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REVIEW

Multiple osteochondroma of the hand: initial and long-term follow-up study

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Abstract
Background The purpose is to determine the location and type of osteochondromas in patients with multiple osteochondroma of the hand as well as the presence of shortening and angulation. Second, it aims to establish longitudinal data on the change in tumors.
Methods Retrospective review of patients with multiple osteochondroma affecting the hand evaluating the location and type of tumors as well as the presence of shortening and angulation is done. We examined radiographs from final follow-up and analyzed them based on patient age at presentation (group I=ages 2–6; II=ages 7–10; III=ages 11–19), to determine changes over time and any differences in the number of tumors, location, and shortening and angulation.
Results The most affected bones were the index and small finger metacarpals with an increase seen around the metacarpophalangeal (MCP) joints. The most shortening and angulation were seen on the ulnar side. Group II had the most tumors and the most bones with angulation. Twenty-three hands had longitudinal follow-up with an overall increase of 2.7 tumors per hand with a range of loss of 8 to gain of 16. There was an increase in the number of bones with angulation and shortening. Group I showed the largest increase in tumors, shortening, and angulation.
Conclusions The ulnar side and bones around the MCP joints are affected most commonly. The largest change was seen as the patients went from young childhood into adolescence, which may be due to rapid growth during this time. This is the largest study of these patients with the longest longitudinal data.

Keywords Osteochondroma · Hand · Multiple hereditary exostoses (MHE)

Introduction

Osteochondromas are common benign bone tumors found predominantly in children and adolescents [5]. They are characterized by cartilage-capped exostoses usually found in the metaphysis of the most rapidly growing ends of long bones. While usually solitary, a condition called multiple hereditary exostoses (MHE) is an autosomal dominant disorder where many osteochondromas can be found in multiple locations. Also known as multiple osteochondroma, this disorder usually shows full penetrance. It is caused by a defect in the EXT1 or EXT2 genes found on chromosome 8 or 11, respectively. These genes play a role in regulating chondrocyte maturation and differentiation and therefore can affect normal endochondral ossification [5].

Reports vary on the percentage of patients with multiple osteochondroma showing hand involvement ranging from 30 to 79 % [1, 3–5]. Most agree that lesions in the hand are generally asymptomatic and do not require any intervention unless severe angulation or rotation develops or they are subungal and disrupting nail growth. However, these can be a difficult problem in patients with multiple lesions and can require operative intervention to address deformity or pain [5].

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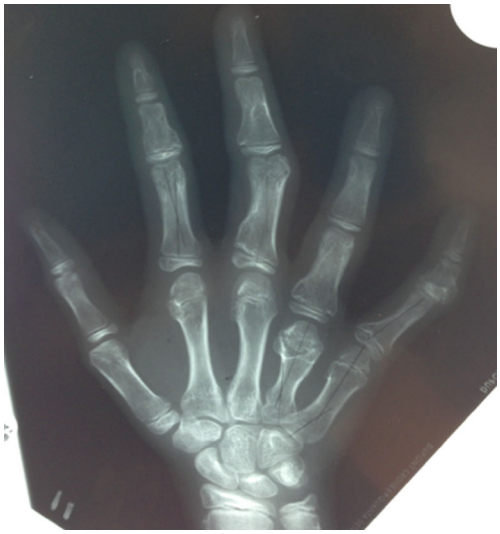

Fig. 1 A patient with multiple osteochondroma showing brachymetacarpia most pronounced in the ring metacarpal and angulation of the ring and small finger metacarpals

Overall, there is little known about the natural history of patients with MHE with regard to the hand. How these lesions change both in terms of their number and size over time is uncertain because there is little long-term follow-up of patients with MHE affecting the hand.

We performed a retrospective X-ray review of patients with MHE of the hand with the purpose of achieving two primary outcome measures. First, we examined all patient radiographs to evaluate the distribution of osteochondromas and their effects on the bones. Second, we reviewed subsequent patient radiographs to determine changes in osteochondroma number and location over time based on different age groups of patients.

Materials and Methods

After obtaining Institutional Review Board (IRB) approval, we completed a retrospective radiographic review of patients with multiple osteochondromas of the hand. In part 1 of this study, the presenting X-rays for each patient were reviewed and analyzed. Then, all patients with longitudinal data had their final X-rays reviewed and the data compared with the initial films. We looked at multiple variables and evaluated X-rays for types of lesions according to a previous study done by Cates et al. [2]. They described three types of lesions—type A lesions are large, globular cavitating lesions with >50 % of bone affected. Type B lesions are smaller, sessile lesions with <50 % of bone affected. Type C lesions are classic pedunculated outgrowths. Other variables assessed were shortening and angulation of the bone, age at presentation, and left or right hand. Shortening was evaluated by visual comparison of bones comparable to the others in the hand (Fig. 1). Angulation was measured by drawing angles and using a goniometer on the plain films and considered angulated if the measurement was >5° from a straight axis (Fig. 2). Overall, there were 83 hands in 46 patients for evaluation at time 1. These patients had an age range of 3–34 years and an average of 11 years old.

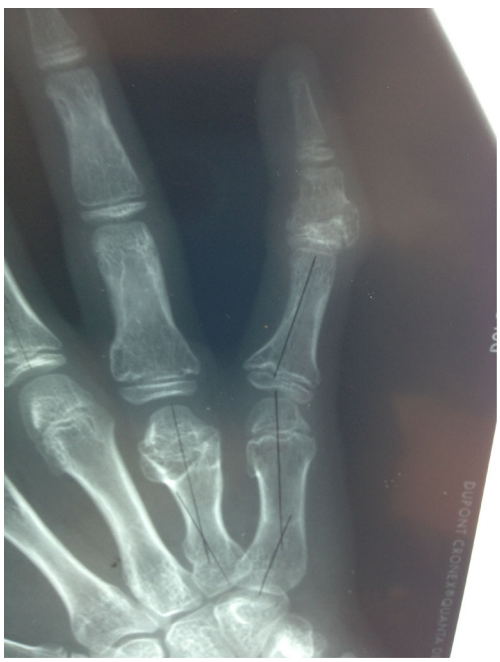

Fig. 2 An example of angulation measurement

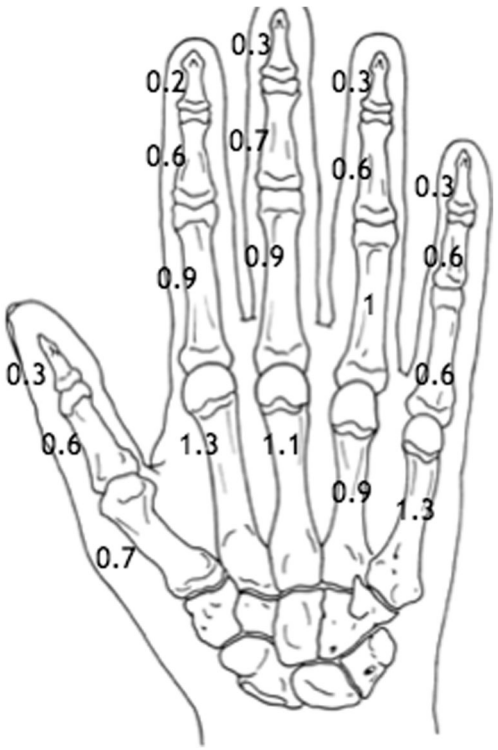

Fig. 3 The average number of tumors per bone

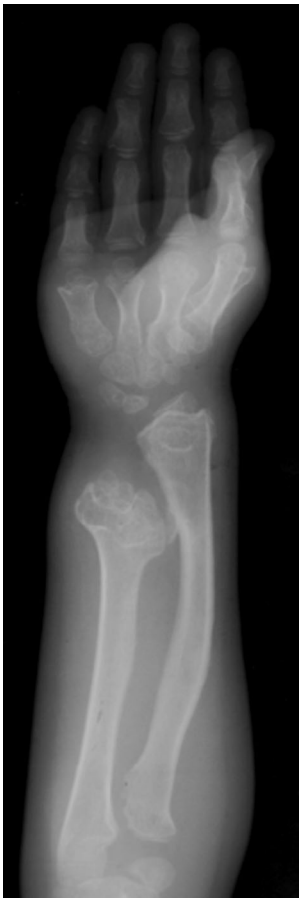

Fig. 4 Radiograph showing brachymetacarpia with angulation of the small finger metacarpal

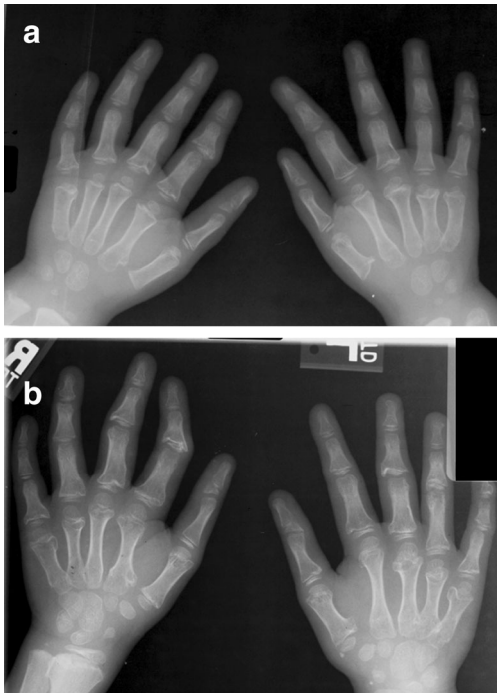

Fig. 5 **a**, **b** A patient from age 3 to age 8 with increased number of tumors and angulation

We then analyzed the same variables but also looked at changes over time in the number of osteochondromas, shortening, and angulation. We separated these data by ages, forming three groups, I=ages 2–6, II=7–10, and III=11–19. These ages were chosen with the intent of having a group of very young children, those in middle childhood, and prepubescent into puberty. There were 23 hands (13 patients) for follow-up data analysis with an average age of 14.1 years and follow-up time of 4.6 years.

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. The IRB approved a waiver of informed consent for this study because it was determined that the study posed minimal risk to subjects and met the criteria for expedited review under the Code of Federal Regulations.

Results

At presentation, we had a total of 83 hands for evaluation in 46 patients. Ages ranged from 3 to 34 years and averaged 11 years at the time of inclusion into this study. The average number of osteochondromas per hand was 13.1 with a range of 2–30. The most affected finger was the small finger (3.3 tumors/finger) > index (2.96) > middle (2.95) > ring (2.7) > thumb (1.63). Type B was the most common type at approximately 98 %, and they tended to be located on the proximal end of the bone (58 %) versus the distal end. An average of five bones per hand had angulation and two bones per hand had shortening.

Table 1 Number of osteochondromas based on age at presentation

Group	Avg age	No. of hands (patients)	Tumors/Hand	Finger with most	Finger with fewest	Most common type	No. bones in hand with shortening	No. bones in hand with angulation
I	4.3	21 (13)	8.8	Small	Ring	B	1.2	4
II	9	22 (13)	15.7	Small	Thumb	B	2	7.3
III	15.8	40 (23)	13.9	Middle	Thumb	B	2.5	4

Group I 2–6 II 7–10; III 11–19

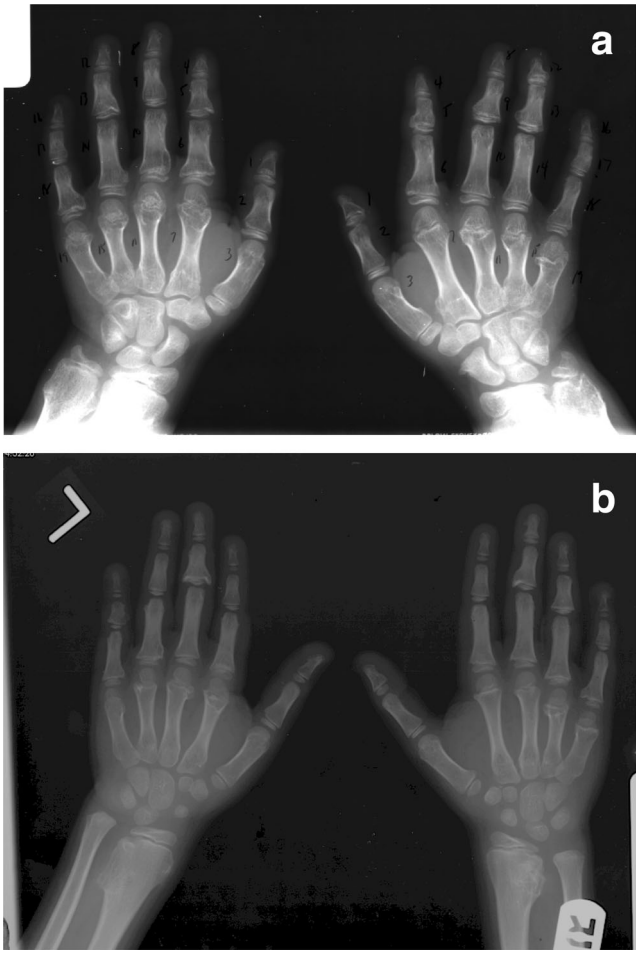

Fig. 6 **a**, **b** A patient with decreased number of tumors as well as resolution of deformity especially seen in the left ulna and small and ring fingers

The most common bones affected were the metacarpals of the index and small fingers that both averaged 1.3 lesions per hand. The least affected was the distal phalanx of the index with 0.2 lesions per hand (Fig. 3). The metacarpals on the ulnar side of the hand tended to have the most shortening, and the metacarpal of the small finger also tended to have the most angulation (Fig. 4).

We then re-analyzed the data based on the age at presentation. Table 1 shows these results.

There were 23 hands in 13 patients evaluated at a second presentation. The average time of radiographic follow-up was 4.6 years (range 1–13 years) with an average age of 14.1 years. The overall change in tumors was +2.7 tumors per hand with the extremes being loss of 8 to a gain of 16 (Figs. 5a, b and 6a, b). These losses were all spontaneous regression and not surgical excision. Most gains were seen in the ring finger +1.7 tumors per hand. There was an average increase in shortening and angulation of 1.3 and 0.8 bones per hand. The ring finger metacarpal and small finger distal phalanx had the greatest increase in numbers of tumors (average 0.5). Changes in shortening and angulation averaged about the same for all bones.

We then looked at changes based on age at presentation in Table 2.

Discussion

Most osteochondromas present at the growing ends of long bones as either sessile (broad based lesion with diameter greatest at base) or pedunculated (diameter increases following a stalk) lesions. Growth usually stops at skeletal maturity, and tumor growth rates are proportional to the overall growth rate of the patient. There have been reports of spontaneous regression of lesions [5], and we observed many such cases in our series. Most patient complaints are of cosmesis or pain. Pain is a result of repetitive trauma to the prominent area, muscles snapping over lesions, or fractures of the tumors. Joint and soft tissue problems can also occur—such as restricted range of motion; bone malalignment; or impingement to tendons, nerves, and vessels.

Some studies show higher prevalence in males, but more recent familial genetic studies have shown no gender difference [5]. Malignant transformation rate is estimated to be between 0.5 and 25 % [1]. Signs of malignant transformation are increasing pain, growth of lesion after skeletal maturity, and adults with cartilage caps >2 cm.

Cates et al. [2] performed a study examining 42 hands in 22 patients. In this study, they found increased involvement of proximal phalanges and metacarpals with the small finger

Table 2 Follow-up number of osteochondromas

Avg age at presentation (years)	Avg age at follow-up (years)	Avg follow- up time (years)	Change in tumors	Change in bones with shortening	Change in bones with angulation
Group I—6 patients, 10 hands					
4.5	12.2	7.7	+2.8 tumors/hand	+1.2 bones/hand	+0.7 bones/hand
Group II—2 patients, 4 hands					
10	13	3	−0.2 tumors/hand	+0.2 bones/hand	0 bones/hand
Group III—5 patients, 9 hands					
14.9	16.8	1.9	+0.1	−0.1	0

metacarpal having highest incidence of 86 %. The distal phalanges and thumb were less affected with the small finger distal phalanx the least affected at 17 %. Wood et al. [6] in 1990 found that the area around the MCP joints of the long, ring, and small fingers were the most commonly affected. In the present study, the metacarpals of the index and small finger were most commonly affected with the thumb being the least affected. Overall, lesions were more prevalent around the metacarpal phalangeal joints. The ulnar side of the hand in this group showed the most shortening and angulation. Cates and Wood also identified type B sessile lesions most commonly in the hand, which is similar to the data seen here [2, 6].

To date, this is the largest known study of MHE involving the hand with 83 hands at presentation and longitudinal data on 23, while the other largest series to date had 42 hands and serial radiographs on 7 patients [2]. When further examining data based on age at presentation, the middle group (ages 7–10) had the most tumors per hand, which may be related to rapid skeletal growth during those ages. They also presented with the most angulation, although the number of bones showing shortening were similar across age groups.

Longitudinal follow-up data show that there is a wide array of changes seen in patients with multiple osteochondroma of the hand. Cates et al. [2] showed a transient exostoses rate of 4.9 % in their seven patients. We had a range of a loss of 8 tumors per hand (by spontaneous regression) to a gain of 16 over the follow-up period with an average of gain 2.7 tumors. There was also an average increase in one bone per hand shortening and angulation. The largest increase was again seen toward the ulnar side of the hand.

Breaking these data down into age groups shows the largest change seen in group I (ages 2–6 at presentation). We anticipate the largest growth spurts during the follow-up time for that group and therefore the largest average increase in tumors of almost three per hand, shortening (one bone per hand), and angulation (one bone per hand). It appears that as patients grew older than 12–13 years of age, their tumors became more stable and little change was seen. There were also a few cases in older children where angulation or shortening was found in bones without tumors or where tumors had resolved. This was also seen by Wood et al. [6], where five patients had shortening of the small and/or ring finger metacarpals without any visible lesions.

The limitation of this study is that the data are purely radiographic without clinical correlation. It was difficult to objectively measure shortening on plain films given the lack of control digits. Also, dividing the follow-up data into groups based on ages did decrease the average follow-up time and number of patients in two of the groups. We did think that the trend of decreased changes in tumors after the age of 12 and increase in tumors and change in the youngest group did add information for this study. It helped to show trends that could be related to rate of skeletal growth. Despite these limitations, the data presented gives the physician information on presentation as well as the natural history through childhood in order to counsel patients and families on the condition.

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Conflict of Interest Julie Colantoni Woodside has no conflicts of interest to disclosure. R.Glenn Gaston has no conflicts of interest directly associated with this manuscript. However, outside of the described study, R.Glenn Gaston is a board member of the American Society for Surgery of the Hand; is on the editorial board of the Journal of Hand Surgery—American; is a paid consultant for Biomet; receives royalties from Biomet; and has been on a speakers bureau or done a paid presentation for Auxilium, Biomet, Smith and Nephew, and BME. Timothy Ganey has no conflicts of interest to disclose.

Statement of Human and Animal Rights All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008 (5).

Statement of Informed Consent The IRB approved a waiver of informed consent for this study because it was determined that the study posed minimal risk to subjects and met the criteria for expedited review under the Code of Federal Regulations.

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