# A HUGE GIANT CELL TUMOUR OF THE EXTENSOR TENDON SHEATH OF THE FOOT: A CASE REPORT AND LITERATURE REVIEW

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## ABSTRACT

Giant cell tumours have a predilection for the hand, where after ganglion cysts, they are the most frequent tumour type. Only 3-5% of giant cell tumours occur in the foot, and even here they tend to occur in the forefoot, with hindfoot giant cell tumours being a rarity. While relatively common overall, their exact nature, as to whether they are truly neoplastic or simply inflammatory, is a subject of significant controversy. They are benign slow growing lesions, best treated with gross total excision under magnification. Despite their subcutaneous nature, they can become markedly infiltrative, and unless completely excised exhibit recurrence rates of between 14% and 44%. Factors predictive of recurrence include pressure erosion on X-ray imaging, interphalangeal joint location, concomitant degenerative joint disease, and incomplete excision. We present an adult female patient who presented to our unit with a giant cell tumour extending over the dorsum and medial side of her foot of an alarming size. While the lesion was largely asymptomatic, she was offered surgery for functional reasons. Through the creation of a local flap over the tumour, and using magnification, we were able to achieve a gross total excision. According to our review of the PubMed literature, this is the largest giant cell tumour described in this location, and as such our case report adds value to the world-body of orthopaedic knowledge on the subject.

**Key words:** Giant cell tumour, Extensor tendons foot

## INTRODUCTION

While giant cell tumours are the second most tumour in the hand after ganglion cysts, their occurrence in the foot is relatively rare. The relative distribution between these two sites is in fact so marked, that while several studies report that 85% of cases occur in the fingers, only 3-5% of giant cell tumours occur in the foot and ankle (1-3). Another study supports the rarity of giant cell tumours in the foot, and notes that while 85% of these tumours occur in the hand, outside of the hand the knee is the second most common site accounting for 12% of cases, with their occurrence in the foot being a relative rarity (4). Another epidemiological clustering occurs in individuals in their 3rd -5th decade, and regarding gender distribution displays a female predominance (5,6). Giant cell tumours were first described by Jaffe in the early 1940's, and in this early study they were proposed to be a variant of pigmented villonodular synovitis, and thus classified as non-neoplastic (7). Ongoing controversy in more recent studies persists, with some proposing them to be neoplastic, while others remain steadfast and regard them as non-neoplastic, polyclonal, and inflammatory (8,9). Evidence for this controversy comes from karyotype analysis which has identified aneuploidy DNA, abnormal mitosis, and genetic aberrations in giant cell tumours indicative of neoplasia, while polymerase chain reaction assays of the X-linked human androgen receptor gene note polyclonal proliferation indicating only a reactive neoplastic hyperplastic process (10). Further evidence for the latter is proposed in another study which notes a history of antecedent trauma, and a subsequently likely inflammatory origin, to be present in approximately 50% of cases (11). Whether neoplastic or not, giant cell tumours are now agreed to be a distinct pathology from pigmented villonodular synovitis. The most important distinguishing fact is that while giant cell tumours may rarely secondarily extend into the intra-articular space, it is largely accepted that they originate from the extra-articular tendon sheath and synovium. Pigmented villonodular synovitis on the other hand arises exclusively from the intra-articular synovium, and hence primarily

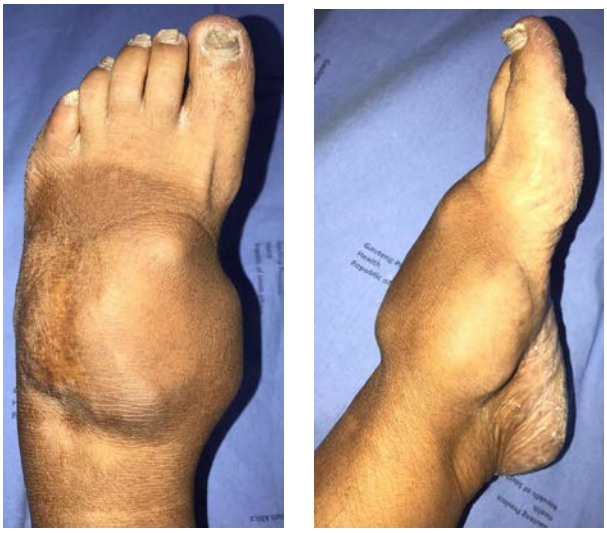
involves the intra-articular chondral and osseous structures, almost invariable destroying the involved joint (1,12). Giant cell tumours are benign, slow-growing, soft tissue tumours of uncertain origin comprising 1.6% of all soft tissue tumours (13). They are largely non-infiltrative, and any bone erosion seen is most commonly only from a local Caspressure effect. Despite these features being present in the majority of cases, bone invasion has been reported to occur in approximately 5% of cases (14), as has malignant transformation (15). Important differential diagnoses for soft tissue tumours of the foot include lipoma, synovial cyst, pigmented villonodular synovitis, fibroma of tendon sheath, synovial sarcoma, undifferentiated pleomophic sarcoma, leiomyosarcoma, and rheumatoid nodule (13,16,17). We present a middle-aged female patient who presented to our unit with an 8-year history of a progressive, painless, soft tissue mass on the dorsum of the left foot, of alarming size, that on histopathological analysis proved to be a diffuse giant cell tumour. According to our review of the Pub Med literature this is the largest giant cell tumour of the foot ever reported, and as such adds value to the world-body of knowledge on the subject.

## CASE REPORT

A 54-year old female patient presented to our unit with progressively increasing in size, painless, mass on the dorsum of the left foot of 8-years duration. What had brought her to the hospital now was purely the fact that she was having difficulty wearing shoes. She denied having any difficulty or pain ambulating. In terms of her medical history she was known with hypertension that was controlled medically, and she had recently been tested and found to be HIV negative. On examination she was a healthy-looking adult female with no stigmata of immunosuppression. General examination was non-contributory, including no popliteal or inguinal lymphadenopathy. Examination of her left leg revealed a normal range of ankle and toe movement on passive testing, however on active testing she had a decreased range of toe flexion. Examination of the mass revealed a 14cm by 11cm multinodular soft tissue mass over the anteromedial aspect of the left foot. The mass had discreet edges, was fixed to the underlying tissues, of firm consistency, with no overlying skin changes. The mass did not trans-illuminate and was non-pulsatile (Figure 1).

**Figure 1**

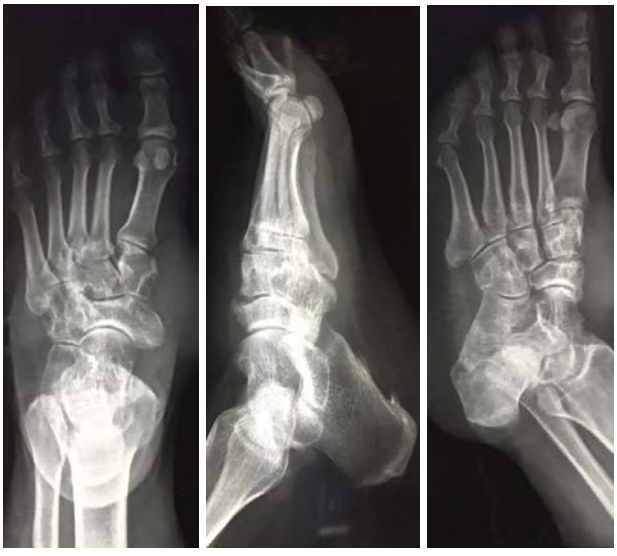
*Pre-operative patient photograph showing the large soft tissue mass on the antero-lateral aspect of the left foot. The alarming size is appreciated. The long history provided alluded to the fact that the mass was in all likelihood benign in anture*



Due to the extensive differential diagnosis of a mass of this size, a fine needle aspiration biopsy was performed which reported the mass to be a diffuse giant cell tumour. We considered this fortunate as malignancy had been excluded, recognized a complete surgical excision to be her best chance of cure, and would primarily consider re-operation in case of recurrence. As part of pre-operative planning X-rays of her left foot were performed which excluded bony involvement by no osteolysis being noted (Figure 2).

**Figure 2**

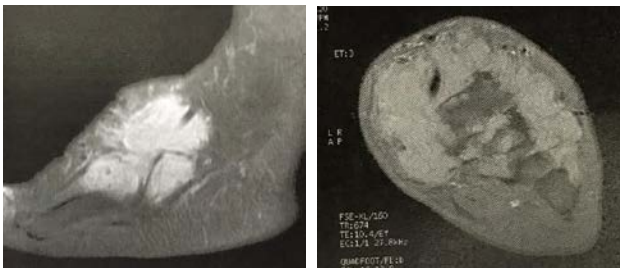
*Pre-operative X-ray of her left foot showed disuse osteopenia however no overt osteolytic focus was noted*



A pre-operative MRI confirmed the multi-lobulated nature of the mass, demonstrated its insinuation between the extensor tendons of the left foot, and demonstrated the circumferential nature of the mass which extended from the dorsum of the left foot over and onto the medial side (Figure 3).

**Figure 3**

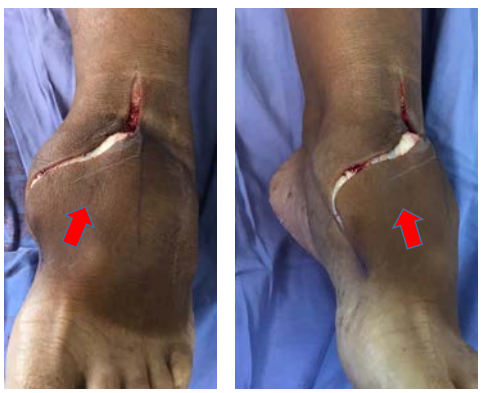
*Pre-operative MRI T2W sagittal and axial views showing the hyperintense multi-lobulated nature of the mass extending over the dorsum and medial aspect of the left foot. The largely subcutaneous nature of the lesion was regarded as favorable and we planned for a gross total resection*



She was taken to the operating room and the tourniquet was insufflated without prior exsanguination to allow for visualization of the vasculature. An 8cm longitudinal skin incision was made over the dorsum of the left foot, and a second 10cm lazy-S skin incision, stemming from the first, was made directed inferior medially over the tumour mass, towards the medial aspect of the foot (Figure 4). The great saphenous vein was identified, ligated, transected, and its ends cauterized.

**Figure 4**

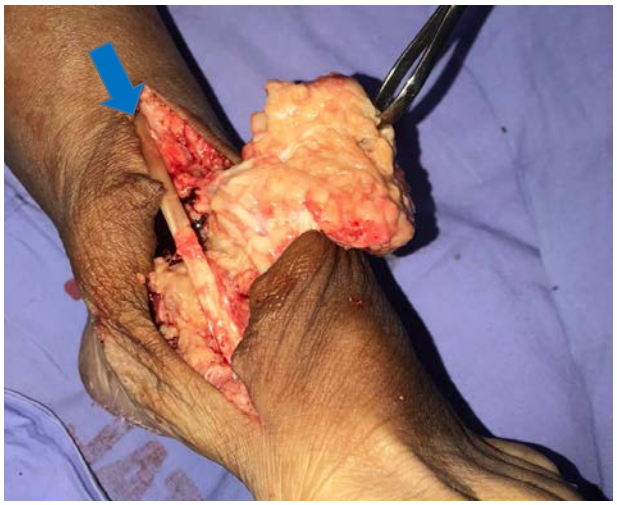
*Intra-operative patient photograph of the skin incision performed to maximize access to the tumour mass. The flap created (red arrow), assisted with this purpose, and was reflected inferiorally. The white fleshy nature of the tumour, directly beneath the skin, was immediately visible*



Using magnification, the tumour was dissected free from the extensor tendon mass, with special attention afforded to the tumour edges to ensure a gross total resection had been achieved. The tumour was noted to have most likely arisen from the synovial sheath of extensor hallucis longus tendon, insinuated between the extensor tendons over the dorsum of the foot, and infiltrated beneath the tibialis anterior tendon on the medial aspect of the foot (Figure 5).

**Figure 5**

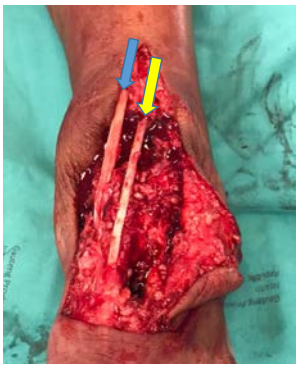
*Intra-operative patient photograph showing the medial aspect of the tumour where it had infiltrated beneath the tibialis anterior tendon (blue arrow)*



A meticulous dissection was performed until a gross total resection, confirmed by magnification, had been achieved. A necessary part of the operation was to resect part of the extensor tendon sheath of the extensor hallucis longus muscle (Figure 6).

**Figure 6**

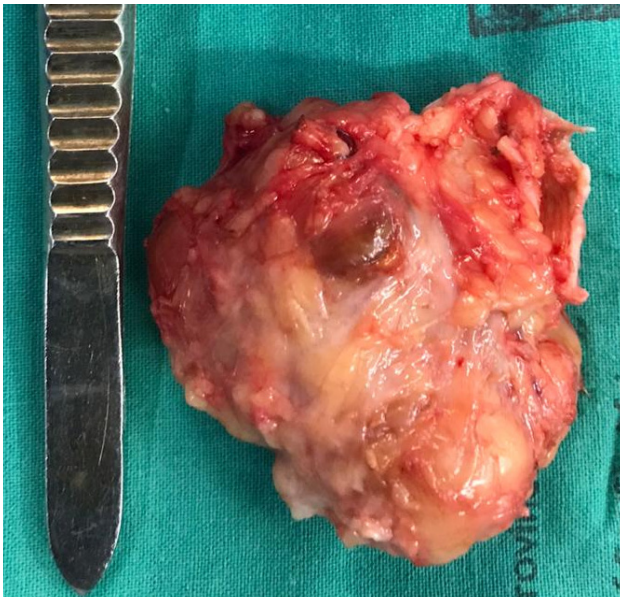
*Intra-operative patient photograph showing the tumour bed after gross total resection of the tumour. The exposed tendons of the extensor hallucis longus muscle (yellow arrow), and the tibialis anterior muscle (blue arrow), can be seen*



The fleshy nature of the main tumour mass was sent for histopathological analysis (Figure 7), the wound irrigated with hydrogen peroxide, washed with saline, and closed in layers. The skin was closed with interrupted nylon mattress sutures.

**Figure 7**

*Intra-operative photograph of the main tumour mass on a sterile drape. The typical solid, fleshy nature, of a giant cell tumour can be appreciated*



The patient was discharged on the 3rd post-operative day, fully ambulant, for out-patient follow-up. At her 2-week follow-up appointment the histopathological analysis became available. According to this report the specimen was mostly comprised of nests of large mononuclear polygonal cells with an eosinophilic cytoplasm, contained within a collagenous hyalinized stroma. Osteoclast-like multinucleated giant cells were randomly distributed within the tumour matrix, as were scattered foam cells. A lymphocytic infiltrate was further noted. Hemosiderin deposits were also seen. Vascular invasion, necrosis, and mitotic figures were not seen. Regarding staining both the mononuclear and multinucleated cells stained positive with CD-68, however only the multinucleated cells demonstrated membrane positivity for CD-45. The entirety of the tumour was negative for S-100 and desmin. The conclusion of the histopathological report was that the lesion was a giant cell tumour- diffuse type. At her 6-week out-patient appointment the patient was noted to be wearing closed shoes, and on examination her wound had healed well. She was booked for 1-year out-patient review. In the event of recurrence, we planned to consider re-operation as our first line intervention and reserved adjunctive radiotherapy for re-consideration depending on the size of the recurrence and findings of the surgery.

## DISCUSSION

Several classification systems exist by which to classify giant cell tumours. The first is a clinical-radiological classification, proposed by Al Qattan, which divides giant cell tumours into three types. Type 1 is a single tumour that is round and multilobulated, such as that in our case. Type 2 refers to two or more distinct separate tumours. Type 3 refers to recurrent satellite lesions after an incomplete primary excision (17). The histopathological classification, proposed by the World Health Organization (WHO), classifies giant cell tumours as fibriohistiocytic tumours. Here a localized nodular type, characterized by a relatively hypocellular nature with numerous giant cells, is differentiated from a diffuse type characterized by hyper cellularity with numerous polygonal mononuclear cells and less frequent giant cells (18). Giant cell tumours of the extensor tendon mass, of whatever histopathological type, are noted to be found in the subcutaneous tissue plane arising from the tendon sheath, with extensions that infiltrate over and under the surrounding structures. The neurovascular bundle is often found displaced by the tumour mass, and hence the importance of magnification is proposed to make separation possible (19). Gross total excision is recognized by several studies to offer the best chance of cure (19-21). Despite this aim, recurrence rates are reported to be between 14% and 44% (21,22). Factors predictive of local recurrence include pressure erosion on X-ray imaging, interphalangeal joint location, concomitant degenerative joint disease, and incomplete excision (22,23). One prospective study, that gave an adjunctive total dose of 20G of radiotherapy, in 2G divided doses, to 14 patients with incomplete excision, mitotic figures, or bone involvement, noted no recurrences. This study proposes a role for adjunctive radiotherapy in the management of this specific post-operative patient subset (21). Regarding the pre-operative diagnosis by fine needle aspiration, as was performed in our patient, this is supported by one study as an important tool for pre-operative surgical planning to prevent recurrence (24). Another study, albeit only a case report, supports our practice and similarly used hydrogen peroxide lavage of the tumour bed post removal of a giant cell tumour from the ankle of a 58-year old adult male. In this study no recurrence had occurred at 2-year follow-up (25).

## CONCLUSION

Giant cell tumours are relatively rare on the dorsum of the foot compared to their incidence on the hand. When they are present, they most commonly affect the forefoot and their presence over the hindfoot is a rarity. One study that considered 14 giant cell tumours of the foot, noted 11/14 (79%) to involve the forefoot, and only 3/14 (21%) involved the hindfoot (26). While benign in nature, their propensity to recur makes gross total excision under magnification the treatment of choice. Recurrence is similarly best treated with re-operation. Our case report provides an account of a giant cell tumour of the extensor tendon sheath of the foot of alarming size. Our study hopes to assist foot and ankle orthopaedic surgeons who will in all probability need to manage this specific tumour at some point in their careers.

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