Images in Rheumatology

Ollier's Disease with Sella Turcica Involvement

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Multiple enchondromatosis (Ollier's disease) is a rare, nonhereditary condition, a dyschondroplasia caused by a developmental error in enchondral ossification that is characterized by multiple, persistent, cartilaginous masses in the metaphyses and diaphyses. The dyschondroplasia leads to shortening and deformity of the extremities,

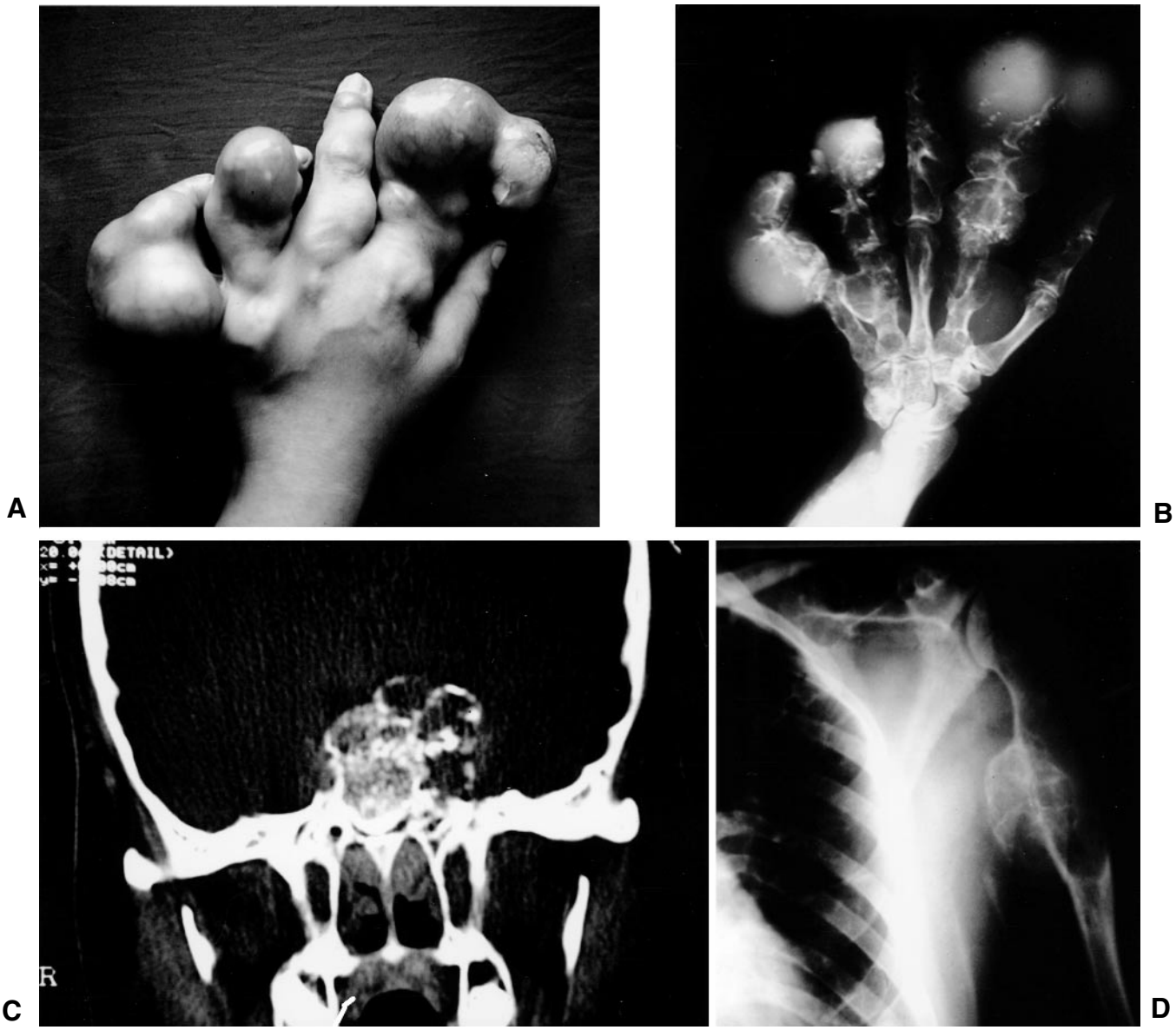

Figure 1. A. The left hand shows deformities of nearly all fingers caused by enchondromas. B. Posteroanterior radiograph of the left hand showing bubbly, expansive lesions in all phalanges and metacarpal bones. C. Preoperative CT scan showing a large mass in the sellar region. D. Radiograph showing an enchondroma in the humerus.

resulting in limb length discrepancies and bone fractures[1].

A 27-year-old man noted multiple hard nodules on his left fingers when he was 10 years old, initially affecting the second metacarpophalangeal and proximal and distal interphalangeal joints, extending later to the fourth and fifth fingers (Figure 1A). At age 13, he developed subcutaneous masses in his left arm and ankle. There was no family history of any vascular or skeletal disorder.

At admission, he complained of pain in the hand with function loss, as well as frontal headache; the symptoms had begun 6 months before and had been progressing for the previous 5 weeks. The classic radiographic pattern was seen in radiographs of the left hand. Expansive masses in all phalanges and metacarpal bones representing enchondromas were present (Figure 1B). Additional lesions were also observed in the ribs, arm, and femur; the enchondromatosis affected mainly the left extremities, and the diagnosis of Ollier's disease was established.

The hand was considered inoperable and amputation was recommended. Three months later, because of worsening of the headache and progressive diplopia, a computed tomographic (CT) scan was performed, which revealed a mass lesion at the cranial base (Figure 1C). A frontal craniotomy was performed, revealing a sella turcica enchondroma; the lesion was removed, and no additional deficits were observed.

He did well during the next 4 years, when he sustained a traumatic fracture to the left humerus (Figure 1D), which required amputation. Afterward he was lost to followup. The involvement of sella turcica by an enchondroma has only rarely been described[2-4].

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