Malignant transformation of solitary phalangeal enchondroma to high grade chondrosarcoma - A case report

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Abstract

Introduction: Chondrosarcoma (CS) is a malignant neoplasm of mesenchymal origin characterized by the formation of cartilaginous matrix by neoplastic cells. It may arise as a primary lesion or occur secondarily at the site of a previous benign lesion such as enchondroma. Primary CS is rare in the hand with a frequency of <0.5–3.2% of all CS. Malignant transformation of a benign solitary enchondroma to secondary CS, especially in the hands, is extremely rare. This transformation is seen more often in multiple enchondromatosis than in solitary lesions, but even this event is rare in the hands. Here, we report a case of solitary enchondroma of proximal phalanx of middle finger initially treated with intralesional curettage and bone grafting, later recurring to high grade (Grade 3) CS.

Case Report: We report the case of a 44-year-old lady presenting with pain and progressive fusiform swelling of the entire right middle finger of 4 months duration. She had a similar history 5 years back for which she had undergone intralesional curettage and bone graft substitute application. Curetted specimen was reported as enchondroma. Radiographical evaluation showed an expansile, intramedullary, and lytic lesion severe cortical destruction over proximal phalanx of the right middle finger. Magnetic resonance imaging detailed the soft-tissue component. She underwent a core needle biopsy which reported as low-grade CS arising from enchondroma. Resection of middle ray including distal 2/3 of 3rd metacarpal was performed. Excised specimen was reported as high-grade CS (Grade 3) with tumor free margins. At 2-year follow-up, she is symptom free without any local recurrence or distant lesions.

Conclusion: Diagnosis of CS in the hand can pose problems to clinicians and pathologists alike. To definitely set a diagnosis of CS in the hand, comparison of histological and radiological finding is paramount. CS are malignancies resistant to both radiation and chemotherapy. CS of the hand is characterized by local recurrence and very low metastatic potential. Hence, wide excision is the recommended approach.

Keywords: Phalanx, Enchondroma, Chondrosarcoma, High grade, Amputation

Introduction

Chondrosarcoma (CS) is a rare malignant tumor of bone, formed from cartilage. Enchondromas are the most common benign cartilaginous bone tumors arising in the medullary cavity of small bones of the hand [1]. In contrast, CS commonly occurs in the pelvis, proximal femur, and humerus [2], but are uncommon in the small bones of the hand [3]. The diagnosis of CS in the hand can pose problems to clinicians and pathologists alike. Because enchondromas are commoner at this site [3, 4] clinicians understandably have a low index of suspicion of malignancy. The possibility of CS arising from solitary pre-existing enchondroma needs to be considered although only a few such cases have been documented [5]. Distinguishing CS from enchondromas in the tubular bones of the hand is difficult [4] because the histologic criteria are not well-defined [6]. This is clinically relevant because CS of the hand requires prompt and more radical treatment as they are locally aggressive [3, 7]. Here, we report a case of a middle aged lady who had undergone intralesional curettage for enchondroma proximal phalanx 4 years back, presenting with local recurrence of pain and swelling. Results of needle biopsy followed by excision biopsy were suggestive of high-grade CS arising from an enchondroma.

Case Report

A 44-year-old lady presented to the orthopedic clinic with complaints pain and swelling over the right middle finger of 4 months duration. She had a similar history 5 years back, which on radiographic evaluation was suspicious of enchondroma for which she underwent intralesional curettage and bone graft substitute application. Curetted specimen was reported as enchondroma. Present swelling was progressive in nature, associated with intermittent, dull aching pain, and without any diurnal variation. No swellings elsewhere in the body noted. No comorbidities or constitutional symptoms noted. Clinically, the patient had a fusiform swelling involving the entire proximal
phalanx of the right middle finger measuring 6.5 cm × 4.5 cm, tender, bony hard in consistency, and immobile. The proximal phalanx could not be palpated separately off the swelling. Skin over the swelling was stretched and shiny.

Movements of third metacarpophalangeal and proximal interphalangeal joints were restricted. Radiograph (Fig. 1) showed an expansile, lytic, lesion involving the entire proximal phalanx of middle finger with severe cortical destruction, bone graft substitute in situ, and soft-tissue effacement. Magnetic resonance imaging (MRI) (Fig. 2) confirmed the severe cortical destruction and soft-tissue effacement. A multidisciplinary tumor board was conducted in which the patient was educated about the tumor and explained about the treatment plan and the recurrence rates. After an informed consent from the patient, we decided to do a radical resection of the tumor. The entire middle ray along with distal 2/3rd third metacarpal was resected followed by webspaces reconstruction (Fig. 4).

Grossly, the amputated specimen (Fig. 5) showed a globular mass encasing the entire proximal phalanx, with cut section showing brown, gelatinous, and myxoid material. Microscopically (Fig. 6), phalangeal bone invaded by cartilaginous neoplasm composed of chondroid lobules with moderate to focal hypercellularity of neoplastic chondrocytes exhibiting moderate nuclear pleomorphism, hyperchromatism, and binucleation with chondroid stroma. Few lobules with marked cellularity and spindling of chondrocytes, nuclear atypia and hyperchromatism, and invasion of adjacent cortical bone and soft tissue. The margins of the tumor with surrounding soft tissues were clear. Diagnosis of high-grade CS arising on the background of enchondroma was made.

She was followed up in the outpatient clinic on 6 weeks, 3 months, 6 months, and at 1-year post-surgery. At 2-year follow-up, she is asymptomatic, without any clinical or radiological evidence of local recurrence, or distant metastasis.

Discussion

CS of the hands has been rarely reported. Nigrisoli et al. [8] in a series of 506 CS found only four cases (<1%) in the hand, and Unni [9] found that only 1.5% of 635 CS were in the hand. Proximal phalanx was most often affected [3]. Malignant transformation of a benign solitary enchondroma to secondary CS, especially in the hand, is extremely rare. Such a change occurred in only two out of 112...
In the diagnosis of CS localized in phalanges, cortical destruction, soft-tissue involvement, periosteal reaction, and intralesional calcification (pop-corn like lesion) are signs of potential malignancy in direct radiography [10, 11]. Particularly, the differentiation of low-grade CS and enchondromas is challenging for both clinicians and pathologists. MRI is the basic radiologic evaluation method for the definition of pathologic and normal bone margins, intramedullary, extraosseous, and soft-tissue invasion. The definitive diagnosis of the patients is established with clinical examination, conventional radiography followed by MRI and biopsy and histopathologic evaluation. Our patient had all the clinical and radiographic features suggestive of CS, and histopathology confirmed the diagnosis.

CS is malignancies resistant to both radiation and chemotherapy [12]. Surgical treatment is the only mean available. Different approaches are used depending on tumor grade. For this reason, it is important to correctly establish the grading of the malignancy. CS is often separated into three histological grades: Atypical cartilagenous tumor (Grade 1), intermediate (Grade 2), and high (Grade 3). The higher the grade, the more likely it is that the tumor will metastasize [13].

Metastasis of the hand CS is rare [3]. In their series, Bovee et al. [3] found no metastasis in 28 of 35 hand and foot CS patients. They reviewed the literature until 1999 and detected only two of 84 patients that distant metastasis occurred and they concluded that phalangeal CS was locally aggressive and had very low metastatic potential. Furthermore, Mankin [7] reported that hand CS had a more benign course than that of other involvements in skeletal system. Patil et al. [11] in their 23-patient-series of metacarpal and phalangeal CS reported no distant metastasis. In our study, there was no distant metastasis during the follow-up period.

Intralesional curettage has been increasing the local recurrence rates [10, 11]. There are 11–50% local recurrence rates reported in the literature [11]. While Bovee et al. [3] did not observe any local recurrence after ray amputation for the 28 of the 35 patients in their series; they reported local recurrence in 10 patients who underwent marginal resection. In another case reported by Demirel et al. [14], a solitary enchondroma was located at the index finger of the right hand. Seven months later, the tumor recurred and was destructive, and amputation material was diagnosed as an intermediate grade CS arising secondary to the former solitary enchondroma. Wide excision of the tumor was done to avoid local recurrence or metastasis.

Amputation of the affected ray or metacarpal is the correct treatment to avoid local recurrence or distant metastasis [3, 10, 11]. Ray amputation was performed for our case to prevent local recurrence or distant metastasis due to extensive cortical destruction, soft-tissue involvement, and needle biopsy diagnosis of CS.

Conclusion

CS involving the hand is very rare. Moreover, very few cases of enchondroma transforming into a CS are documented in the literature. CS located in the hands is a diagnostic and therapeutic challenge. Distinction between benign and malignant lesions is not always easy. During diagnosis, clinical examination findings and conventional radiologic studies must be evaluated well, and MRI studies must be performed to evaluate soft-tissue involvement and support the diagnosis of CS. These neoplasms are believed to have a more benign behavior compared to CS located elsewhere: They are locally aggressive, but show poor tendency to metastasize. CS requires a prompt and more radical treatment than enchondroma. Wide excision is recommended to avoid local recurrence or metastasis.

Clinical Message

The differentiation of phalangeal CS from enchondromas is rare and difficult both for pathologists and the clinicians. If there is any doubt in the radiographic and clinical evaluation, MRI and histopathologic evaluation must be performed. Limited and
marginal interventions lead to a rise in local recurrences. The proper and definitive treatment is amputation.

References


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