

Thumb Distal Phalanx Giant Enchondroma. A Case Presentation and Literature Review

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ABSTRACT

Background: Enchordoma of the distal phalanx of the thumb is extremely rare.

Case presentation: We report a case of 31-year-old man who presented with a pathological fracture of the left thumb. Imaging evaluation revealed a lytic lesion and surgical curettage with bone graft was performed after fracture healing. Histological examination confirmed the diagnosis of enchordoma. The postoperative period was uncomplicated without signs of recurrence.

Conclusion: Lytic lesions in the thumb are uncommon occurrences and necessitate a comprehensive examination to determine their potential causes. Given the significant functional role of the thumb compared to other fingers, it is crucial to undergo radiological assessment and further investigation of these lytic lesions.

Keywords: enchordoma, thumb, distal phalange.

INTRODUCTION

Enochordoma is the most common benign tumor of the hand, occurring mostly on the proximal phalanges and metacarpals (1). Enchondromas usually occur in the first to fourth decades of life, affecting the ulnar-side tubular bones of the hand. They may present incidentally or with pain, swelling and stiffness following a pathologic fracture, in

about 40-60% of cases. Histopathologic examination reveals hyaline cartilaginous lesions and bone cells (1). Lytic lesions of the thumb, especially in the distal phalanx, are rare and need increased awareness, since the function of the hand depends on their accurate diagnosis and specified treatment (2). According to the existing literature, enchondromas of the distal phalanx of the thumb are extremely rare (3). The following case report presents the diagnostic procedure

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and management of an enchondroma of the distal phalanx of the thumb, a location where successful treatment is of paramount importance for the function of the hand.

CASE REPORT

A 31-year-old right-dominant male presented to our outpatient clinic with a painful and swollen thumb of his left hand following a low-energy trauma. The patient recalled that edema and nail changes had been present for a year prior to examination. Clinical examination revealed a limited range of motion of the PIP and tenderness on palpation.

Radiographs revealed a well-defined lytic lesion of the distal phalanx of the thumb, accompanied by a pathological fracture (Figure 1). A spica cast was applied and both computed tomography (CT) evaluation and magnetic resonance imaging (MRI) were performed. The CT scan showed a 1.5 cm lytic lesion with sharply defined margins, which were disrupted due to fracture and cortical thinning expanding over 2/3 of the distal phalanx, without matrix mineralization (Figure 1). The MRI revealed low signal in T1 sequence with a peripheral enhancement of gadolinium. Moreover, T2 showed a high signal lesion and soft tissue edema, possibly due to



FIGURE 2. Magnetic resonance imaging showing low signal in T1 frequency

fracture hematoma (Figure 2). Aneurysmatic cyst, giant cell tumor, chondrosarcoma and enchondroma were all included in differential diagnosis. Consequently, an open needle aspiration via tru cut biopsy was decided, but the histological results were non-diagnostic.

The spica cast was kept for five weeks until the fracture healed. As the cavity was still radiologically evident, the patient was scheduled for surgical intervention. The operation was performed under general anesthesia with a tourniquet application. A lateral approach protecting the neurovascular bundles and flexor tendons was used. A thorough curettage of the lesion through the lateral wall of the phalanx was performed. The tumor was removed by spoon and the cavity was filled with autologous iliac bone graft. Thereafter, the skin was sutured and, after wound closure, a thumb spica cast was applied. The patient was discharged from hospital on the same day. The open biopsy revealed islands of mature hyaline cartilage into normal bone cells, characteristics that indicated enchondroma.

The hand was immobilized for two weeks and the X-ray was repeated in a month. The cavity was filled with bone. The swelling subsided and the patient had painless full range of motion and returned to work in 15 days. How-



FIGURE 1. Enchondroma of distal phalanx of the thumb (X-ray and CT evaluation)



FIGURE 3. Lesion healed, with no signs of recurrence, one year postoperatively

ever, the patient complained of persistent donor site pain for three months postoperatively.

One year later, the patient had a DASH score of 96, with a full, painless range of motion, although a slight deformity of the phalanx was evident. X-rays and CT did not reveal any sign of recurrence, and the lesion was completely healed (Figure 3). On his last follow-up visit, two years postoperatively, the patient had a fully functional hand with no complaints.

DISCUSSION

Although enchordomas are common tumors of the hand, they are extremely rare on distal

phalanges specifically of the thumb (4). Generally, enchordomas located on the distal phalanx represent only 10% of cases, and the involvement of the thumb is exceptional. Furthermore, they are usually located in the diaphysis of hand bones, which is in contrast to the metaphyseal region of long bones (5). The lesion often presents as a pathologic fracture following a low-energy trauma or as an avulsion fracture of the deep flexor tendon (6-9).

Differential diagnosis includes giant cell tumor, enchondroma and aneurysmatic cyst. Moreover, special attention should be paid to differential diagnosis of chondrosarcoma, which is usually presented as a painful lesion with locally aggressive features on X-rays and MRI. More specifically, development of lucencies, calcification, periosteal reaction and soft-tissue mass are radiological features of chondrosarcoma (10). Giant cell tumor often presents with persistent pain without a fracture, bone devastation and soft tissue extension. Frequently, aneurysmatic cyst occurs in proximal phalanxes and has diaphragms on radiological assessment. Aneurysmatic cysts usually occur in children and due to low frequency in adults are believed to resolve spontaneously. In our case, the patient reported being asymptomatic before the fracture.

The treatment of enchondromas of the hand may vary and is largely symptomatic. In case of asymptomatic, painless enchordomas, conservative treatment and evaluation with serial plain radiographs is recommended (18). Indications of surgical treatment are pathological fractures and

Author	Sex	Age (years)	Initial presentation	Surgery	Result
Gomez-Fernadez (11)	F	21	Progressive pain and deformity	Curettage of the lesional cavity and filling with autologous bone graft, k-wire	Full healing without recurrence
Iannicelli (12)	F	35	Painless, swelling distal phalanx, limited range of motion	Curettage of the lesion with bone grafting	Complete healing without recurrence
Prietzel (13)	M	56	Swelling, nail deformity	Curretage, allograft	Complete healing without recurrence
Yamamoto (14)	F	81	Dorsum nodule, asymptomatic, nail deformity	Curretage, hydroxyapatite	Complete healing without recurrence
Milliez (15)	M	32	Swelling	Resection	Complete healing without recurrence
Bachoura (16)	F	32	Fracture	Consolidation	Missing in follow up
Lu (17)	F	56	Clubbed finger, infection	Drainage and calcium phosphate cement	Complete healing without recurrence

F=female; M=male

TABLE 1. Cases of enchodromas of the thumb

symptomatic lesions. Complete curettage and filling of the defect via autologous bone graft even with hydroxyapatite or calcium phosphate cement have been described as surgical options (19, 20). The placement of bone graft or bioactive glass has been recently investigated, but no significant difference existed in the published studies (21). A study demonstrates that bone graft is essential only in cases where the cortex is thin or the lesion is near the articular surface (22).

Yamamoto *et al* use hydroxyapatite and Yasuda injects calcium phosphate cement into the bone defect. The cement approximates the strength of intact bone; however, it is unknown if this correlates clinically to a quicker return in range of motion or healing. In cases of massive involvement of the phalanx or metacarpal, Bickels complements tumor curettage with the intramedullary placement of two Kirschner wires, filling the cavity with bone cement (PMMA) (14, 19, 20). In our case, no use of k-wires was required due to the high stability of the lesion and autologous bone graft was preferred.

Adjuvant therapies are often used to reduce the recurrence rate during the curettage and bone grafting procedure. Two common modalities include instillation of alcohol and the use of a high speed burr (23).

Controversy exists regarding the treatment for an enchondroma with an associated pathologic fracture. In some studies, the functional outcome after surgical treatment with or without pathologic fracture remains similar (24). In more complicated cases, such as avulsion fracture of the flexor tendon or instability of PIP joint, fusion and tenodesis are recommended (25). In the literature, the timing of surgery in case of fracture presence is not yet clear. In our case, surgery was performed after the healing of the fracture in order to avoid instability on the fracture site postoperatively, and the use of Kirschner wires provided better curettage and bone graft placement. Furthermore, it allowed a better surgical approach and wound healing as the soft tissues did not present swelling. The purpose of surgical treatment is to avoid pathologic fractures and deformity of the thumb.

Enchondroma of the distal phalanx of the thumb is extremely rare and only few cases have been reported in the literature so far. Most of the authors prefer bone graft rather than hydroxy-

apatite or calcium phosphate cement. Full healing without recurrence is demonstrated in cases of enchondromas (11-17).

The radiological features of the enchondroma are usually characteristic, being a well-defined lytic lesion, with the presence of small calcifications, and thinned cortex, with a sclerotic halo around it. In our case, the absence of calcification confused the diagnoses. Takigawa *et al* classify enchondromas according to their location and extension, from central to eccentric, with cases of extensive involvement and others of a polycentric nature. Our case is considered giant as it occupies the entire distal phalanx (5).

The recurrence rates in the literature search are variable. In a review conducted by Bachoura *et al*, the incidence of the recurrence rate was 2.0% after curettage, 3.2% when autograft was used, 0% in the group that employed other osteoconductive materials and 0% in the group treated with cement. Furthermore, infection is the most frequently reported complication according to the same study. Montero *et al* recommend a longer follow-up period because there is the possibility of recurrence after five years (26).

CONCLUSION

The lytic lesions of the thumb are rare entities and require a thorough evaluation for their differential diagnosis. The thumb is a finger of high functional importance in comparison to the other digits, thereby radiological evaluation and further assessment of lytic lesions are mandatory. The time of surgery is still controversial, since waiting or not for the fracture to heal may play a crucial role intraoperatively. Thorough curettage and bone grafting was an effective option for our patient. ■

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