Tumours of the foot and ankle

Zeeshan Khan ∗, Shakir Hussain, Simon R. Carter

Bone Tumour and Adult Reconstruction Unit, The Royal Orthopaedic Hospital, Bristol Road South, Birmingham B31 2AP, UK

Abstract

Sarcomas are rare tumours and particularly rarer in the foot and ankle region. The complex anatomy of the foot and ankle makes it unique and hence poses a challenge to the surgeon for limb salvage surgery. Other lesions found in the foot and ankle region are benign bone and soft tissue tumours, metastasis and infection.

The purpose of this article is to discuss the relevance of the complex anatomy of the foot and ankle in relation to tumours, clinical features, their general management principles and further discussion about some of the more common bone and soft tissue lesions. Discussion of every single bone and soft tissue lesion in the foot and ankle region is beyond the scope of this article.

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1. Introduction

It is estimated that sarcomas comprise 0.2% of all tumours and only 2% of all the sarcomas arise in the foot and ankle region [1]. Tumours are reasonably common in the foot and ankle region but based on just clinical examination it is difficult to distinguish benign from malignant lesions. This can lead to the inadvertent “whoops procedure” necessitating further treatment due to lack of surgical clearance, high risk of local recurrence and metastasis due to the rich vascular and lymphatic network in this region [2,10]. Negligence in clinical workup and awareness of some common tumours in this region along with abiding by the general management principles of any lump would reduce error.

2. Anatomy

The foot is a unique anatomic structure with many critical functional units in close proximity. The unique feature of this region is the presence of small bones, small epiphysis, thin cortices, muscles being arranged in various layers in very close proximity to each other and lack of fascial barriers. Because of these features there is early bone destruction which along with the high vascular and lymphatic network makes wide excision very challenging for the surgeon and amputation may be the only safe option.

3. Clinical presentation

The commonest presentations of any tumour in the foot and ankle region are pain and a lump [7]. Certain tumours are common in particular age groups and this helps in narrowing down the diagnosis along with specific location within the bone.

Pain at rest should be considered a worrying feature and sometimes its response to a particular analgesic, non-steroidal anti inflammatory drugs, can point to a specific diagnosis (Osteoid osteoma). Because of the complex and tight anatomical structure of this region, any lump would be expected to declare itself early in the disease process, particularly over the dorsal aspect of the foot due to the relative scarcity of soft tissues in comparison to the plantar side.

Occasionally, a lump may have been present and dormant for many years but it suddenly starts growing or become symptomatic, in which case the physician should be alerted to the possibility of malignancy. Any change in a pigmented lesion should also be investigated properly as melanomas are not uncommon in this region [11]. Occasionally tumours in this region can present with neurological symptoms and this maybe a clue to its extent and location.

A detailed past medical history is also relevant as metastasis and secondary sarcomas (radiation induced) can also occur in this region. Acrometastasis from primary lung cancer are reported to be common but a study published from the senior authors institution suggests infra diaphragmatic cancers to be a more common cause [7].

4. Physical examination

A systemic approach to the examination of the foot and ankle region should be adopted. Comparison should be made with the contralateral side where possible. The commonest findings would be a lump, tenderness, pain on movement of the involved joints, sensory or motor neurological deficit and occasionally a deformity. The site, size, depth, consistency, tenderness, trans illumination and pulsatility of soft tissue lumps, adherence to joints and tendons should be evaluated. Although rare in sarcomas, but loco regional lymph nodes should be palpated. In case of any neurological symptoms, a detailed neurological examination should be conducted along with examination of the spine.

5. Management

The standard work up for any foot and ankle disorder will include an anteroposterior, lateral and oblique weight bearing radiograph. This is relatively cheaper, easily available and yields a lot of information. Bony and joint destruction in case of aggressive tumours is easily visible along with an outline of soft tissue lesions. Calcification within a soft tissue lump can point to a synovial sarcoma [12].

A well demarcated lesion in the bone with no significant cortical reaction or destruction will generally point towards a more benign lesion along with a narrow zone of transition and homogeneity. To gain further 3 dimensional detail about the bony architecture, CT scan is the investigation of choice particularly in cases where a surgeon would consider limb salvage surgery.

In order to assess the soft tissues, Ultrasonography and MRI scans are the investigations of choice. The former investigation is cheap, quick and easily available but its major drawback is operator dependence. Furthermore, it can also demonstrate the vascularity of the lesion.

Magnetic resonance scan provides extensive details about both soft tissue and osseous lesions including extent of tumour, constituents, anatomical relationship with other important structures and bony oedema. Infection and stress fractures can also be ruled out with this modality if there is a suspicion based on history and other investigations. A detailed history provided to the radiologist prior to the scan can aid them in obtaining the best sequences to obtain the diagnosis.

Nuclear medicine bone scans are performed in cases where metastasis are considered and also in the case of a known primary with no known previous metastasis. As a standard work up for any sarcoma, a chest X-ray or CT scan of the chest is also
routinely obtained for systemic staging. Blood tests including a full blood count, inflammatory markers, various tumour markers, bone profile and calcium levels are also part of the standard work up.

6. Biopsy

Biopsy is the gold standard for diagnosis and the best type is the one which yields the appropriate type and amount of tissue for the histopathologist to make a certain diagnosis. Hence, discussion with the histopathologist about the type of biopsy prior to the procedure would reduce the risk of repeat procedures. A biopsy can be fine needle aspiration, open trucut or excisional and can be performed either by the orthopaedic surgeon or a radiologist under image guidance depending on the location of the lesion [13].

Regardless of the type of biopsy, the principles of biopsy should be strongly adhered to.

Some of the common principles of biopsy are as follows:

1. It should ideally be performed by the surgeon who will undertake definite management or after consultation with them and should always be longitudinal.
2. It should be minimally invasive.

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**Fig. 2.** Giant cell tumour of tendon sheath.

**Fig. 3.** Sagittal view (MRI) showing extensive involvement of the ankle, subtalar and midfoot joints by pigmented villonodular synovitis (PVNS).

**Fig. 4.** Plain radiographs of the patient in Fig. 2, demonstrating extensive bony and joint destruction with PVNS.
Fig. 5. Subcutaneous lipoma over the dorso lateral aspect of right foot causing problems with foot wear.

Fig. 6. Peripheral nerve sheath tumour (Schwannoma) of the right ankle demonstrated on axial view of MRI scan.

(3) It should not contaminate intercompartmental normal tissue.
(4) The incision for biopsy should be in line with the definite incision employed for further surgery.
(5) It should be a representative sample of the lesion and necrotic areas should be avoided.
(6) Meticulous haemostasis should be achieved to prevent local spread of tumour.
(7) If a drain is used, it should be in line with the main incision.
(8) The sample should be assessed by an appropriate histopathologist.

7. Soft tissue lesions

Soft tissue tumours may present as a lump, which may or may not be painful, and or a deformity. Because of the complex anatomy of this region it is of utmost importance to establish a definitive diagnosis before embarking on any surgical treatment.

Fig. 7. Clinical photograph of a lump around the left ankle, dorsal aspect, which on further investigations turned out to be a synovial sarcoma.

Table 1

<table>
<thead>
<tr>
<th>Benign lesions</th>
<th>Malignant lesions</th>
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<tbody>
<tr>
<td>(1) Ganglion</td>
<td>(1) Synovial sarcoma</td>
</tr>
<tr>
<td>(2) Plantar fibromatosis</td>
<td>(2) Malignant melanoma</td>
</tr>
<tr>
<td>(3) Fibroma</td>
<td>(3) Clear cell sarcoma</td>
</tr>
<tr>
<td>(4) Giant cell tumour of tendon sheath</td>
<td>(4) Epithelioid sarcoma</td>
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<tr>
<td>(5) Pigmented villonodular synovitis</td>
<td>(5) Haemangioepithelioma</td>
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<td>(6) Lipoma</td>
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<td>(7) Schwannoma</td>
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<td>(8) Haemangioma</td>
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<tr>
<td>(9) Glomus tumour</td>
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<tr>
<td>(10) Nodular fascitis</td>
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Benign tumours form the major bulk of soft tissue tumours in this region [3] (Table 1).

Sarcomas are more common in the younger (10–40 years old) and older (above 60 years) age groups. In the UK, any patient presenting to their primary care physician with a lump with any of these following features is referred to the local sarcoma unit:

(1) Lump size more than 5 cm.
(2) Increase in size.
(3) Deep to the deep fascia.
(4) Painful.
(5) Recurrent.
If any of these features are present, malignancy should be excluded. A lump with all of these features present has an 86% positive predictive value for malignant potential [13].

Presence or absence of pain, size of the lesion and duration of symptoms in the foot and ankle region cannot confidently exclude malignancy [3]. The mean size of a sarcoma at presentation in a case series published from the senior author’s institution was 10 cm [14].

8. General management principles

The aim of any surgical intervention in sarcoma surgery is to achieve wide margins. There is no universal consensus on what constitutes wide margins but generally 1 cm of normal tissue or intact fascial layer is considered adequate.

Because of the complex anatomy of the foot and ankle region, one may not be able to achieve these safe margins. In order to achieve this and to reduce the risk of local recurrence, ablative surgery maybe the only feasible option.

Occasionally a surgeon may compromise on wide margins in order to perform limb salvage surgery (spare major neurovascular structures) and leave a planned positive margin after discussing this with the patient and its implications. Along with the wide margins, the risk of local recurrence is also dependent upon the grade of the tumour [18].

Adjuvant radiotherapy is the standard treatment for all intermediate and high grade sarcomas and has similar results in comparison to radical resection with the added advantage of preservation of function [19].

Low grade tumours generally do not necessitate radiotherapy but should be considered in complex anatomical sites such as the foot and ankle where achieving wide surgical margins may not be possible.

The role of chemotherapy in soft tissue sarcomas is yet to be established and is not considered a standard treatment in the UK.

It is predominantly reserved for trials, metastatic disease, patients older than 65 years and high grade tumours. The overall prognosis depends on the grade, size and depth of the tumour and age of the patient [13,14].

8.1. Benign tumours

A ganglion is the commonest benign lesion in this region after hands [15]. It is commonly found in areas of frequent physical stress i.e. Joint capsule and tendon sheath. They present as a firm subcutaneous lump which may spontaneously disappear or rupture. They may cause symptoms in relation to its location and foot wear (Fig. 1). They trans illuminate on the torch test which along with an ultra sound scan and aspiration help yield the diagnosis. A thick gel like fluid is aspirated which occasionally is difficult to perform depending on the consistency of the fluid. If they are symptomatic, we recommend surgical excision with dissection up to the stalk of the ganglion to reduce the risk of recurrence which is a common complication.

Plantar fibromatosis, desmoid or Ledderhose disease is a locally aggressive, benign, hyperproliferative disorder of plantar aponeurosis. It is more common in men and its greatest prevalence is in middle age and beyond. It can be associated with other fibromatosis such as Duputyrens and Peyronies disease. It is characterised by fibrous proliferation of abnormal fibrous tissue in the plantar fascia in the form of nodules and cords which can be extremely painful on weight bearing [16]. Risk of recurrence is high with marginal excision and hence resection of the nodule along with the overlying skin is advisable, in which case skin graft may be required.

Fibromas are benign fibroblastic tumours interspersed with thick collagen fibres. They present as firm, subcutaneous, well localised nodules and are usually non tender. Marginal excision of symptomatic lumps is the recommended treatment with a low risk of recurrence locally.
Giant cell tumours of the tendon sheath are mostly asymptomatic and patients may only seek attention after pressure symptoms from footwear (Fig. 2). These are benign lesions formed predominantly of giant cells, foam cells and reticulum cells. They present as well defined firm nodules and may disappear spontaneously but in any case recurrence after marginal excision is very low.

Pigmented villonodular synovitis (PVNS) is a benign locally aggressive tumour which can cause bony erosions and affects the knee and ankle most commonly (Figs. 3 and 4). The midfoot is the commonest involved region in the foot and this can involve multiple bones. Clinically they will present as swelling and effusion along with discomfort over the involved joints. The synovium is usually brownish and on X-rays there are bony erosions near the joints. Isolated nodules can be resected arthroscopically if possible, but in advanced cases, total or near total synovectomy is advisable to reduce the risk of recurrence [17]. In advanced cases reconstructive surgery of the involved joints may be needed.

Lipomas are benign encapsulated lesions of adipose tissue which are subcutaneous and doughy in consistency. They are usually found on the dorsum of the foot and are asymptomatic unless they cause pressure symptoms (Fig. 5). Marginal excision is advisable for the symptomatic ones with a low risk of recurrence.

Schwannomas are well encapsulated benign solitary tumours of the nerve sheath. They have a positive percussion test and if symptomatic, careful enucleation of the lesion from the nerve sheath is advisable to reduce the damage to the nerve (Fig. 6).

A Glomus tumour usually presents a dark discolouration under the toe nail, which is sensitive to cold and pressure. X-rays of the foot usually show characteristic scalloping of the terminal phalynx. Marginal excision of the symptomatic lesions is advisable.

8.2. Malignant tumours

Soft tissue sarcomas are common in the middle and old aged patients but also account for 7–10% of malignancies in the paediatric population [13].

Synovial sarcoma is the commonest malignant soft tissue tumour in this region. It has a variable presentation and may present as pain or a lump which has been latent for a long time and then suddenly starts growing and become symptomatic. It commonly presents in the young or middle aged adults with a firm and fixed mass [4]. Involvement of loco regional lymph nodes is more common with synovial sarcoma and they must be examined. Intralesional calcification on radiographs is a characteristic finding in these tumours. Although surgery is the mainstay of treatment, adjuvant radiotherapy has a role in its management. Wide margins are aimed for during surgery and this should not be compromised for limb salvage surgery and on occasions amputation maybe the only safe treatment due to the complex anatomy of this region (Figs. 7 and 8).

Malignant melanoma is another common soft tissue sarcoma in this region. Any change in the character, shape and size of a pigmented lesion should raise the suspicion of malignancy. Wide resection is the treatment of choice which may or may not require soft tissue reconstruction.

Fig. 10. Enchondroma of 5th metatarsal head.
Table 2
Common bone tumours.

<table>
<thead>
<tr>
<th>Benign bone tumours</th>
<th>Malignant bone tumours</th>
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<tbody>
<tr>
<td>Osteochondroma</td>
<td>Osteosarcoma</td>
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<tr>
<td>Enchondroma</td>
<td>Chondrosarcoma</td>
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<tr>
<td>Osteoid osteoma</td>
<td>Ewing's sarcoma</td>
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<td>Osteoblastoma</td>
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<td>Simple bone cyst (SBC)</td>
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<tr>
<td>Aneurysmal bone cyst</td>
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<tr>
<td>Giant cell tumour</td>
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<tr>
<td>Chondroblastoma</td>
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<tr>
<td>Subungual exostosis</td>
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9. Osseous lesions

The commonest bone tumours are benign with some of the commonest lesions of both varieties mentioned in Table 2.

9.1. Benign bone tumours

Osteochondromas are usually solitary, sessile or pedunculated, benign lesions found in the metaphyseal region growing away from the physis (Fig. 9). They maybe multiple and part of a rare autosomal dominant condition called hereditary multiple exostosis (HME). Malignant change can happen in the cartilage cap (1% in solitary lesions and 5–25% in HME) in which case it is usually a grade 1 chondrosarcoma [5]. Simple excision at the base is usually adequate for solitary osteochondromas if they are symptomatic.

Enchondromas are benign, intramedullary, cartilaginous tumours which are often solitary but in cases of multiple enchondromas, it is called Ollier's disease (Figs. 10 and 11). They are most commonly found in the tubular bones of hands and feet. Isolated lesions are usually asymptomatic and are incidental findings but can occasionally present with pathological fractures. Painful or at risk lesions can be treated with curettage after confirmation of diagnosis which may or may not also need bone grafting. Ollier’s disease in association with soft tissue haemangiomas is called Maffucci’s syndrome and in both cases the risk of malignant transformation of these enchondromas is approximately 30%.

Osteoid osteoma is a benign self limiting neoplasm which presents mainly with non-activity related pain in younger patients (5–30 years old) and can be nocturnal. This pain classically responds well to non steroidal anti inflammatory drugs (NSAIDS) which is a diagnostic clincher. They are more common in long bones but can present in the foot and ankle region [9]. Plain radiographs may not be able to show the small radiolucent nidus (<1.5 cm) with the surrounding rim of sclerosis and CT scan is a better choice in this case. A bone scan will always be hot in this case. If left on their own, they resolve over a few years but if the symptoms are distressing surgical intervention in the form of image guided radiofrequency ablation (RFA) is the preferred treatment [8] (Fig. 12). If RFA is not available, then open surgery in the form thorough curettage of the lesion is recommended which may or may not include bone grafting as well.

If the radiolucent nidus is bigger than 1.5 cm, usually 2 cm or more, then the lesion is called an Osteoblastoma (Fig. 13). It is less common than osteoid osteoma and presents mostly in the 15–30 years age group with pain. They will usually present near the growth plate but most commonly in the dorsal talar neck. They do not respond so well to NSAIDS as their other counterpart and continue to grow and curettage is the treatment of choice.

Simple bone cysts (SBC) are benign fluid filled lesions thought to be formed due to medullary bone malformation. They are not as common in the foot and ankle region and usually found in the os calcis. They are usually asymptomatic and if they cause symptoms then thorough curettage and bone grafting is the treatment of choice (Fig. 14).

Aneurysmal bone cysts are benign lesions common in the <20 years age group with the commonest bone involved in this region being the metatarsals. Plain radiographs will demonstrate an expansile lesion with a thin cortex and multiple fluid levels noted...
Fig. 14. Simple bone cyst in os calcis.

Fig. 15. T1 image, axial view of os calcis demonstrating multiple fluid levels in an aneurysmal bone cyst.

Fig. 16. Giant cell tumour of bone in 1st metatarsal with its classical epiphyseal and peri articural location.

on MRI scans (Fig. 15). Curettage and bone grafting is the treatment of choice with a risk of recurrence.

Giant cell tumours (GCT) are benign bony tumours with a high risk of recurrence, approximately 50%, and a 2% risk of systemic metastasis [20]. They usually present as a painful mass in the epiphyseal region, commonly in 30–40 years age group with the os calcis and talus being most commonly affected (Fig. 16). Thorough curettage with high speed burrs is the main stay of treatment with other adjuvant treatments being proposed with good but variable success rates (bone grating, cementation, cryotherapy and radiotherapy).

9.2. Malignant bone tumours

Malignant osseous lesions in the foot and ankle region are relatively uncommon. The commonest malignant lesions in this region are Ewing’s sarcoma, Chondrosarcoma and Osteosarcoma. Delayed or missed diagnosis is a common presentation in this region which can make limb salvage surgery very demanding and ablative surgery maybe the only option [7].

Ewing’s sarcoma has the worst prognosis of all primary sarcomas and it usually presents as a lytic expansile mass in the metatarsals with onion peel appearance (Fig. 17). Multi agent chemotherapy is used for systemic control and surgery and or radiotherapy is used for local control [6]. Surgery usually involves amputation depending on the bones involved and maybe in the form of ray amputation for toes or metatarsals and below knee amputation in case of midfoot and hind foot involvement.

In contrast, chondrosarcomas are slow growing tumours usually presenting in middle aged to older patients (Fig. 18). Surgery in the form of wide excision is the recommended treatment with very poor response to chemo or radiotherapy.
Less than 1% osteosarcomas occur in this region and it has a bimodal age distribution. It usually presents as a painful, enlarging hard mass. Multi agent chemotherapy and wide surgical resection is the mainstay of treatment.

For any malignant lesion, the clinical and radiological follow up in the senior authors institution is at 3 monthly intervals for the first 2 years, then 6 monthly up till year 5 and then on annual basis up till year 10.

10. Metastatic disease

Metastasis in foot and ankle are rare (less than 1%) and are associated with poor prognosis. These patients present with pain and swelling with lung, colon and genitourinary tumours being the commonest primary tumours. Palliative radiotherapy and surgery for local control is the main stay of treatment [7].

Conflicts of interest

None declared.

References
