Case Report

Journal of Orthopaedic Education
Volume 2 Number 2, July - December 2016
DOI: http://dx.doi.org/10.21088/joe.2454.7956.2216.4

A Rare Case of Secondary Chondrosarcoma of Third Metacarpal

Jain Saurabh*, Jain Anil Kumar, Dhammi Ish Kumar*****

Author Affiliation: *Assistant Professor, Mahatma Gandhi Memorial Medical College, Indore, Madhya Pradesh, India. **Professor and Head, ***Specialist, Department of orthopaedics, University College Medical Science and Guru Teg Bahadur Hospital, Delhi, India

Reprint Request: Saurabh Jain, Mahavir Hospital, A-2, Sch 71, Sec C, Footi Khothi Square, Ring Road, Indore, Madhya Pradesh India 452009.
E-mail: jaindrsaurabh@yahoo.com

Abstract

Hand is very uncommon site for primary malignant bony tumors. Chondrosarcoma, which can form *de novo* as a primary lesion or secondarily from malignant transformation of a pre-existing benign lesion, like enchondroma, is common in pelvis, proximal femur and proximal humerus and rarely occurs in hand. Biologic behaviour of chondrosarcomas in hand whether primary or secondary, which have similar histologic features as chondrosarcomas located elsewhere is not known, but they are locally aggressive lesion, rarely metastasises and had relatively favorable prognosis. We report here such a rare case of third metacarpal chondrosarcoma, suspected to be a secondary chondrosarcoma due to malignant transformation of a benign lesion, which was successfully treated with ray excision. As pre conversion stages are benign, can be differentiated from malignant lesion on clinic-radiologically features and are amenable to salvage surgery, thus regular follow-up are suggested to prevent conversion of benign lesion to malignant lesion.

Keywords: Chondrosarcoma; Hand; Metacarpal; Tumor.

Introduction

Hand is very uncommon site for primary bony tumors, of which enchondroma, a primary benign intramedullary cartilage tumor, located predominantly in the phalanges, is most common [1,2]. Chondrosarcoma, a malignant tumor, which can form *de novo* as a primary lesion or secondarily from malignant transformation of a pre-existing benign lesion, like enchondroma, are further rarer in hand [1-3]. We here report a rare case of third metacarpal chondrosarcoma, in a 43 years male, successfully treated with ray excision, suspected to be a secondary chondrosarcoma due to malignant transformation of a benign lesion. This case emphasis on regular follows up and prompt treatment of the benign lesions, to prevent rare but possible transformation of the benign enchondroma to secondary chondrosarcomas, as benign lesions are amenable to salvage.

Case Report

A 43 years old male, with right hand dominance, and farmer by occupation presented to the orthopaedic department with complains of severe progressive swelling over the dorsum of the left hand, since 6 years. There was mild pain and tenderness present over the swelling. Constitutional symptoms were absent. He had taken no treatment, except some analgesics and had never consulted a doctor.

The swelling was insidious in onset, initially small but had gradually progressed over span of 6 years to its present size of around 6 cm in breadth

and 8 cm in length, localized on the dorsum of the left hand centred over the entire third metacarpal, and around it with dilated tortuous vessel with shinning skin seen over the swelling (Figure 1). On palpation, it was hard in consistency and immobile, whereas due to the swelling the deeper metacarpals could not be palpated. Movements of third metacarpo-phalangeal joints were restricted. Bleeding or ulceration was absent. Investigations revealed no metastasis and no other similar lesions elsewhere in the body.

Radiological examination of the hand showed radiolucent lytic permeative expansile destruction of the entire third metacarapal bone with cortical destruction and soft tissue shadow with areas of speckled calcification. The proximal phalanx and rest of the metacarpals were uninvolved (Figure 2). CT or MRI could not be done.

After the informed consent and consultation with the patient explaining him about the tumor, treatment and recurrence, the patient underwent an extended ray amputation of middle finger of the left hand with removal of the entire third metacarpal along with the surrounding muscles along with carpo-metacarpal k wire fixation for 2nd and 4th metacarpals (Figure 3). Suture removal was done after 2 weeks and k wire removed after 6 weeks (Figure 4).

The resected specimen grossly showed a lobulated mass encircling the metacarpal bone surface. Microscopically showed bony trabeculaes entrapped by solid lobules of hyaline cartilage with neoplastic chondrocytes with chondroid matrix exhibiting pleomorphism, hyperchromasia, mitosis, areas of myxoid degeneration, necrosis and atypical in focal areas. Tumor cells showed infiltration into the adjacent soft tissue. The margins of the tumors with the surrounding soft tissues were clear. The histology result showed a chondrosarcoma grade 3 which had been adequately excised. Patient provided written informed consent for use of patient data and photographs for publication.

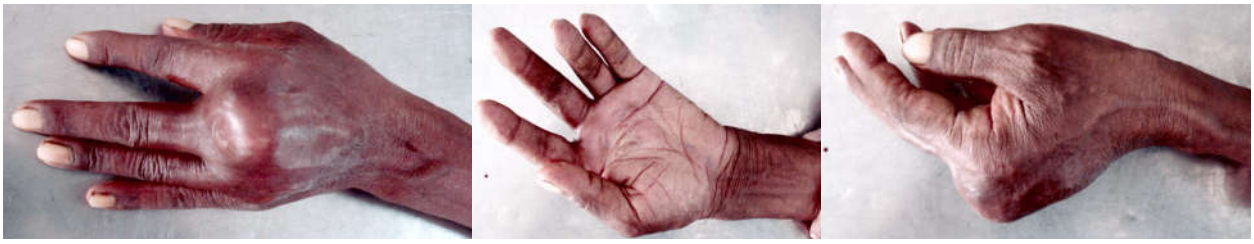

Fig. 1a Fig. 1b Fig. 1c

Fig. 1: Clinical photograph of the patient's hand dorsal **(a),** volar **(b)** and lateral view **(c)** showing the tumour centred over the entire third metacarpal.

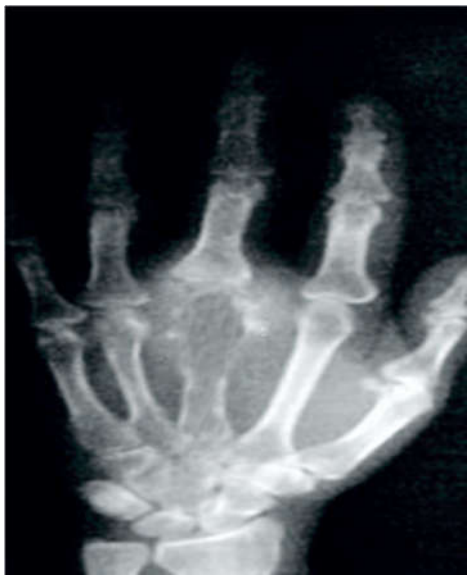

Fig. 2: Pre op X rays of the hand AP view showing radiolucent lytic expansile destruction of the third metacarapal bone with cortical destruction and soft tissue shadow with areas of speckled calcification. With normal proximal phalanx and rest of the metacarpals.

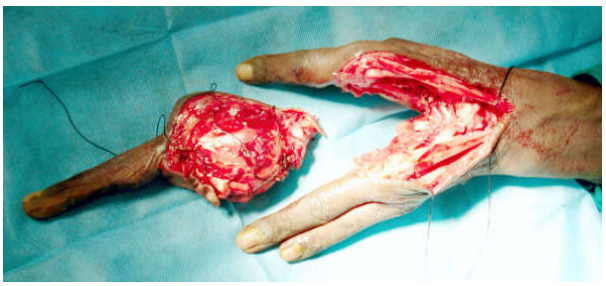

Fig. 3 a:

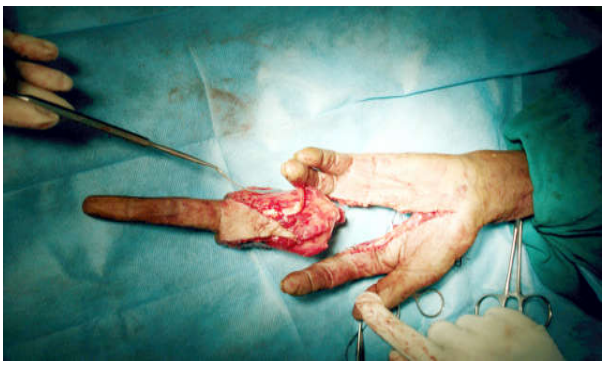

Fig. 3 b:

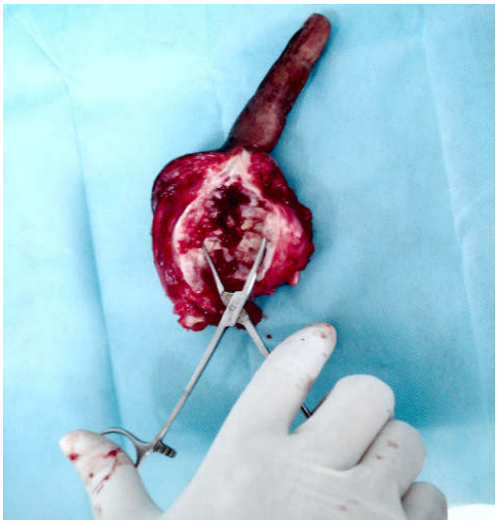

Fig. 3c:

Fig. 3: Intra –op clinical photograph dorsal (a) and volar **(b):** view of the patient after extended ray amputation of middle finger with removal of the entire third metacarpal along with the surrounding muscles. The resected specimen **(c):** showing the entire cortical destruction of the third metacarpal.

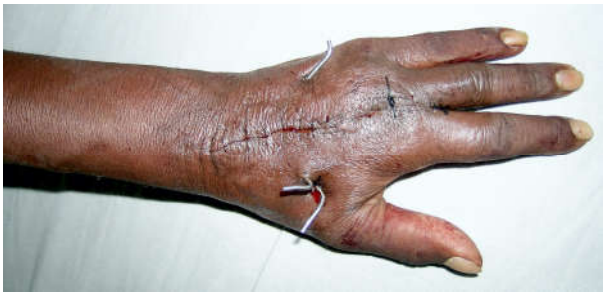

Fig. 4a:

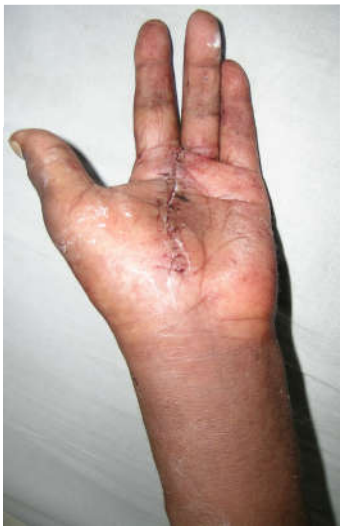

Fig. 4b:

Fig. 4: 2 weeks post of clinical photograph dorsal (a) and volar (b) view of the hand after suture removal showing carpo-metacarpal k wire fixation for 2nd and 4th metacarpals which were removed after 6 weeks.

Discussion

Enchondroma is the most common primary benign skeletal neoplasm of the hand, located predominantly in the phalanges. Chondrosarcoma, a malignant bone tumour with tumour cells that produce cartilage, representing 9%, of primary osseous neoplasms, is common in pelvis, proximal femur and proximal humerus and rarely occurs in hand [1-5]. Short tubular bones of the hand are very rare site, constituting less than 0.5% of all chondrosarcomas [6,7]. Less than 200 cases have been described, of which 50% are located in phalanges, with metacarpal involvement, still rarer [3,4,6-9]. To our knowledge only very small numbers of cases with limited follow-up of metacarpal chondrosarcoma, which are part of a larger cohort, have been described to date.

Chondrosarcoma can form *de novo* as a primary lesion or secondarily from malignant transformation of a pre-existing benign lesion, like enchondroma (more common with multiple enchondromas like ollier or maffucci syndrome than solitary enchondroma), osteochondroma, synovial chondromatosis, chondromyxoid fibroma, periosteal chondroma, chondroblastoma, and fibrous dysplasia [1-3]. Primary chondrosarcoma peaks in adults between 40 and 60 years, whereas secondary chondrosarcomas usually develop at 20 - 40 years [1-3,10]. Similar to most bone tumors, the incidence is slightly higher in males. Race predilection is not significant [6,7]. Differentiating primary or secondary chondrosarcoma clinic-radiologically or histologically is difficult, as they have same clinical and histological appearance showing neoplastic chondrocytes with abundant chondroid matrix exhibiting pleomorphism, hyperchromasia, mitosis, hypercellularity, plump nuclei, myxoid degeneration, necrosis and focal atypical [5,11].

Although clinic-radiologically we can differentiate enchondroma (benign) with chondrosarcoma (malignant) by pain in the absence of a pathological fracture, deep endosteal scalloping of more than two-thirds of cortical thickness, cortical destruction with irregular matrix calcification, soft-tissue invasion and cartilaginous cap larger than 2 cm in a skeletally mature patient in CT scan or MRI, all strongly suggesting chondrosarcoma [1-4,12-15].

The clinical behaviour, treatment and prognosis for patients with chondrosarcoma depends mostly on the size, grade, and location of the lesion [3,8]. Although little is known regarding the biologic behaviour of chondrosarcomas in hand whether primary or secondary, which have similar histologic

features as chondrosarcomas located elsewhere, but both behaves as a locally aggressive lesion, rarely metastasises and had relatively favorable prognosis [3,8,10]. Since in our case chondrosarcoma, was grade 3, located at expendable location, we did extended ray amputation of middle finger of the left hand with removal of the entire third metacarpal along with the surrounding muscles for our case, to optimise local disease control and reduce the frequency of metastatic spread and recuurence [1,3,5,8].

Our case was histologically proven chondrosarcoma grade 3, but since no previous imaging was available, dilemma exists, whether it was a primary or secondary chondrosarcoma. In our case, due to rarity of location in hand, which was central intramedullary in location, that erode the cortex and invade the surrounding soft tissue and long slow progressive history for several years before medical attention, along with other clinico-radiological feature, we suggest our case to a rare transformation of a solitary enchondroma into a secondary chondrosarcoma. Whether a solitary benign enchondroma has the potential to give rise to a secondary chondrosarcoma is difficult to determine. But if we can be diagnose the potential enchondroma earlier than conversion to secondary chondrosarcoma, the salvage procedure can be done as the pre conversion stages are benign and ameneable to salvage surgery. But since the incidence of this conversion is very rare and treatment of primary or secondary chondrosarcoma is also not different and is the same, prophylactic treatment of asymptomatic enchondromas is not recommended.

Conclusion

It is important to regularly follow the asymptomatic or potential enchondromas by radiographs, CT scan or MRI, and treat them adequately so as to prevent conversion of benign lesion to malignant secondary chondrosarcoma. Further long-term follow-up of the operative site and the chest is also imperative so that treatment can be initiated promptly in the event of a recurrence or metastasis.

Conflict of Interest

Nil

Acknowledgements

Nil

References

1. Hatori M, Watanabe M, Kotake H, Kokubun S. Chondrosarcoma of the ring finger: a case report and review of the literature. Tohoku J Exp Med. 2006 Mar; 208(3): 275-281.
2. Essadki B, Belabidia B, Fikry T, Moujtahid M, Lamine A, Essadki O, Zryouil B. Chondrosarcoma of the hand. Diagnostic and therapeutic discussion (three case reports). Chir Main. 2001 Apr; 20(2): 164-71.
3. Spiegel K, Joseph E. A rare appendicular skeleton chondrosarcoma. SA J Radiol. 2007 Nov; 11(2): 28-31.
4. Dahlin DC, Salvador AH . Chondrosarcomas of bones of the hand and feet. A study of 30 cases. Cancer. 1974 Sep; 34 (3): 755-760.
5. Thanigai S, Douraiswami. Chondrosarcoma of metacarpal – A rare presentation. J Clin Orthop Trauma. 2013 Jun; 4(2): 85–88.
6. Kanjirakkadu PK, Moothiringode CS, Ramani KC, Puthoor D. Periosteal chondrosarcoma of metacarpal bone: Report of a rare entity. Australas Med J. 2012 Nov; 5(11): 589-92.
7. Wirbel RJ, Remberger K. Conservative surgery for chondrosarcoma of the first metacarpal bone. Acta Orthopaedica Belica 1999 Jun; 65(2): 226-9.
8. Murphey MD, Walker EA, Wilson AJ, Kransdorf MJ, Temple HT, Gannon FH. From the archives of the AFIP imaging of primary chondrosarcoma: Radiologic-pathologic correlation. Radiographics. 2003 Sep-Oct; 23(5): 1245-78.
9. Mittermayer F, Dominkus M, Krepler P, Schwameis E, Sluga M, Toma C, Lang S, Grampp S, Kotz R. Chondrosarcoma of the hand. Clin Orthop. 2004 July; 424: 211–215.
10. Bovee JV, Vander Heul RO, Taminiau AH, Hogendoorn PC. Chondrosarcoma of the phalanx: a locally aggressive lesion with minimal metastatic potential: a report of 35 cases and review of the literature. Cancer. 1999 Nov; 86(9): 1724-32.
11. Springfield DS, Gebhardt MC, Mc Guire MH. Chondrosarcoma: a review. J Bone Joint Surg (Am). 1996 Jan ; 78(1): 141-149.
12. Muller PE, Durr HR, Nerlich A, Pellengahr C, Maier M, Jansson V. Malignant transformation of a benign enchondroma of the hand to secondary chondrosarcoma with isolated pulmonary metastasis. Acta Chir Belg. 2004 Jun; 104 (3): 341-344.
13. Murphey MD, Flemming DJ, Boyea SR, Bojescul JA, Sweet DE, Temple HT. Enchondroma versus chondrosarcoma in the appendicular skeleton: differentiating features. Radiographics. 1998 Sep-Oct; 18 (5): 1213-1245.
14. Demireli P, Çavdar DK, Vural S, Ovali GY, Özalp T. Secondary chondrosarcoma arising from a solitary enchondroma at the index finger of the right hand:

a case report. Turk J pathology. 2006 Jan; 22(1): 53-56.

15. Muller PE, Durr HR, Wegener B, Pellengahr C, Maier M, Jansson V. Solitary enchondromas: is radiographic follow-up sufficient in patients with asymptomatic lesions? Acta Orthop Belg. 2003 Apr; 69(2): 112-118.