

Simple presentations of sinister pathology to an upper limb orthopaedic clinic

Abstract

Background: Bone and soft tissue sarcomas present to orthopaedic and rheumatology clinics in district general hospitals with a diverse array of symptoms and signs. Delays in diagnosis adversely effect prognosis and referral pathways have been set up in order to centralise care of these complex and rare tumours to improve outcomes.

Methods: We present a series of 7 interesting cases that presented with bone and soft tissue tumours to an elective upper limb clinic over a period of 3 years and managed using the appropriate referral pathways.

Results: In this series 1 patient presented with a metastatic bone deposit from a renal primary and had treatment locally after investigations suggested it was metastatic. Six patients were seen with soft tissue swellings, had magnetic resonance imaging and were subsequently referred to the sarcoma unit for further assessment. Two patients underwent biopsy and were found to have uncommon benign pathology – nodular fasciitis and tumoural calcinosis. The remaining 4 patients were found to have sarcomas with different histological features – spindle cell, epithelioid, alveolar soft part and follicular dendritic (recurrent). Each of these patients was referred expeditiously from the orthopaedic clinic but despite this later developed metastasis from their primary sarcoma.

Discussion: Recognition of bone and soft tissue tumours is not always straightforward and it is not unusual to see delays in diagnosis and consequent referral. Late referral to specialist orthopaedic services has a significant impact on prognosis and guidelines have been developed to facilitate this. Awareness of the red flag symptoms and signs to arrange prompt referral leads to better outcomes.

Keywords: Soft tissue sarcoma, Tumour, Nodular fasciitis, Tumoural calcinosis

Volume 2 Issue 2 - 2015

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Received: December 16, 2014 | **Published:** March 4, 2015

Abbreviations: CT, Computed Tomography; GP, General Practitioner; MRI, Magnetic Resonance Imaging; NICE, National Institute for Health and Care Excellence; PET, Positive Emission Tomography; UK, United Kingdom

Introduction

Bone and soft tissue tumours are seen infrequently in district general hospitals and diagnosis requires a good history and examination with good recognition of red flag symptoms offered by the patient. Delay in diagnosis of these potentially life changing diagnoses have an effect on treatment options and the potential for curative therapy. In the UK, referral to specialist bone and soft tissue tumour centres allows for rapid diagnosis and multidisciplinary lead treatment. Primary bone tumours are rare with metastatic tumours occurring more frequently with the primary often found in the lung, kidney, thyroid, breast or prostate. Sarcomas account for 0.2% of all new cancer cases in the UK.¹ Sarcomas are often referred from general practice to local orthopaedic clinics despite 2-week wait targets from NICE with guidelines to suggest urgent referral to a soft tissue.² We present a series of 10 cases presenting to an elective upper limb clinic over a 3-year period, between August 2010 and August 2013.

Case Series

Case 1: Renal cell carcinoma

This man in his seventies was referred by the GP with a 3-month gradual shoulder swelling. Examination revealed a 9x8cm firm swelling over the lateral third of the clavicle and an X-ray revealed

a destructive bony lesion. A staging CT scan was arranged and the patient was found to have a probable renal cell carcinoma and was treated with radical nephrectomy and chemotherapy. No specific treatment was given to the clavicular metastasis (Figure 1).

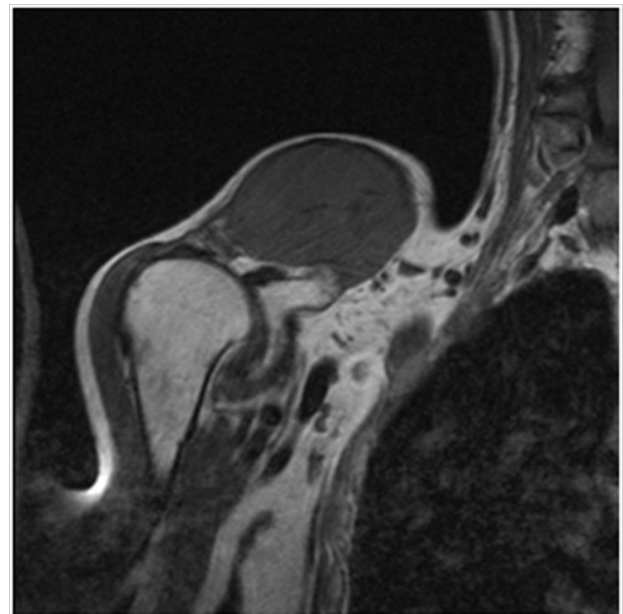


Figure 1 MRI of metastatic Renal Cell Carcinoma of the distal clavicle.

Case 2: Follicular dendritic cell sarcoma

Our second patient in her late fifties was initially seen by the Rheumatologists with shoulder pain following a referral from the GP but was referred to the orthopaedic clinic. A mixed lytic sclerotic lesion of the scapula was found and following referral to the local bone tumour service. She had a history of follicular dendritic cell sarcoma of the neck excised in the late nineties with a breast cancer the following year. Scans suggested it was a solitary metastasis and a decision was made to perform scapulectomy. A few weeks later she was found to have a solitary lymph node in her neck which was also removed and found to suggest metastatic follicular dendritic cell sarcoma. She consequently received radiotherapy but developed further metastasis despite this Figure 2.



Figure 2 X-ray of post-op scapulectomy for metastatic Follicular Dendritic Cell Sarcoma.

Case 3: Epithelioid sarcoma

This gentleman in his forties was referred by the GP with a 3-month history of gradual swelling of the proximal forearm. In clinic it was found to be 5x3cm in size with dysfunction of the wrist flexors. An urgent MRI was arranged for further characterisation and was found to arise from flexor digitorum superficialis with some smaller satellite lesions. Urgent referral to the local tumour service was completed and biopsy revealed an epithelioid sarcoma. PET scan noted axillary lymphadenopathy and following discussion the patient opted for a forequarter amputation and axillary node clearance with curative intent. Histology revealed clear resection margins and he was not found to have local recurrence at follow up. CT scan 9 months postoperatively unfortunately revealed multiple lung nodules and was further treated at another hospital (Figure 3).

Case 4: Alveolar soft part sarcoma

This gentleman was seen in clinic following a delayed presentation to the GP reporting variable pain and swelling of the proximal forearm for approximately 2 years. He thought he had pulled a muscle in his arm and felt the swelling was coming and going and so did not seek medical attention. He started getting paresthesia and aching which

prompted him to attend his GP. In clinic he was found to have a soft tethered swelling and an MRI demonstrated an irregular 4x7cm mass in the flexor compartment of the forearm. A CT scan revealed multiple pulmonary nodules with one larger mass measuring 6cm in the right lower lobe. An opinion from the sarcoma unit was sought and subsequent biopsy confirmed an alveolar soft part sarcoma with lung metastasis and primary palliative treatment was suggested Figure 4.

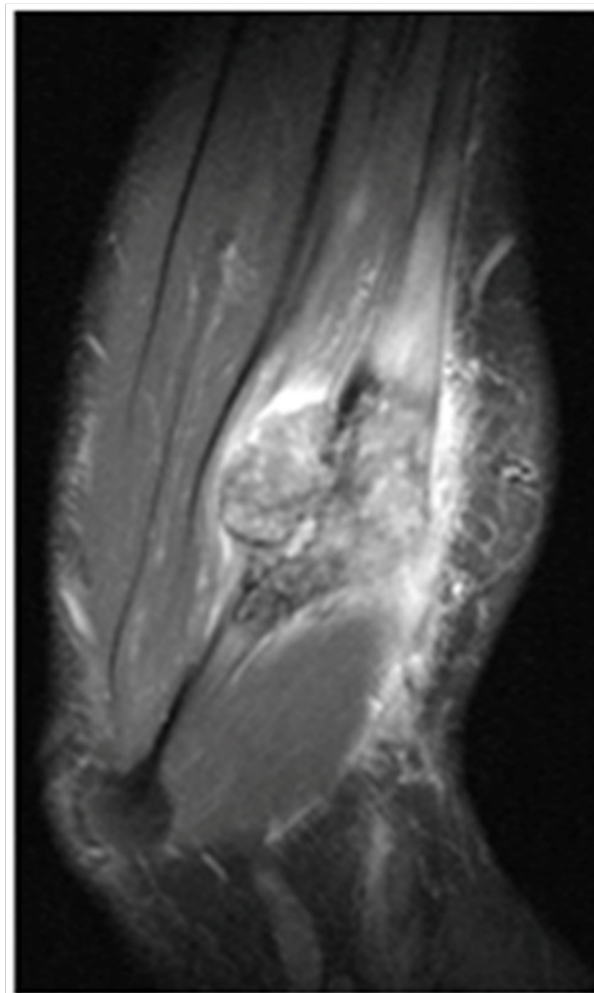


Figure 3 MRI of Epithelioid Sarcoma in the proximal forearm.

Case 5: Spindle cell sarcoma

This elderly gentleman noticed a slow growing lump on his right arm over a period of 2 months with substantial increase in size in the 2 week period prior to review. The initial impression from the GP had been a lipoma but due to the steady rate of growth he was referred to the upper limb clinic. An MRI was in keeping with a soft tissue sarcoma and as such he was sent to the bone tumour service. He was seen in clinic and a biopsy was taken due to the concern that this was a sarcoma. Histology was positive for spindle cell/pleomorphic sarcoma. This was subsequently excised but recurrence had occurred at 6 week follow up with a 2 cm mass at the surgical site. Staging CT scan was clear of metastatic deposits. A wider excision was performed and he had post-operative adjuvant radiotherapy to the surgical field (Figure 5).

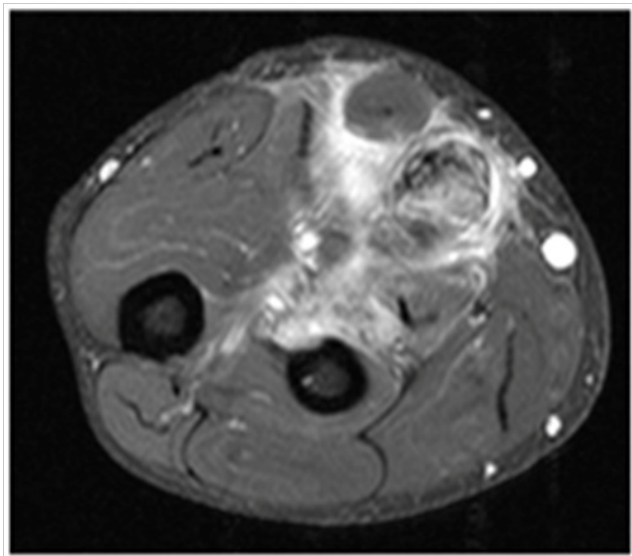


Figure 4 MRI of Alveolar Part Sarcoma in the forearm.

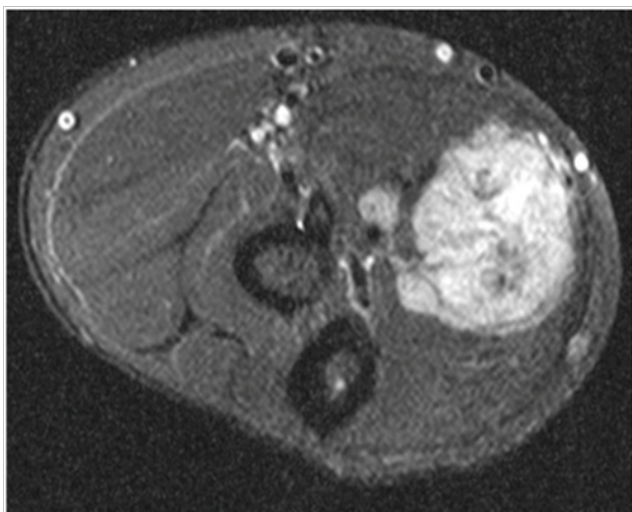


Figure 5 MRI of Spindle Cell Sarcoma in the upper arm.

Case 6: Tumoural calcinosis

A young girl was referred with a 3-year history of a mass overlying her olecranon. This had been previously treated abroad with excision but it had since recurred with a discharging sinus. Treatment with oral antibiotics cleared the infection however due to the persistence of the mass the GP referred to clinic. Clinically it was a non-tender mobile multi-lobulated mass and imaging revealed a calcified lesion confirmed by MRI and referral was made to the tumour centre. Biopsy revealed tumoural calcinosis, a metabolic disorder sometimes associated with Milk-Alkali syndrome or renal disease and a referral was made to the paediatricians for follow up Figure 6.

Case 7: Nodular fasciitis

The final patient was referred with a painless soft tissue swelling on the volar aspect of the distal forearm. It had been present for 9 months with no change in neurovascular status and was found to be mobile.

An MRI revealed a solid appearance of unknown diagnosis and as such he was also referred on for further assessment. He underwent biopsy and it was found to be a benign lesion with features consistent with nodular fasciitis and had conservative management.

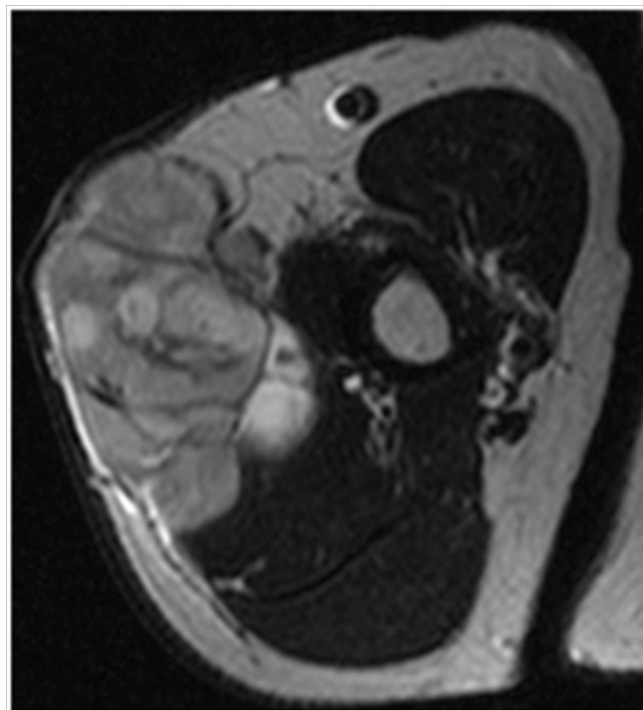


Figure 6 MRI of Tumoural Calcinosis overlying the Olecranon.

Discussion

This set of cases include some important learning points as patients with early referral and diagnosis had better prognosis than those with delayed presentation to the GP or delayed referral to specialist services. A good understanding of common orthopaedic bone and soft tissue tumours also helps ensure appropriate referrals to the bone tumour service with metastatic lesions investigated and treated by specialists within the same hospital if possible.³ Grimer and Briggs⁴ highlighted the referral process and importance of early diagnosis of soft tissue and bone tumours. Criteria for further investigation of soft tissue tumours include lesions greater than 5cm in size, deep-to-deep fascia, increasing in size, painful or recurrence.⁴ This correlates with survival data suggesting that 10 year survival in tumours less than 5cm in size is more than 60% with bigger tumours having worse prognosis. Importantly masses less than 5cm with other concerning features may represent early tumours warranting further investigation.

Conclusion

Consideration for referral of bone tumours to specialist centres is well defined, however the history from the patient often gives many clues as to whether a primary or metastatic lesion is at fault. If there is concern about a primary bone tumour then specialist advice from a bone tumour unit is advised.²

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