Hereditary Multiple Exostoses

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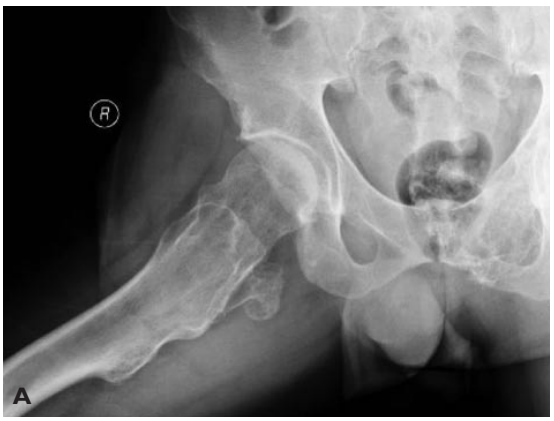

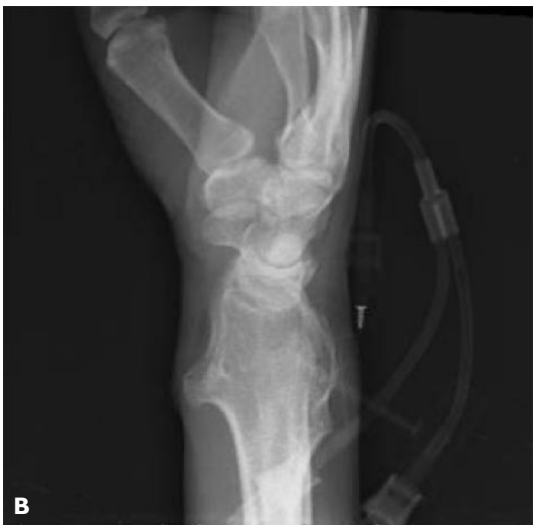

A 29-year-old Caucasian man was noted to have a bony prominence of his ventral wrist during a routine physical examination on morning rounds after he had been admitted the previous evening for acute intoxication. Upon questioning, the patient revealed that he had been diagnosed with hereditary multiple exostoses (HME) during adolescence. He had an extensive surgical history, with multiple orthopedic procedures performed to restore range of motion to his right lower extremity. Hip and wrist radiographs revealed bony excrescences with extraosseous calcification (**Image A** and **Image B**).

HME (also referred to as hereditary multiple osteochondromata and multiple cartilaginous exostoses) is a rare autosomal dominant genetic disorder of bone growth characterized by cartilage-capped benign bone tumors that grow outward from the metaphyses of long bones. The reported prevalence of HME is approximately 1 case per 50,000 persons.[1] The prevalence is higher among certain ethnic groups, with Native Americans of Manitoba, Canada, having an incidence of 1 case per 77 persons.[2] Approximately 10% of individuals with HME acquire the condition as a result of a de novo gene mutation.[3]

In HME, exostoses arise during childhood, grow while the metaphyses are open, and stop growing once skeletal maturity is reached. These growths are typically benign, but they may disturb normal bone growth when they occur near the growth plate or cause reduced range of motion when they occur near a joint. Exostoses may also impinge on surrounding soft tissue, such as nerve sheaths, and may require surgical removal for symptomatic relief. They may form a bony protuberance visible through the skin, which may be cosmetically intolerable for some patients. Malignant transformation is seen in approximately 1% of patients with HME.[4] Recognizing the radiographic evidence of multiple exostoses, especially in a child, can help the physician set long-term preventive plans. Patients with HME should be monitored with radiographs periodically to screen for malignant transformation. Proactive surveillance may allow for resection of low-grade lesions before progression to invasive chondrosarcoma. **HP**

REFERENCES

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