

# Clinical Quiz

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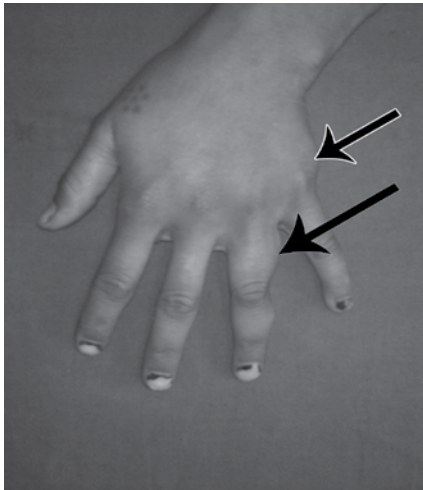
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## Enchondromas in the small bones of the hand

### Clinical Presentation

A 12-year-old female patient was brought to the orthopedic outpatient department with complaints of swelling in her left hand and fingers for the last 7 months. She also complained of minimal pain occasionally. There was no history of trauma, fever, and other constitutional symptoms. On examination, the swellings involved the metacarpal and phalanx of the little and ring finger of her left hand. Swellings were non-tender, bulbous, and were not associated with signs of inflammation (Figure 1). The rest of the skeletal survey was normal. Anteroposterior radiograph of the left hand was performed (Figure 2).



**Figure 1** - Clinical photograph of patient's left hand showing multiple bulbous swellings involving the metacarpal and phalanx of the little and ring finger (arrow heads).



**Figure 2** - The anteroposterior radiograph of the left hand shows multiple small intramedullary lytic lesions (arrow heads).

## Questions

1. What are the features seen on the radiograph?
2. What is the diagnosis?
3. What is the management?

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## Answers

1. The anteroposterior radiograph of the left hand shows multiple small intramedullary lytic lesions in the metacarpal, and phalanx of the little and ring finger with a distinct zone of transition, expansion of bone, thinning of overlying cortex, that has a somewhat scalloped appearance, and without any evidence of periosteal reaction and pathological fracture (Figure 2). The lesion is mainly involving the diaphysis and seems to be approaching the end of the bone near the metacarpophalangeal and inter-phalangeal joints.
2. The clinico-radiological diagnosis is enchondromatosis of the small bones of the hand. When multiple enchondromas coexist, the diagnosis of enchondromatosis should be considered. Multiple enchondromas may occur in Ollier's disease, and Maffucci's syndrome.<sup>1</sup>
3. Curettage remains the treatment of choice for enchondromas. Additional bone grafting or the use of bone substitutes is contentious.<sup>2</sup> Autologous bone grafting with a biological material that is completely incorporated into the cancellous bone is considered to be the gold standard,<sup>3</sup> though other authors have advocated simple curettage as sufficient.<sup>4</sup> Schaller and Baer<sup>2</sup> concluded that there is no major difference between the curettage with or without bone grafting in the treatment of enchondromas of the hand, and consequently additional bone grafting is uncalled for. The exceptions remain special indications such as preventing pathological fractures in extremely weakened out cortical bone and lesions in close proximity to a joint.

## Discussion

Enchondroma is a type of benign bone tumor that originates from cartilage. An enchondroma most often affects the cartilage that lines the inside of the bone, and substitutes normal bone with mineralized, or unmineralized hyaline cartilage.<sup>1</sup> The bones most often involved are the small bones of the hands and feet. In fact, enchondromas are the most frequent tumors of the hand. In rare instances, multiple tumors can be seen in syndromes like Ollier's disease and Maffucci's syndrome. Malignant transformation to secondary chondrosarcoma is extremely rare.<sup>5</sup> The lesions likely crop up from cartilaginous rests that are displaced from the growth plate. A characteristic radiological pattern of calcifications, described as rings, and arcs, is pathognomonic when it is seen in the small bones of the hand.<sup>5</sup>

Enchondromas are generally painless because of their sluggish growth, negligible peritumoral reaction, and avascularity. The tumor is detected accidentally, unless there is a pathological fracture. Malignant transformation of a solitary enchondroma into secondary chondrosarcoma in the hand is extremely rare.<sup>5,6</sup> The diagnosis of an enchondroma in most cases is made by clinico-radiological examination. If there is uncertainty left, a biopsy may be performed. Curettage remains the treatment of choice for enchondromas. Surgical treatment often can be avoided if the diagnostic imaging suggests a benign typical lesion with normal follow-up.<sup>5</sup> Moreover, a histological confirmation of the diagnosis through a biopsy usually is obligatory before definitive treatment, because resection of an enchondroma is less aggressive than of a chondrosarcoma.<sup>5</sup>

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