Clinical History:

A 43-year-old woman presented with a painless swelling over the ulnar aspect of the right hand. There were no associated signs or symptoms, nor was there a history of trauma. Radiographs (Figure 1) were obtained.

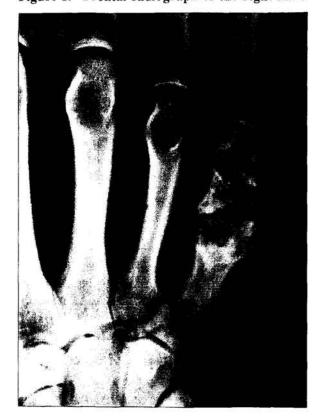


Figure 1: Frontal radiograph of the right hand

Answer on page 405

What is the diagnosis?

- a) Aneurysmal bone cyst
- b) Enchondroma
- c) Giant cell tumour
- d) Fibrous dysplasia
- c) Epidermoid inclusion cyst

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Answer:

b) Enchondroma

Radiological findings

The radiograph (Figure 2) shows a rounded intramedullary lesion causing expansion of the shaft of the 5th metacarpal. The lesion is well-defined, and has a narrow zone of transition with thin sclerotic margins. It is mainly radiolucent with areas of calcification and

Figure 2: Same radiograph as Figure 1 with the addition of arrows. An expanded lytic lesion, with well-defined thin sclerotic margins (arrowheads), is seen. Punctate (some indicated with long arrows) and flocculent (curved arrows) calcifications, as well as "rings and arcs" ossifications (small arrows), are present within the lesion matrix



ossification within. No cortical breach, associated periosteal reaction nor soft tissue swelling is detected.

Discussion

Enchondroma

The radiographic features and site of involvement are typical of those of enchondroma. Enchondroma is a benign cartilaginous tumour located in the medullary cavity. It arises from a failure of normal enchondral ossification. Bones preformed in cartilage are typically affected. Patients with these lesions are usually asymptomatic and lesions are usually discovered incidentally at the ages of 10-50 years, with a peak in the third decade. Occasionally, patients may complain of a painless swelling of the affected bone leading to its discovery. There is no sexual predilection.²

Enchondroma affects the small bones of the wrists and hands in up to 50% of the cases.³ In the hand, it commonly affects the metacarpals, and the proximal and middle phalanges, occurring more often in the ulnar three digits, particularly the fifth digit.¹ The other frequent sites of involvement include the distal femur and proximal tibia.⁴ Within these bones, enchondromas are distributed mainly in the metaphyseal or diametaphyseal locations. Enchondromas are the commonest tumour occurring in the phalanges and after osteochondromas, are the second most common benign neoplasm of cartilaginous origin.⁴

Radiographically, enchondroma is seen as an oval to round, well-defined radiolucent lesion with sharp, thin sclerotic borders. It is typically centrally located in a tubular bone, causing bulbous expansion with cortical thinning and endosteal scalloping. As it is of cartilaginous origin, matrix calcification is often seen. Patterns of calcification include the punctate, stippled or flocculent types. The "rings and arcs" forms of ossification may be present. Finding of these types of mineralisation greatly assists the diagnosis.

Malignant degeneration of solitary enchondromas of the hands and feet is extremely rare, thus lesions

involving these sites are best managed conservatively. However, if there is an enlarging lesion after cessation of bone growth, progression of bone destruction, margin irregularity or the onset of painful symptoms, then an underlying malignant process should be suspected. In the long bones, incidence of malignant change is higher, reported to be in the range of 15-20%.² Associated pathological fractures should raise the suspicion of malignant transformation of long bone lesions. On the other hand, small stress fractures are commonly encountered in phalangeal enchondromas.⁴

Multiple enchondromatosis (Ollier's disease or dyschondromatosis) is a non-hereditary condition in which multiple bones contain proliferating benign cartilaginous masses. This entity presents in childhood and has a variable appearance. It may appear as radiolucent streaks arranged in a linear manner perpendicular to the growth plate, representing cartilaginous rests in the bony medulla, or as large osteolytic lesions, with or without a calcified matrix. These lesions may lead to bony deformity or leg length discrepancy. Multiple soft tissue haemangiomas may be associated with enchondromatosis (known as Maffucci's syndrome). These soft tissue masses characteristically contain phleboliths. Malignant transformation is reported to be higher in enchon-dromatosis (up to 30% of affected individuals) than in solitary enchondroma.1.4

Aneurysmal bone cyst

Patients with aneurysmal bone cyst (ABC) commonly present at 10-30 years of age, with a slight female predilection. ABC is seen as a radiolucent lesion affecting the metaphysis of long bones. It does not cross the growth plate until after fusion. When it involves the spine, it tends to be located in the posterior elements. The lesion is usually purely radiolucent, typically causing marked eccentric expansion of the bone with cortical thinning but without periosteal reaction. The common presenting symptoms are pain and swelling. In the spine, cord or nerve root compression may occur. Intra-lesional fluid levels may be demonstrated on computed tomography or magnetic resonance imaging. This lesion is rare in the hands and does not contain internal calcification, hence this diagnosis can be safely excluded in our patient.

Giant cell tumour

Giant cell tumour (GCT) typically presents in young adults between 20 and 40 years of age. About 85% occur in long bones (50-60% around the knee) and 15% in flat bones. It is most often seen after epiphyseal plate fusion and is characteristically subarticular in location. GCT appears as a well-demarcated, expansile, radiolucent lesion, eccentrically located in bone, with no sclerosis, periosteal reaction or internal calcification. It may be complicated by a pathological fracture. The diametaphyseal site and presence of calcification in the lesion of our patient makes this diagnosis very unlikely.

Fibrous dysplasia

Fibrous dysplasia is a benign fibrous developmental anomaly of unknown origin, usually diagnosed between the first and second decades of life. It is monostotic in 85% of cases, mainly affecting the proximal femur, tibia, ribs, and craniofacial bones. The polyostotic form (15%) occurs in the long bones, pelvis, ribs, feet, spine and clavicle. In long bones, it affects mainly the metaphysis with extension into diaphysis. It is radiolucent, typically described as having a "ground glass" appearance, with a well-defined thick sclerotic border of reactive bone. It may contain internal calcifications. Patients may present with pain due to incomplete fractures, deformity or complete fractures. Associated endocrine abnormalities include the McCune-Albright syndrome. Although fibrous dysplasia may mimic many other bone tumours, it rarely affects the hands and mineralisation, if present, tends to be distributed throughout the lesion rather than in a discrete or "rings and arcs" pattern.4

Epidermoid inclusion cyst

Epidermoid cyst, also known as implantation cyst or intraosseous keratin cyst, occurs in the second to fourth decade and affects males more than females. It is seen as a solitary radiolucent lesion, usually occurring in the terminal tuft of the fingers and less commonly, the toes.

It has well-defined margins with cortical expansion being a frequent finding. There is usually no calcification, soft tissue swelling or periosteal reaction. This lesion is thought to develop from a traumatic episode in which epidermoid cells of the skin are implanted intraosseously. In our patient, the site of involvement in the metacarpal and presence of calcification/ossification makes this diagnosis highly unlikely.

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