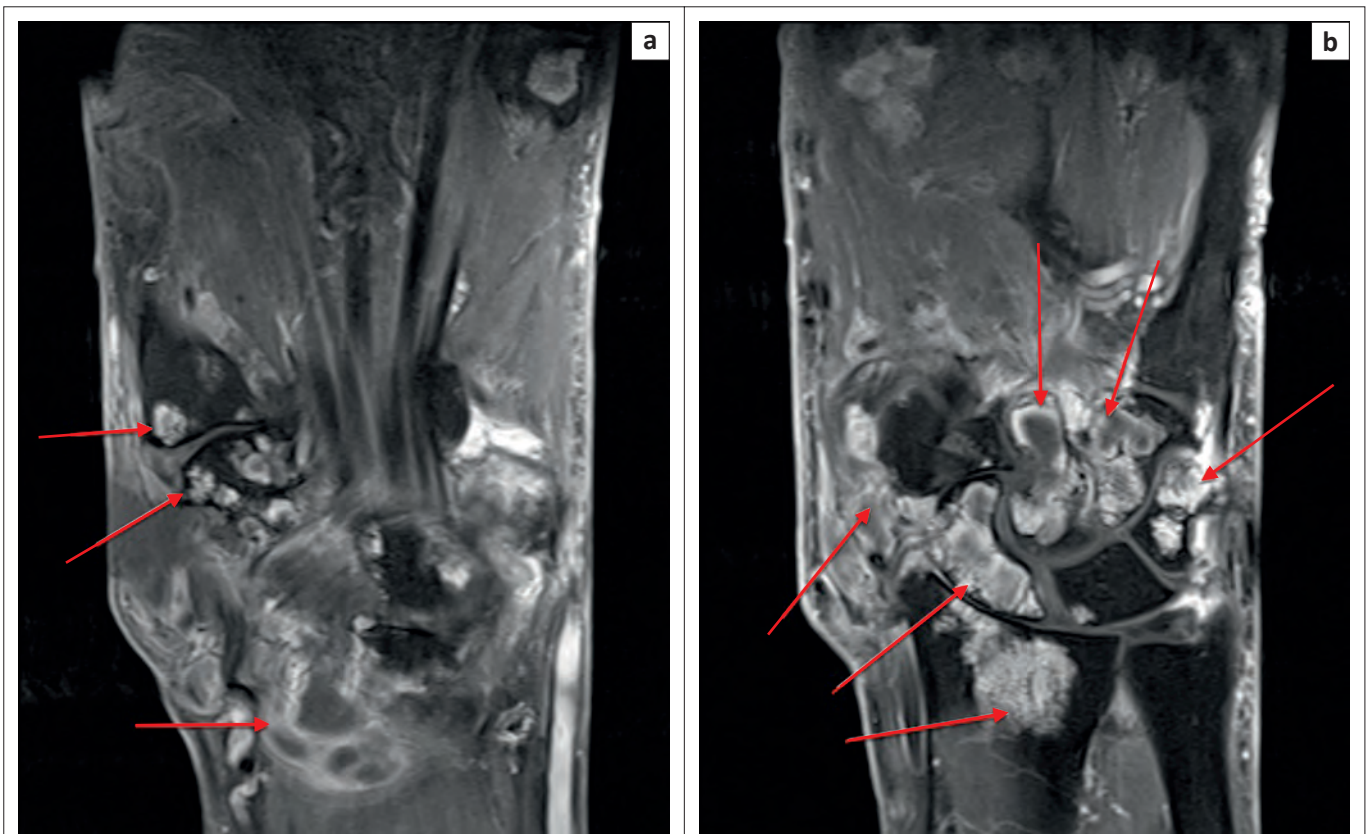


FIGURE 5: Coronal T1-weighted post intravenous gadolinium images (a and b). Intense, predominantly peripheral enhancement of the thenar mass and osseous erosions (red arrows).



FIGURE 6: Coronal T1 FSE-weighted image shows an isointense lobulated mass in the thenar eminence with contiguous spread into the pronator quadratus, the radius, the carpal bones and the bases of the metacarpal bones (white arrows).



FIGURE 7: At surgery, a complex, infiltrating, locally aggressive cystic mass was confirmed.

On ultrasound, the lesion may appear as a complex cystic mass (hypo- or anechoic with increased through-transmission and posterior acoustic enhancement) or may even appear solid. It is hypovascular.¹ Computed tomography (CT) typically shows a lesion denser than water but less dense than muscle.⁸ The lesion on MRI is iso- to hypointense on precontrast T1W, inhomogeneously hyperintense on T2W, and demonstrates mild to moderate diffuse or peripheral contrast enhancement.⁸ Typical MRI findings also include mild perilesional oedema, owing to an incomplete fibrous capsule, and a split-fat sign as a result of adjacent muscle atrophy.⁸

JAM and intramuscular myxoma share the same histological characteristics, but are differentiated by lesion location and the high recurrence rate of JAM. The differential diagnosis of a lesion with imaging findings of a soft-tissue mass that mimics a cyst includes synovial cyst, bursa, ganglion, neurogenic neoplasms, myxoid liposarcomas and myxoid malignant fibrous histiocytoma (MFH).^{5,7}

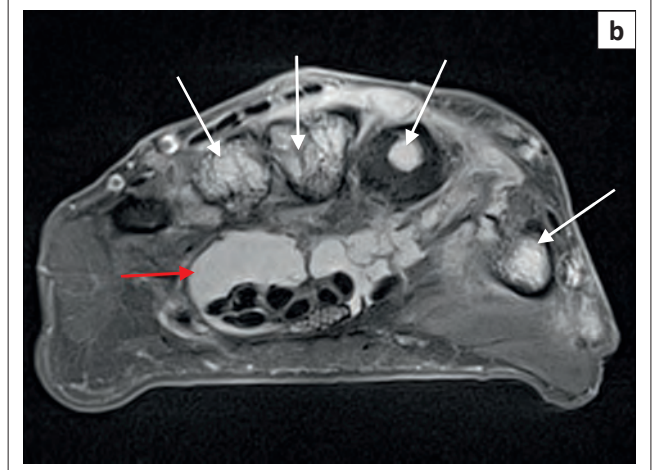
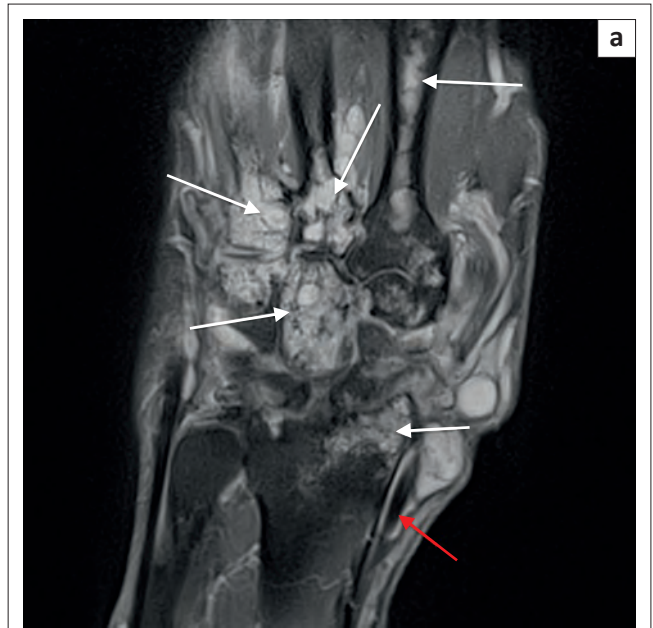


FIGURE 8: Coronal (a) and axial (b) PD fat-saturated images with marked flexor and extensor compartment tenosynovitis (red arrows) and large hyperintense erosions in the radius, carpal and metacarpal bones (white arrows).

Conclusion

JAM is a benign mesenchymal soft-tissue tumour that is a variant of myxoma, associated with large joints and joint degeneration. The present case demonstrates a histologically proven case of JAM in an unusual location, with an unusually aggressive growth pattern and prominent local tissue infiltration.

Acknowledgements

Competing interests

The author declares that they he has no financial or personal relationships that he might have inappropriately influenced him in writing this article.

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