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Ossifying fibromyxoid tumor (OFMT) – A rare cause of a painful thumb

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ABSTRACT

INTRODUCTION: Ossifying fibromyxoid tumor (OFMT) is an uncommon soft tissue and bone neoplasm with just over 100 cases being reported in the literature. They present as small, slow-growing asymptomatic subcutaneous nodules in the soft tissues of the trunk and extremities.
PRESENTATION OF CASE: We present a case of a 25-year-old, right hand dominant gentleman who complained of a seven-year history of pain and discomfort in the dorsal aspect of the right thumb. He was unable to move his interphalangeal joint (IPJ) but had good function otherwise. Examination revealed a localized tender swelling over the dorsal aspect of the IPJ. The thumb was fixed in extension. X-ray revealed marked abnormal soft tissue swelling around the interphalangeal joint, cystic abnormalities and new bone formation. Biopsy showed fibrous tissue containing nodules of tumor with cells in a myxoid background, rounded and histiocytoid to elongated and spindle shaped. CD57, type IV collagen, smooth muscle actin were found and CD56 and EMA were focally positive.

These were in keeping with ossifying fibromyxoid tumor with an atypical immunophenotype.
DISCUSSION: The tumor was formally excised with the flexor pollicis longus tendon. A two-month review revealed his pain had settled. As the tumor had an atypical immunophenotype he was referred to the regional sarcoma team.
CONCLUSION: OMFT can present with atypical clinical, radiological and histological features. It is managed in a multidisciplinary setting and often requires lifetime follow up to detect a recurrence given the uncertain nature of these lesions.

1. Introduction

Ossifying fibromyxoid tumors (OFMT) are rare tumors of unknown origin. They often present with non-specific pain in the extremities. The rarity of the lesion and its atypical features may result in delayed diagnosis and challenges in patient management. We describe a case of atypical OFMT with unique histological and radiological findings. We have also discussed management strategies of the various types of OMFT on the basis of it being a subset of upper limb sarcomas.

2. Case report

A 25-year old Nigerian man experienced seven years of gradually increasing pain and discomfort in his dominant right thumb, post injury in a martial arts competition. He was treated by an allopathic Doctor in his homeland prior to being seen in the United Kingdom. Clinically he presented with an intermittent ache along the dorsal aspect of his thumb, which woke him at night. He was unable to move his interphalangeal joint (IPJ) but had good function. Examination revealed a localized tender swelling beneath a scar over the dorsal aspect of the IPJ. The thumb was fixed in extension.

X-ray revealed marked abnormal soft tissue swelling around the interphalangeal joint, cystic abnormalities and new bone formation adjacent to the head of the proximal phalanx of his right thumb (Fig. 1). Biopsy macroscopically revealed a mass of scar/fibrotic tissue surrounding the IPJ. Microscopy revealed fibrous tissue containing nodules of tumor tissue divided by fibrous septa of varying thickness. The tumor consisted of a variably dense population of cells in a myxoid background, the shapes of which were rounded and histiocytoid to elongated and spindle shaped. CD57, type IV collagen, smooth muscle actin were found and CD56 and EMA were focally positive (Fig. 2). These features were in keeping with ossifying fibromyxoid tumor with an atypical immunophenotype. Two months following his biopsy he reported pain on the volar aspect of his thumb. MRI scan revealed a tumor with myxoid content within the soft tissues of the volar aspect of the proximal phalanx of the thumb extending across the interphalangeal joint. The tumor, which presented macroscopically as dense white fibrous tissue,

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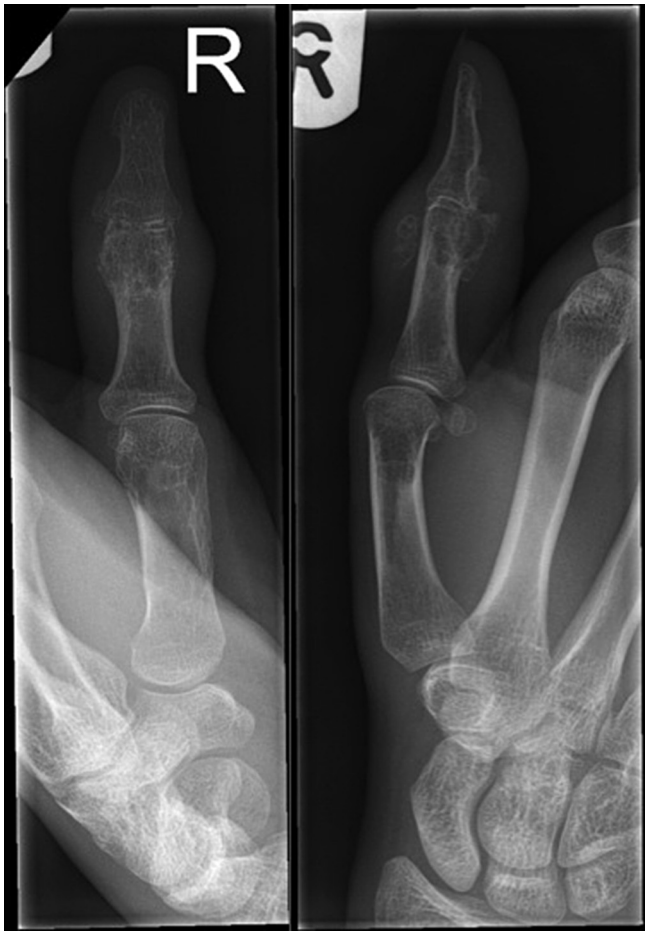

Fig. 1. X-rays (AP and lateral) demonstrating abnormal soft tissue swelling around the interphalangeal joint, cystic abnormalities and new bone formation adjacent to the head of the proximal phalanx.

was formally excised with the flexor pollicis longus tendon as it was seen to be invading the head of the proximal phalanx (Fig. 3). Further histological analysis revealed ovoid or short spindle cells in nests and variable myxoid change. Other immunostains found included MUC4, calponin, h-caldesmon, CD10, D2-40. Subsequent histology reported the tumor to be completely excised.

Two months post-operatively his pain had settled apart from one small area dorsally. As the tumor had an atypical phenotype he was referred to the regional sarcoma team. A multidisciplinary team consisting of plastic surgeons, radiologists, oncologists and the patient assessed the various options of monitoring with yearly MRI scans, radiotherapy or amputation with toe to thumb transfer. The patient opted for monitoring and has shown no signs of recurrence in the past year.

3. Discussion

Ossifying fibromyxoid tumor (OFMT) is an uncommon soft tissue and bone neoplasm with just over 100 cases being reported in the literature. First described by Enzinger et al. in 1989,[1] they most commonly affect middle-aged male adults with a mean age 50 years. Clinically they present as small, slow- growing asymptomatic subcutaneous nodules in the soft tissues of the trunk and extremities.[2] Histological analysis of typical cases reveals lobules of small ovoid to polygonal bland cells arranged in cords and/or nests embedded within a fibromyxoid matrix.[1,3] A band of dense collagen with spicules of metaplastic bone is commonly encountered at the tumor periphery.[4] Most tumors display low cellularity, low nuclear grade, absent necrosis or vascular space invasion and low mitotic rates. As a result they are often classified as benign to at most low grade malignant, however several reports have documented rare atypical and malignant variants of OFMT, with metastasis.[5]

Historically there has been an ongoing debate to define the characteristics of malignancy in OMFT in order to plan follow up and subsequent treatment for these patients. In one of the largest series to date, Folpe et al. classified OFMTs into benign, atypical and malignant. Aggressive OFMT was found to be associated with high nuclear grade, cellularity and mitotic activity. These lesions are labeled as malignant OMFT as there is significant potential for metastasis.[6] Atypical OMFT on the other hand do not fit the appearance of typical OMFT but not meeting the criteria for malignant OMFT. The overall proposed recurrence and metastatic rates of atypical OMFT are 13% and 6% respectively hence the typical and atypical forms of this tumor are considered as lesions of intermediate malignancy.[1,7,8] In our case there was minimal nuclear atypia and no mitotic or apoptotic cells which excluded a diagnosis of malignant OMFT.

A subsequent case study by Graham et al. attempted to further elucidate the natural history of OFMT and reassess the validity of the Folpe and Weiss classification. They examined 46 cases and concluded that the classification system adopted by Folpe et al. was accurate in elucidating malignant OFMT.[3]

The exact origins of these tumors are unclear and debatable. In its first description by Enzinger a neural or cartilaginous origin was proposed as two thirds of the cases contained the immunoreactive s-100 protein and an incomplete shell of mature bone in the capsular region of the tumor. Hence the non- committal

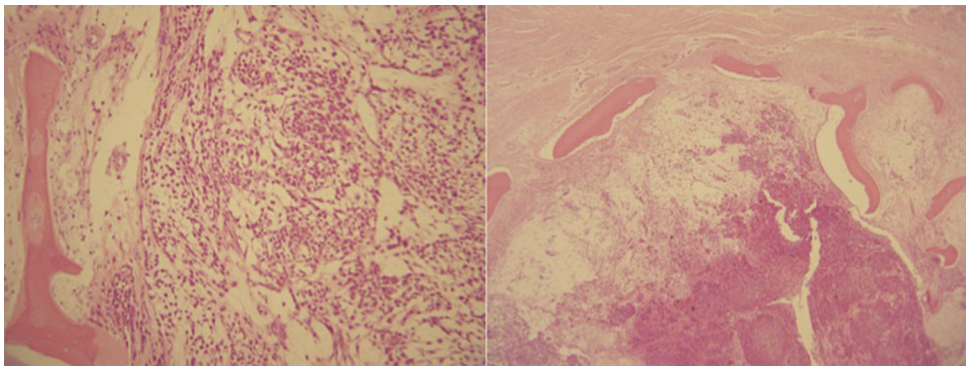

Fig. 2. H&E staining showing nodules of tumor divided by fibrous septa. The tumor consists of cells in a myxoid background. The pale pink area reflects bone formation. The figure to the right has a magnification of ×40 and left ×100.

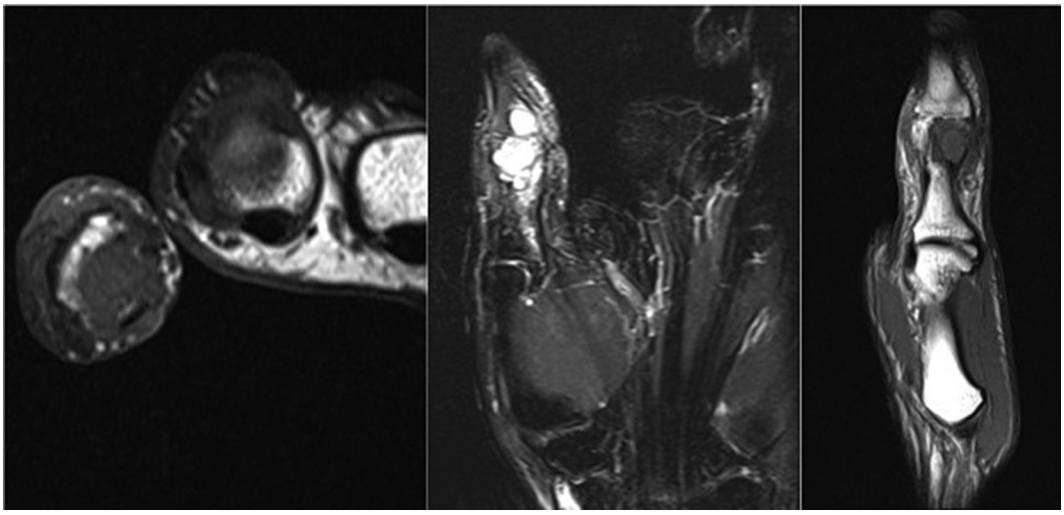

Fig. 3. MRI illustrating the tumor with myxoid content within the soft tissues and bone surrounding the IPJ of the thumb.

label of ossifying fibromyxoid tumor of soft parts.[1] In addition to histology, immunohistochemistry is an integral part of the diagnostic pathway in OMFTs. In the four largest case series s100, desmin and smooth muscle actin were found to be most commonly associated with OMFT. In our case immunohistochemistry revealed cd57, type IV collage, smooth muscle actin, CD56 and EMA. This caused a diagnostic quandary and a differential diagnosis of myxoid perineuroma, nerve sheath myxoma and OMFT was given. However taking into consideration the patient's clinical picture, radiological and histological findings, a diagnosis of atypical OMFT was favored, as it is known to display variable immunophenotypes.

The exact management once the lesion has been excised depends on its classification. Folpe et al. reported a clinical follow up of 27 cases (mixture of typical and atypical OFMT) of about 55 months duration and found all patients to be alive without disease.[6] Graham et al. reported no cases of metastasis in patients with atypical or typical OMFT, Miettinen et al. reported no cases of metastases in typical OMFT although local recurrence was present in 22% of patients at follow up. Given the uncertain history of this rare lesion we recommend a multidisciplinary approach to the management of these patients. Operative intervention is often required for adequate clearance of the tumor and reconstruction depending on functional needs. As this would fall within the realm of a sarcoma follow up would be life long consisting of clinical and radiological assessment (MRI) for recurrence due to the uncertain malignant potential of this rare lesion.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest

No conflicts of interest.

Funding

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Ethical approval

Consent obtained and granted.

Authors contribution

Miss Sharma wrote up the case report and initial draft of the literature review, organised the manuscript and collected images from the radiology department. Dr Hughes, a consultant pathologist provided slides and information on OMFT. Miss Harper is the consultant hand surgeon, primarily responsible for the care of the patient. She is the senior author who revised the report and analysis.

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