Clinical History:

A young man in his early twenties presented after a mild injury to his left wrist. Plain radiographs were performed (**Figures 1 and 2**).

Figure 1: Frontal radiograph of the left wrist

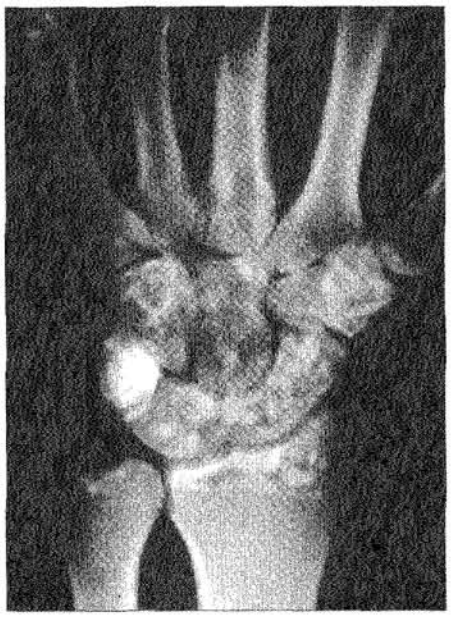
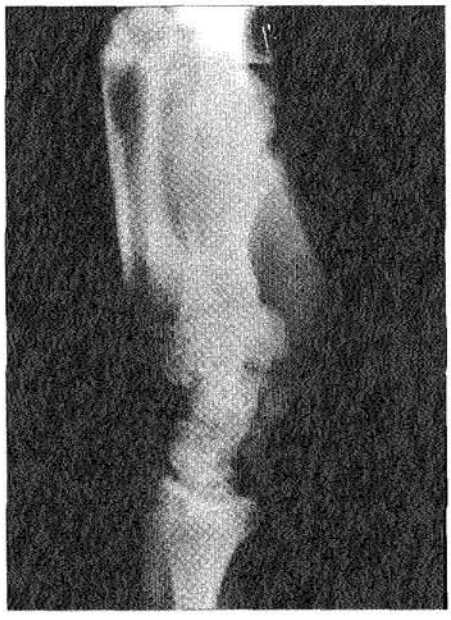


Figure 2: Lateral radiograph of the left wrist



Answer on page 347

What is the diagnosis?

a) Bony metastases

b) Osteopoikilosis

c) Chronic osteomyelitis

d) Osteopathia striata

c) Multiple exostoses

This radiology case was prepared by: **Dr. L.L.S. Wong,**
Medical Officer.
Professor W.C.G. Peh,
Department of Diagnostic Radiology,
The University of Hong Kong,
Queen Mary Hospital.

RADIOLOGICAL CONFERENCE

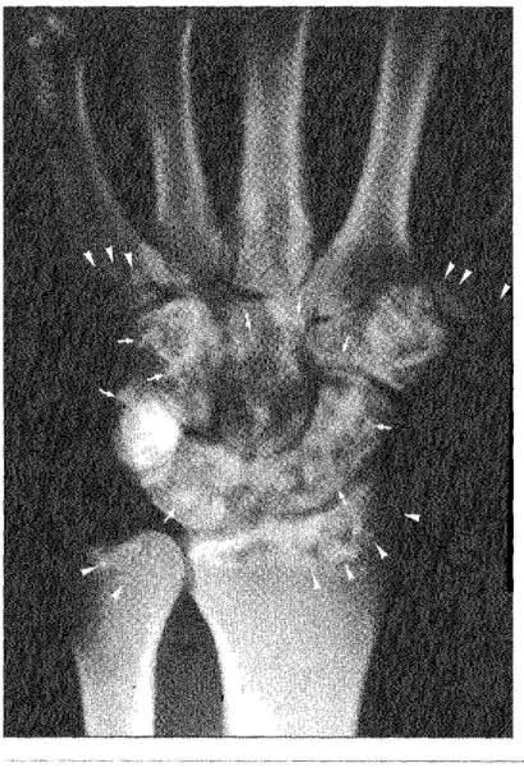
Answer:

b) Osteopoikilosis

Radiological findings

There is no evidence of a fracture or dislocation on plain radiographs of the left wrist. Multiple, small, well-defined, round and oval sclerotic lesions are present (**Figure 3**). They are distributed in the epiphyses of the distal radius and ulna, and the proximal ends of some of the metacarpals. The carpal bones are similarly involved. No cortical breaching, periosteal reaction or soft tissue swelling is noted. The joints around the wrists are normal.

Figure 3: Same radiograph as Figure 1 with the addition of arrows. Multiple small dense foci are scattered within the medullary cavity of the ends of the long tubular bones (arrowheads), as well as in the carpal bones (small arrows). The shape of the bones and the joints are otherwise normal



Discussion

Osteopoikilosis (osteopathia condensans disseminata; spotted bones) is an osteosclerotic dysplasia initially described by Albers-Schönberg, and Ledoux-Lebard and associates in early twentieth century. It is a rare hereditary condition with an autosomal dominant pattern of genetic transmission. There is a male predominance for the lesion. It develops during childhood though it is uncommon below 3 years of age. It may become evident at any age and usually persists throughout life.[1]

Osteopoikilosis is most frequently discovered by chance on radiographs although cutaneous lesions may be present in 25% of cases.[1] Being benign, it is usually not diagnosed until detected as an incidental finding.[2] Radiographical features are considered diagnostic, with a typical pattern of numerous small (2-10 mm), well-defined, oval or round foci of increased radiodensity. The oval foci are often orientated with their long axes parallel to the shafts of the tubular bones. These lesions are symmetrically distributed, and have a predilection for the epiphyseal and metaphyseal regions of long tubular bones, as well as the carpal and tarsal bones. In the pelvis and scapulae, they are concentrated at the acetabuli and glenoid, respectively. The skull, ribs, spine and clavicles are rarely involved. On serial radiographs, these lesions may increase or decrease in size and number, or disappear. Radionuclide bone scan usually reveals no increased activity within these lesions. Pathologically, these lesions are foci of compact lamellar bone within the spongiosa, with histological features identical to bone islands.[1-4]

The cause and pathogenesis of osteopoikilosis are not known. The systemic nature of the condition with widespread bone involvement suggests that an overlying neurovascular anomaly may be responsible for the subsequent new bone formation.[2] Affected bones are not prone to infection or trauma. In summary, osteopoikilosis has virtually no clinical significance other than that it may be confused with diffuse osteoblastic metastases.[5]

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Bony metastases

Metastases should be included in the differential diagnosis of a bone lesion in any patient over the age of 40 years.[5] The clinical history of a known primary carcinoma is vital and should always be sought in older patients. Symptoms such as pain, swelling, pressure effects on neighbouring structures, and pathological fractures may be present. They can have a wide variety of appearances, though they are most commonly lytic. Osteoblastic metastases are typically seen in carcinoma of the prostate. Other primary tumours which may produce sclerotic metastases include carcinoid tumour, and carcinoma of the breast, bladder, testis, stomach and colon.

Metastatic deposits are usually asymmetrical in distribution and commonly involve the axial skeleton. They often show variation in size, may be associated with osseous destruction, and typically have positive scintigraphic findings. This diagnosis can be excluded in our patient, based on the radiographical appearances and distribution of lesions.

Chronic osteomyelitis

The alterations in bone from acute or chronic infection may produce a variety of radiological changes. Chronic osteomyelitis may present radiographically as an irregular sclerotic lesion, with expansion, cortical thickening and adjacent periosteal new bone formation. When osteomyelitis occurs near a joint, the joint itself is usually involved with effusion and cartilage loss.[5] The presence of a bony sequestrum (or an island of dead bone) is highly suggestive of the diagnosis. The radiographical pattern of involvement in our patient does not fit that produced by chronic osteomyelitis.

Osteopathia striata

Osteopathia striata, also known as Voorhoeve's disease, is a rare disease manifested by the radiographical findings of linear striations within bone. Multiple, 2-3 mm thick, linear sclerotic bands are seen aligned parallel to the long axis of long bones, and in the pelvis. This condition is asymptomatic and is usually an incidental finding.[2,5] The radiographical features of this entity are not present in our patient.

Multiple exostoses

Exostoses (or osteochondromas) are well-defined eccentric growths that protrude from the surface of bone. The cortex and trabeculae are continuous with those of the parent bone. The lesion has a cartilaginous cap, and is typically seen in the metaphysis of the distal femur, proximal tibia, proximal humerus, pelvis and scapula. Large exostoses may be associated with modelling deformity of the adjacent bone. Multiple exostoses (also known as diaphyseal aclasis) has a strong familial tendency and is inherited as an autosomal dominant trait. The significance of multiple exostoses is the increased incidence of malignant degeneration, reported to be in the region of 20% of cases. This diagnosis can be excluded in our patient, based on radiographical grounds. ■

References

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2. Milgram JW. *Radiologic and Histologic Pathology of Nontumorous Diseases of Bones and Joints.* Northbrook Publishing, Northbrook, IL. 1990; pp.181-200.
3. Benli T, Akalin S, Boysan E, *et al.* Epidemiological, clinical and radiological aspects of osteopoikilosis. *J Bone Joint Surg* 1992;74B:504-506.
4. Lagier R, Mbakop A, Bigler A. Osteopoikilosis: a radiological and pathological study. *Skeletal Radiol* 1984;11:161-168.
5. Helms CA. *Fundamentals of Skeletal Radiology.* 2nd ed. W.B. Saunders, Philadelphia, 1995.

CORRIGENDUM Errors occurred in this Radiological Conference by Dr. A.C.W. Chin & Professor W.C.G. Peh, *HK Pract* 20(5):288-290. Page 289, **Figure 3** is upside down and on page 290, under "Rheumatoid arthritis" second sentence "The classical pattern of joint disease is that of a symmetrical erosive arthritis of the synovial joint". It should be "a symmetrical" not "asymmetrical".