

Thumb polydactyly associated with secondary chondrosarcoma arising from a solitary enchondroma: A rare case report

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ABSTRACT

The most common type of polydactyly in the hand is thumb polydactyly. Enchondromas are benign cartilaginous tumors and usually present in phalanx of hands. Transformation of enchondromas into chondrosarcomas is very rare, which is commonly associated with multiple enchondromatous lesions. Association of thumb polydactyly and secondary chondrosarcoma arising from a solitary enchondroma is even rarer. We present a 50-year-old female patient with right thumb polydactyly associated with secondary chondrosarcoma arising from a solitary enchondroma. To our knowledge this association has not been described previously.

Key words: Chondrosarcoma, enchondroma, polydactyly

INTRODUCTION

The most common type of polydactyly in the hand is thumb polydactyly. It arises from excessive cell proliferation and disturbed cell necrosis of preaxial ectodermal and mesodermal tissues during early gestational periods.^[1] It has a sporadic occurrence with an incidence of 8 in 100,000 in both black and white populations.^[2] Isolated polydactyly is usually autosomal dominant, while syndromic polydactyly is often autosomal recessive.^[3] In order to categorize different types of polydactyly and to guide respective surgical procedures, the Wassel classification is universally accepted.^[4]

The most common primary benign bone tumors of the hand are enchondromas involving phalanx and they rarely transform into chondrosarcomas. Transformation

of enchondromas into chondrosarcomas is very rare which is commonly associated with multiple enchondromatous lesions.^[5,6]

A histopathological differentiation between enchondroma and chondrosarcoma is usually difficult. For differential diagnosis, radiological as well as pathological findings are required.^[6-8] To avoid local recurrence or distant metastasis this type of categorization is necessary because chondrosarcomas of the hand require wide excision.^[8]

We report a rare case of right thumb polydactyly associated with secondary chondrosarcoma arising from a solitary enchondroma.

CASE REPORT

This patient, a 50-year-old female presents with duplication of right thumb since birth and swelling and pain lasting for 6 months at ulnar side thumb of the right hand. Earlier she was able to use her hand well for grasping and pinching small object by her ulnar side thumb. Following development of swelling and pain she experienced work related difficulties in her daily activities. Physical examination revealed right sided preaxial hexadactyly with thumb duplication with hard, tender swelling in proximal

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phalanx of ulnar side thumb. Radial side thumb was slightly hypoplastic with normal sensation, but the patient was not able to move it independently.

Radiographs showed extra thumb containing duplicated phalanges and articulating on the common metacarpal Type IV polydactyly, according to the Wassel classification [Table 1]. Plain radiograph of the swelling showed well-demarcated lytic lesion with intact cortex suggestive of benign nature.

Excisional curettage and biopsy of the swelling revealed to be an enchondroma. Six months later, the swelling recurred at the same location. Plain radiograph of the proximal phalanx of the ulnar side thumb showed expansion of bone, cortical destruction, and soft-tissue extension with invasion of adjoining metacarpal bone [Figure 1]. Computed tomography (CT) of the thumb was suggestive of chondrosarcoma [Figure 2]. The duplicated thumb was amputated at carpometacarpal joint. Healing was good with no occurrence of surgery related problems. Histopathologically, surgical margins were negative. Microscopically, section from thumb showed lobules of

chondrocytes of variable size separated by fibrous stroma. Permeation of bony trabeculae seen focally. These findings were suggestive of chondrosarcoma [Figures 3 and 4]. After that, smears of excisional curettage and radiographs of the primary lesion were reevaluated and comparison was done with those of the secondary lesion. Following reevaluation and comparison, the diagnosis of well differentiated chondrosarcoma secondary to solitary enchondroma was made. Since the surgical margins were negative, pollicization was planned.

DISCUSSION

The most common congenital digital anomaly of the hand and foot is polydactyly. The different types of polydactyly are preaxial, central, and postaxial. Preaxial polydactyly is the most common type in which first digital ray is duplicated.^[9] In order to show the skeletal elements in the rudimentary digit, radiographs of the affected parts are recommended. The exact operative procedure depends on to what extent digit is angulated and surface area of articulating surface.^[10] Appropriate surgical procedure is performed to create a normal thumb usually around 1 year of age.^[11]

It is very important to simultaneously evaluate and treat the skin, nail, bone, and the ligaments in order to obtain a good result of reconstruction and to minimize both the complications and the need for subsequent operations.^[12]

Our patient had preaxial polydactyly since birth. Because this patient did not experience work related difficulties, therefore she did not receive any treatment. The patient presented when she developed swelling and pain in her ulnar side thumb.

Table 1: Wassel classification

Wassel type	Anatomic descriptions
I	Bifid distal phalanx
II	Duplicated distal phalanx sharing common distal interphalangeal joint articulation
III	Bifid proximal phalanx
IV	Duplicated proximal phalanx sharing common proximal interphalangeal joint articulation
V	Bifid metacarpal
VI	Duplicated metacarpal sharing common carpal articulation
VII	Triphalangeal thumb

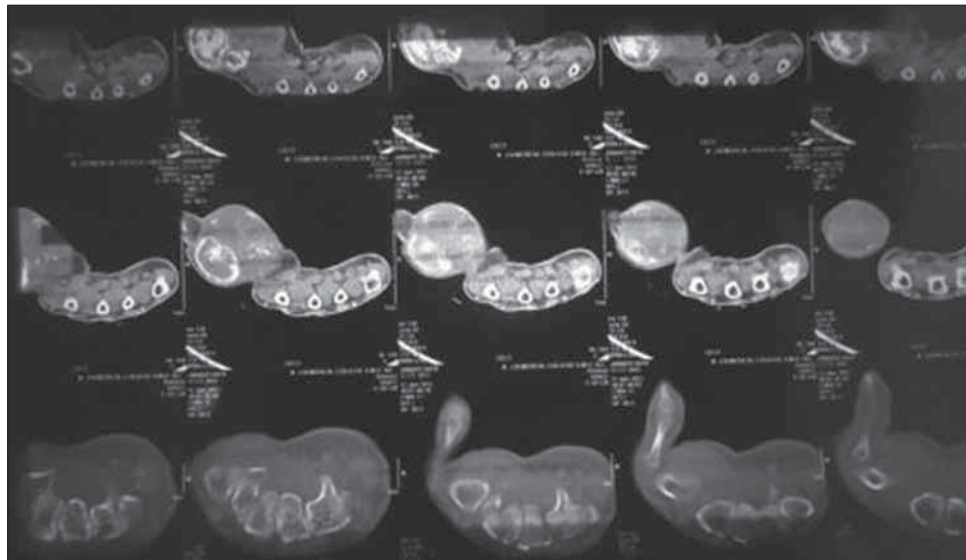


Figure 1: Computed tomography of the thumb suggestive of chondrosarcoma

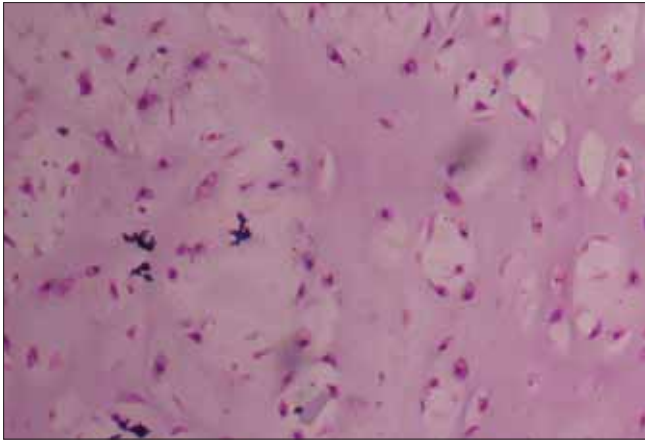


Figure 2: Low power view of well differentiated chondrosarcoma with nuclear atypia

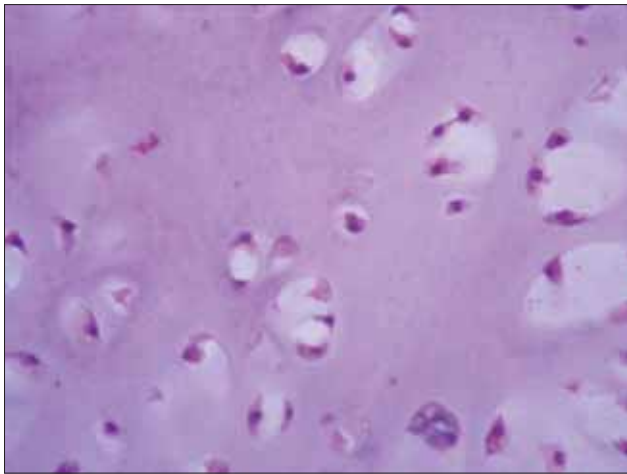


Figure 4: High power view of well-differentiated chondrosarcoma with nuclear hyperchromatism and two or more cells per lacuna

Enchondromas are cartilaginous tumors of benign nature and they are usually present in phalanx of the hands.^[6] Transformation of enchondromas into chondrosarcomas is very rare which is commonly associated with multiple enchondromatous lesions.^[5,7,8] In our patient transformation occurred because of malignant degeneration of monostotic enchondroma to chondrosarcoma. Though this phenomenon is rare, but has been well described in the literature.^[13]

Due to similar clinical findings, it is very difficult to differentiate between enchondroma and chondrosarcoma.

Radiograph showing cortical destruction, periosteal reaction, and soft tissue mass strongly suggest the diagnosis of chondrosarcoma.^[6-8] Therefore, it is used as an important tool in differentiating these two entities.

CT scan in our patient showed features of chondrosarcoma of proximal phalanx of ulnar side thumb invading adjacent metacarpal bone. Therefore, we performed amputation at the carpometacarpal joint [Figure 5].



Figure 3: Plain radiograph showing expansion of bone, cortical destruction, and soft tissue extension with invasion of adjoining metacarpal bone



Figure 5: Postop X-ray of the patient's thumb

Destruction of cortex and invasion of soft tissue are seen in chondrosarcomas.^[5,8] Permeation of bony trabeculae is also seen. These features were present on histopathological examination of the specimen of the thumb in our patient.

For excisional curettage of enchondroma, dorsal or lateral approach can be used. First of all biopsy should be done by limited exposure and diagnosis confirmed by frozen section analysis. After confirming the diagnosis by frozen section analysis, a wider exposure is used. But in our center, facility of frozen section analysis is not available. So it could not be performed in this case.

In order to fill the defect after excision and curettage, iliac crest or distal part of radius has been most commonly used.^[14,15] But recently, many researchers have found good results by simple curettage without filling the defect.^[16]

Most of the of chondrosarcomas of hands are of high histologic grade.^[8] Chondrosarcomas of the hand is less aggressive in nature contrary to other areas of the body.^[8] However, chondrosarcoma of the hand requires early radical treatment. To avoid local recurrence or metastasis wide excision is recommended.^[8]

CONCLUSION

It can be concluded that differentiation between enchondroma and chondrosarcoma by using radiological as well pathological modalities is very important, because chondrosarcoma of the hand requires early radical treatment than enchondroma. Following excisional curettage of enchondroma, patient should be kept on regular follow-up keeping in mind the transformation of enchondroma to chondrosarcoma.

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