Original Article

Three different methods for treating multiple enchondromatosis in one hand

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Abstract: Ollier’s disease remains comparatively rare, and is a non-hereditary cartilage dysplasia of bone. It is usually associated with problems such as deformity and fracture. Three different methods were used in a one-hand of 15-year-old boy reporting his pain in the left hand and swellings. After the curettage of tumor, regarding as the differences of all parts of the bone structure reconstruction in the patient’s hand, we chose three following methods for this boy, i.e. fixed by the locking plate with calcium phosphate cement, filled with allograft bone, curetted the tumor without any bone graft. After the surgery, the patient was able to perform full motion of the operated hand. No evidence of recurrence was noted four years after surgery. To choose the different ways with bone grafts or not that relies on the patients’ conditions for bone structure reconstruction. However, patients with large osseous defects or pathological fracture, we demand full bone graft and reliable internal fixation. After surgery, early exercises can reach a desirable result and functional recovery.

Keywords: Ollier’s disease, enchondromatosis, bone graft

Background

Enchondromatosis is a form of osteochondrodysplasia characterized by a proliferation of enchondromas. Multiple enchondromatosis (Ollier’s disease), originally reported by Ollier in 1899, is a rare and non-hereditary cartilage dysplasia of bone, consisting of multiple, asymmetrically distributed, intraosseous cartilaginous foci and subperiosteal deposition of cartilage, either exclusively or predominantly involving one side of the body. It arises in the medullary cavity and grows in the cortex, forming a prominent endogenous mass of the bone. Some patients will not accept the medical treatment until a pathological fracture occurs. And also the enchondroma will not be discovered until some patients take radiographs after trauma. We report a 15-year-old boy’s case with three different treatments in one hand. There have not been any articles reporting different treatments that are used on the same patient.

Case presentation

A 15-year-old teenage boy came to the Hand Clinic in the First affiliated hospital, Zhejiang University, complaining of the pain that was lasting for half a year in his left hand. We found swellings and deformities on his dorsal of hand and in his proximal phalanx of middle and ring fingers respectively. There was no history of injury, systemic symptoms or operation. The skin and tendon were unaffected. Laboratory studies were within normal findings. Radiographs showed expansion of bone and thinning of cortex, endosteal scalloping, cortical thinning in the 3rd~4th metacarpal shaft and the proximal and middle phalanx of the middle and ring fingers respectively. The lesions caused deformities of the 3rd~4th metacarpal shaft and the 4th metacarpal shaft had a pathologic fracture. The middle phalanx of the index finger had only thinning of one side cortex. The radiographs findings showed no signs of aggressive activity (Figure 1). Based on the clinical presentation and radiographic findings, we diagnosed this case as a benign tumor. An incisional biopsy procedure could not be performed in order to avoid the wound contamination.

A dorsal skin incision was made under general anesthesia so as that the overlying extensor tendon was exposed longitudinally and mobilized medially or laterally. A cortical window was
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Three days after the surgery, the patient was able to perform full motion of the operated hand. Three months after the surgery, the patient could do all weight exercises (Figure 2). One year after the surgery, the locking plate fixation was removed and the patient’s functions presented normally. No evidence of recurrence was noted four years after surgery. Bone regeneration was noted both radiographically and clinically (Figure 3).

Discussion

Ollier’s disease, a rare non-hereditary skeletal disorder, is characterized by multiple enchondromas with a predilection for unilateral cosmetic problems such as deformity and fracture. The enchondromas grow large with progressive skeletal growth, become more evident and disfigure. After the cessation of growth and development, the enchondromas do not enlarge [1]. The rate of occurrence of this disease is 1/100,000, which is equal between men and women [2]. In general, enchondromas are involved in the short tubular bones of the hands and feet, and also found incidentally in the distal femurs, and proximal humerus. They undergo malignant transformation in less than 1% of cases [3]. Malignant transformation and development of extra skeletal cancers are the main complications of multiple enchondromatosis. It has up to 30% risk of malignant chondrosarcomatous transformation [4]. Patients with enchondromas of the small bones of the limbs have the lowest risk to develop malignancy (14%), whereas enchondromas affecting the long axial bones show the highest risk (44%--55%) [5]. Multiple enchondromas and hemangiomas of soft tissue constitute a condition known as Maffucci’s Syndrome. It is less common than Ollier disease, but has a higher risk of chondrosarcomatous change. Other cartilaginous lesions include bone infarct, low-grade chondrosarcoma, epidermoid inclusion cyst, chondromyxoid fibroma, unicameral bone cyst, giant cell tumor and fibrous dysplasia. Our case didn’t destruct the cortex or enter the soft tissues, so benign tumors should be firstly considered.

Surgery remains the only effective solution to remove the enchondroma and prevent malignant transformation.

Surgical resection to multiple enchondromas of the hand includes curettage, curettage with autografts or allografts and amputations [6-8]. According to Klausmeyer MA et al. and Hasselgren G et al., despite large bony deficits after resection, bony regrowth occurs under the circumstance of without bone autograft [9, 10].
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Figure 2. After three months postoperative radiograph of the left hand, showing Pathologic fracture has healed, internal fixation has presented stable and bone grafting is without bone absorption.

Figure 3. After four years postoperative radiograph of the left hand, showing No evidence of recurrence was noted. Bone regeneration repairs bone defect.

Our case obviously showed bony defects and pathological fracture. Concerned about stable fixation and the initiation of early range-of-motion exercises, we chose the calcium phosphate cement to fill with the defect of metacarpal shafts and then the metacarpal shafts were fixed with the locking plate. Proximal phalanx of the middle finger, proximal and middle phalanx of ring fingers were filled with allograft bone. The middle phalanx of the index finger was only curettage of the intramedullary. Three different treatments in one hand had a satisfactory result.

Conclusions

This case report emphasizes that according to the patient’s conditions of bone structure, we choose different treatments including bone grafts and without bone grafts. If patients have large osseous defects or pathological fracture, we demand bone graft and reliable internal fixation. After surgery, early exercises can reach a desirable result and functional recovery.

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Disclosure of conflict of interest

None.

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