



A preventable complication of a slow growing cancer: Lessons learned and a literature review

Alethea Tan¹, Parneet Gill¹, Haitham Kashaba¹, Sally Hales², Fahmy Fahmy¹

¹Department of Plastic Surgery, Countess of Chester Hospital, Chester, United Kingdom,
²Department of Histopathology, Countess of Chester, Chester, United Kingdom

Address for correspondence:
Alethea Tan, Plastic Surgery, Countess of Chester Hospital, Chester, United Kingdom.
E-mail: alethea.tan@doctors.org.uk

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ABSTRACT

Malignant tumors of the hand are rare. Chondrosarcoma presenting in the hand are usually low grade with low risk of metastasis. To our knowledge and following literature review, there are nine reported cases of metastatic chondrosarcoma of the hand worldwide in the English literature, with only two reported cases of axillary lymphadenopathy. We describe a case of an unusual presentation of chondrosarcoma of the left little finger with distant axillary and pulmonary metastases. We also review management issues unique to this patient and reflective learning points to improve care. Our patient could have had a satisfactory outcome if treated early. We recommend referral of suspicious hand lesions to a specialist hand surgeon, for early assessment and management, in order to reduce patient morbidity and mortality.

KEY WORDS: Chondrosarcoma, hand, literature review, metastases

INTRODUCTION

Malignant tumors of the hand are rare. The complex anatomy of the hand involves multiple interconnecting compartments containing vital structures, all at potential risk from tumor invasion. Chondrosarcoma is the most common malignant tumor in the hand, but only accounts for 1.5% of all cases found in the body [1]. Large hyperchromatic bi- or trinucleated cartilage cells on histology are pathognomonic. Radiological features include osteolytic lesions, with or without calcification, commonly affecting the proximal phalanx or metacarpal head but can extend to soft tissue.

Chondrosarcoma presenting in the hand are usually low grade with low risk of metastasis. To our knowledge, there are nine reported cases of metastatic chondrosarcoma of the hand in English literature, with only two reported cases of axillary lymphadenopathy [2-9]. We describe a case of an incidental chondrosarcoma of the left little finger with distant axillary and pulmonary metastases.

CASE REPORT

A 95-year-old Caucasian female presented to the medical team in a district general hospital, following an unwitnessed collapse

secondary to a suspected seizure. A concurrent left sided neck of femur fracture required fixation with a hemiarthroplasty. Significant past medical history included a right parietal lobe meningioma, previous transient ischemic attack, and epilepsy.

A referral was made to the plastic surgery team for a laceration on the dorsum of the patient's left hand, sustained during the fall. Examination of the left little finger was hindered by a bulky dressing. Removal of the dressing exposed a large, odorous, fungating mass, measuring 13 cm in maximal diameter engulfing the entire finger. Ipsilateral axillary lymphadenopathy was noted. Radiographs [Figure 1a] showed a large osteolytic lesion with calcification engulfing all three phalanges of left little finger, loss of cortex definition, and extension into the soft tissue. We also noted an old fracture of the fifth metacarpal neck.

Further history was extrapolated from the next of kin, who described the initial presentation of the lesion over a decade ago. Early medical assessment by the general practitioner resulted in conservative management, in view of the patient's extensive comorbidities and high risk of mortality with a general anesthetic. No further opinion was sought despite an increase in size of the lesion and subsequent functional deterioration. The patient later had several hospital admissions for seizures. However no

formal specialist referral had been made for the lesion on her left little finger. 11 years later, an incidental finding by an observant reviewer led to a thorough assessment, where the patient was deemed fit for surgical intervention.

A ray amputation of the left little finger was performed, with split skin graft reconstruction [Figure 2 a-f] under a regional block. The specimen weighed a massive 468 g [Figure 2c and 3c]. Post-operative regime involved hand physiotherapy exercises following graft check. The wound has healed well, with 100% graft take. Histopathology reported the specimen as a chondrosarcoma [Figure 3 b-c] with evidence of Grade II and Grade III disease. Prompt referral to a regional sarcoma multi-disciplinary team (MDT) and a staging computed tomography (CT) was organized. CT scan [Figure 1b] revealed multiple left axillary lymphadenopathy with a solitary pulmonary nodule suspicious of metastases.

DISCUSSION

Primary bone malignancies are rare, with chondrosarcoma most commonly affecting pelvic bones, proximal humerus and femur. Its incidence peaks between the fourth and seventh decade with a slight preponderance in females. It is commonly associated with enchondromatosis, oller's disease and maffuci's syndrome [1]. Histopathologic characterization consists of cytologic atypia, hypercellularity, binucleated, prominent nucleoli, and large nuclei. Early radiological signs include the presence of osteolytic lesions, bone cortex thickening, and expansion of the bony shaft. Delayed radiological signs include cortical bone rupture, stippled calcification, and gross soft tissue invasion [2].

Microscopic grading of chondrosarcoma has a significant prognostic value, with Grade I disease offering the best prognosis. Enchondromas have a similar histological appearance to low grade chondrosarcomas. Hence, lesions occurring in long bones should be considered chondrosarcoma until proven otherwise. Grades II and III chondrosarcomas have poor

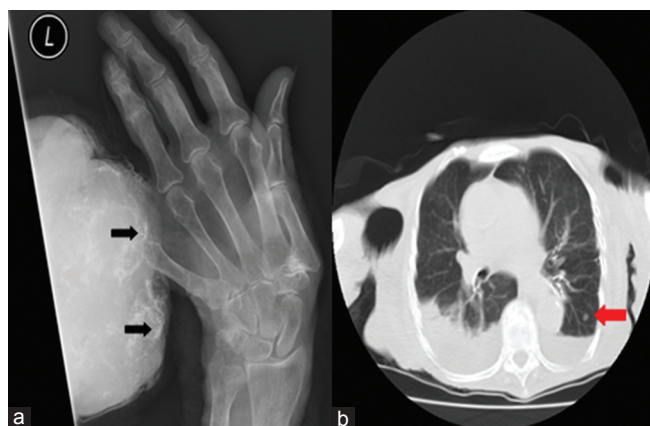


Figure 1: Imaging findings: (a) Plain film showing a grossly enlarged lesion engulfing phalanges of the left little finger, with stippled calcification and soft tissue extension. (b) Computed tomography scan showing solitary pulmonary nodule (arrow) in the left lower lobe and evidence of chronic bilateral effusions

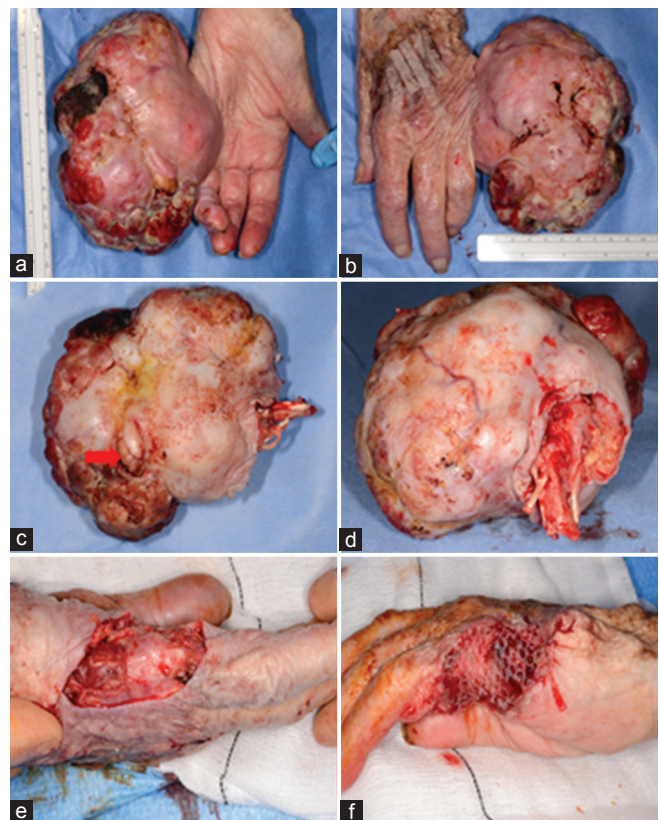


Figure 2: Surgical approach described: (a and b) Fungating ulcerating mass engulfing left little fi nger measuring maximum diameter of 13 cm. (c) Lesion post removal with fingertip and nail plate visible close to its superior pole (arrow), (d) View showing level of amputation comprising base of fi fth metacarpal, flexor tendons, neurovascular bundles, and partial extensor digiti minimi. (e) Post ray amputation showing skin defect and remaining muscle cuff used to provide padding, (f) Lateral and volar views; meshed split skin graft harvested from right thigh used to resurfaced defect.

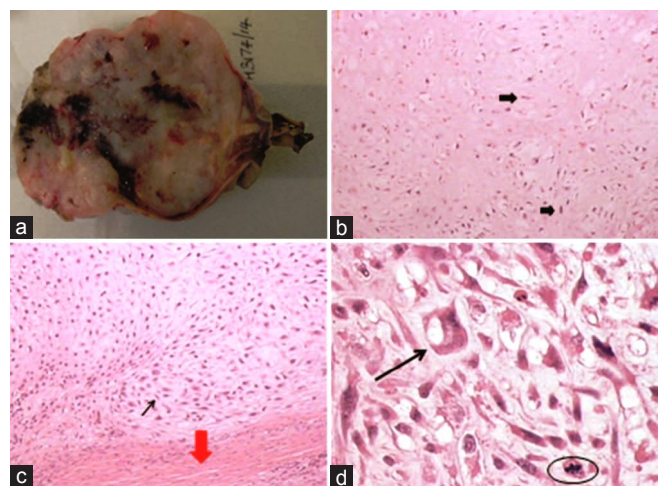


Figure 3: Histopathology section and slides explained: (a) Post calcium stripping, a 3 week process, (b) Presence of binucleate cells (black thick arrows), (c) Likely longstanding enchondroma denoted by increased nuclear pleomorphism (black arrow) surrounded by a fibrous cap (red arrow), (d) Severe nuclear pleomorphism (black arrow) with presence of abnormal mitotic figure (circled), now classifiable as sarcoma variant.

Table 1: Reported cases of hand chondrosarcoma with metastases

Reference	Age (year)	Sex	Primary site	Metastases	Management	Additional interventions
Hamada <i>et al.</i> 2010	48	M	Second metacarpal	Pulmonary metastases	Ray amputation with 'wide excision margins'	
Justis and Dart 1983	47	F	Fifth metacarpal	Pulmonary metastases	Ray resection 5 th digit	Lung metastases treated with chemotherapy
Wu <i>et al.</i> 1980	87	M	Extrasosseous dorsum hand overlying second and third metacarpals	Pulmonary metastases	Excision	Radiotherapy
Dahlin and Salvador 1974	60	M	Lesser multangular	Pulmonary metastases	Excision lesion, ray amputation, amputation forearm	
Dahlin and Salvador 1974	47	M	First metacarpal	Disseminated metastases	Excision X4 and amputation	
Coley and Higgin-Botham 1954	53	F	Fourth metacarpal	Bilateral lung metastases	Partial amputation of hand	
Cruickshank 1945	66	M	Middle phalanx middle finger	Pulmonary and cutaneous metastases	Amputation MP joint	
Allendale 1936	14	F	Fifth metacarpal and multiple phalanges of the hand	Pulmonary metastases	Disarticulation ring finger; curettage and cauterization of phalanx of thumb	
Popovici 1934	28	M	Multiple bilateral tumors of metacarpals, phalanges	Axillary lymphadenopathy, cutaneous metastases, suspected pulmonary metastases	Radiotherapy as primary mode of treatment	

prognostic value, presenting locally with advanced disease and metastases. These are often grouped together as a single entity in the histological classification [10]. The 5 years survival rate for high-grade tumors is 20%. Dedifferentiated chondrosarcoma is a separate group comprising 10% of all chondrosarcomas and has the worst prognosis with a 5 years survival rate of 10% [11].

Chondrosarcoma in the small bones of the hand is rare, accounting for 0.9-1.5% of total reported cases in the body [1]. There are reports of solitary enchondromas undergoing malignant transformation to chondrosarcomas in the hand. A case series showed 78% of the lesions in hand arose primarily with no evidence of previous enchondroma or bone tumor [12]. The majority of chondrosarcoma occurring in the hand are low grade with slow disease progression, rarely metastasizing. To date, there are nine published metastatic hand chondrosarcoma cases available in the English literature, of which two had axillary lymphadenopathies. Only one described axillary lymphadenopathy with dyspnea, another described disseminated disease and the remaining cases were all pulmonary metastases [Table 1].

Current European Society of Medical Oncology guidelines recommend surgical excision with adequate margins for peripheral low-grade disease and wider excision for high-grade disease. Recurrence rate was higher in patients who underwent curettage compared to surgical excision with clear margins [10]. Chondrosarcoma has a poor response to chemotherapy and radiotherapy [12], however, selective palliative skull base chondrosarcomas may respond with excellent local disease control [10].

We present a unique case of an incidental finding of chondrosarcoma affecting the left little finger with associated ipsilateral axillary and pulmonary metastases.

There was no documented histologic diagnosis to indicate a solitary enchondroma prior to the current presentation. Current literature review revealed the largest reported tumor dimensions to be 10 cm × 12 cm [13]. Our patient's tumor spanned the length of her palm (13.5 cm × 9 cm × 7 cm) involving the metacarpal head. Due to the tumor size, ray amputation was carried out to the base of the metacarpal to ensure adequate resection margins. Following histological confirmation of the diagnosis, a staging CT scan was organized and prompt referral was made to the regional sarcoma MDT.

Our unique case report highlights a late incidental diagnosis of an initially small benign tumor of the hand, which underwent malignant transformation. Earlier excision may have spared the digit and prevented spread to the lymph nodes by reducing tumor bulk.

In summary, chondrosarcoma originating from the phalanges of the hand is slow growing, rare, with very low reported metastatic rates. Evidence has increased our understanding of the pathophysiology of chondrosarcoma in the hands, leading to early detection and prompt management. Our patient could have had a satisfactory outcome if treated early. We recommend urgent referral of suspicious hand lesions to a specialist hand surgeon, for early assessment and management, in order to reduce patient morbidity and mortality.

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