Multiple Enchondromas of the Hand in Children: Long-Term Follow-Up of Mean 15.4 Years

Assaf Kadar, Geffen Kleinstern, Mohamed Morsy, Endre Soreide, Steven L Moran

Abstract:

Background: Multiple enchondromatosis of bone, termed Ollier's disease, or Maffucci syndrome when associated with hemangiomas, is a rare disease that can affect the pediatric hand. This condition often causes a finger mass, deformity, pain and possible pathologic fractures, and has been associated with malignant transformation to chondrosarcoma. The aim of our study is to describe the long-term sequela of multiple enchondromatosis of the hand in the pediatric population, specifically the rates of malignant transformation, tumor recurrence, rates of pathologic fracture, and phalangeal growth arrest. Methods: We examined 15 pediatric patients who were treated in our institute with a total of 127 phalanges and metacarpals lesions. Only patients with follow-up of at least 4 years were included. We retrospectively reviewed patients' chart and hand radiograph for symptoms including pathologic fractures, indications for surgery, and postoperative complications including tumor recurrence, and malignant transformation. We assessed phalangeal growth arrest with radiographs and normalized phalangeal growth charts. Results: Mean age of diagnosis was 5.8 years and mean follow-up time was 15.4 years. Pathologic fractures were common at 46% of pediatric patients, but ceased to occur once reaching adulthood. Outcomes of pathologic fractures were excellent, regardless of treatment. Malignant transformation occurred in 1 patient and did not occur during childhood. A total of 80% of patients and 29% of lesions underwent surgical treatment of curettage and bone graft for the lesion, yet recurrence was common and affected 33% of treated patients. Phalangeal growth arrest was the most common long-term sequela and affected 11% of phalanxes and metacarpals. This sequela was significantly more prevalent in patients who had surgical excision of the tumor. Conclusions: Our findings reassure that malignant transformation of enchondromatosis of the hand is unlikely in the pediatric population. Pathologic fracture is common, but has excellent outcomes. When considering surgery, parents should be counseled about the possibility of phalangeal growth arrest and recurrence of the lesion.

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