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Radiology of Metabolic Bone Disease: Workshop Report

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Ed. Note - This workshop was originally given at the International Symposium on Clinical Disorders of Bone and Mineral Metabolism, May 9-13, 1983.

S.R. Kottamasu illustrated an overview of metabolic bone disease with several clinical and radiologic case studies.

Case 1
A 55-year-old man with pain and limitation of motion of the left lower extremity. Radiographs demonstrated hyperostosis of the left hemipelvis, femur, tibia, and medial rays of the foot. The findings in the hemipelvis resembled Paget’s disease, although a wavy and sclerotic bone contour extending along one side of tubular bones, like flowing candle wax, is characteristic of MELORHEOSTOSIS (Figs. 1, 2).

Case 1. Melorheostosis.

Fig. 1
Case 1. Melorheostosis.

Fig. 2
Case 1. Melorheostosis.

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Case 2
A 15-year-old boy with blunt trauma to the foot. Radiographs demonstrated numerous, well-circumscribed foci of increased density in various bones, an incidental finding, characteristic of OSTEOPOIKILOSIS (Fig. 3).

Pathologically, the focal sclerotic areas were foci of compact bone in spongiosa.

Case 3
A 5-year-old boy with waddling gait and muscular weakness. Radiographs of lower extremities showed symmetric cortical thickening involving mid-diaphyses of femora (Fig. 4).

Radiographs of a 40-year-old man, father of Case 3, demonstrated sclerosis and marked cortical thickening with increase in the outer diameter of the bones. Metaphyses and epiphyses were conspicuously unaffected (Fig. 5). The findings were characteristic of ENGELMANN’S DISEASE (progressive diaphyseal dysplasia).

Case 4
Radiographs of the spine in a 45-year-old man showed typical “bone-within-bone” appearance. One of the classic conditions in the differential diagnosis of this radiologic finding is OSTEOPETROSIS. Subsequent radiographs demonstrated generalized osteosclerosis, absence of corticomedullary demarcation, and pathologic fractures of the proximal femur.

Case 5
Rickets in osteopetrosis. Some children with the congenital form of osteopetrosis develop radiologic changes of...
rickets and abnormal biochemical changes (low serum calcium, phosphorus, and increased alkaline phosphatase). The pathogenesis is not well understood, and treatment is unsatisfactory.

Case 6
A 55-year-old man with pain in the left lower thigh. Radiographs of the femur demonstrated multiple lytic lesions involving the distal femur. Some of the lesions were intracortical, and the lesions extended to the distal articular surface of the femur. The proximal end of the lesion had a “blade of grass” appearance, characteristic of the osteolytic phase of Paget’s disease, OSTEOPOROSIS CIRCUMSCRIPTA.

Case 7
Radiographs demonstrated multiple lytic lesions in the fronto-parietal region of the skull. Follow-up radiographs showed sclerosis and thickening of bone at the sites of previous lytic lesions (Fig. 6).

This case demonstrated spontaneous progression of the lytic phase of FIBROUS DYSPLASIA to the more commonly recognized sclerotic phase. This transition is less frequently documented in fibrous dysplasia than in Paget’s disease.

Case 8
A 45-year-old woman with skin rash and hepatomegaly. Skeletal radiographs demonstrated generalized mixed osteosclerotic and lytic pattern. This, associated with a brownish macular skin rash that urticated and hepatosplenomegaly, suggested the possibility of SYSTEMIC MASTOCYTOSIS (Fig. 7).

Of patients with mastocytosis, 15-20% have detectable bone lesions. Skeletal lesions in mastocytosis may be lytic, sclerotic, or mixed, and they may demonstrate focal, multifocal, or diffuse distribution.

Case 9
A 60-year-old man with generalized and progressive, severe skeletal pain and tenderness. Serum chemistries were normal except for a modest elevation of alkaline phosphatase (Fig. 8).

Radiographs demonstrated indistinct cortices, coarse trabecular pattern, and spotty areas of increased bone density. Outer diameters of all the bones of the hands were increased symmetrically, a feature characteristic of FIBROGENESIS IMPERFECTA OSSUM. There was loss of birefringence of collagen fibers on polarizing microscopy.
Case 10
A 60-year-old with generalized osteosclerosis and lytic lesions involving two ribs. A bone scan showed only minimal increased uptake in the lytic lesions in the ribs. Serum protein electrophoretic pattern was normal.

Bone biopsy demonstrated an OSTEOSCLEROTIC FORM OF MULTIPLE MYELOMA (Figs. 9,10). Table I lists the major causes of generalized osteosclerosis.

Case 11
A 35-year-old man presented with a pathologic fracture through a lytic lesion in the right tibia. The skeletal survey demonstrated additional lytic lesions involving the left tibia and fibula. Follow-up films in six months showed SPONTANEOUS HEALING OF BROWN TUMORS WITH SCLEROSIS FOLLOWING PARATHYROIDECTOMY.
Case 12
A 14-year-old girl with bilateral slipped capital femoral epiphysis. Subperiosteal resorption involved the phalanges of the hand. There was a lytic lesion in the left distal clavicle, consistent with a Brown tumor. The laboratory findings revealed persistent hypocalcemia, elevation of serum phosphorus, alkaline phosphatase and iPTH, and resistance to the administration of exogenous PTH.

This case represented RENAL RESISTANCE TO PARATHYROID HORMONE WITH OSTEITIS FIBROSA (pseudohypo-hyperparathyroidism).

Discussion
H.E. Meema discussed the periosteal and juxtaperiosteal components of hyperparathyroid bone resorption in chronic renal failure. Subperiosteal resorption in finger phalanges is a pathognomonic sign of hyperparathyroidism. It is generally considered to represent centripetal osteoclastic bone resorption from the periosteal surfaces, i.e. surface erosion. This does not fully explain why in some cases there remains a thin remnant of periosteal bone covering large juxtaperiosteal resorption spaces. A longitudinal microradioscopic study of the evolution of subperiosteal resorption in patients with chronic renal failure showed increased juxtaperiosteal resorption (a variant of intracortical resorption) in many areas where subperiosteal resorption subsequently developed. It thus appears that intracortical resorption correlates significantly with the extent of elevation in iPTH, although the correlation is higher for subperiosteal than for intracortical resorption.

H.K. Genant reviewed the clinical applications of computed tomography of the spine.

### TABLE I
Differential Diagnosis of Generalized Osteosclerosis

| 1. Osteoblastic metastatic bone disease |
| 2. Hyperparathyroidism - primary or secondary |
| 3. Myelosclerosis |
| 4. Osteopetrosis |
| 5. Lymphoma |
| 6. Pyknodysostosis |
| 7. Fluorosis |
| 8. Systemic mastocytosis |
| 9. Tuberosclerosis |
| 10. Van Buchem's disease |
| 11. Osteosclerotic multiple myeloma |
| 12. Fibrogenesis imperfecta ossium |

The normal ranges of vertebral mineral and age-related bone losses were determined from cross-sectional studies of 120 normal men and 203 normal women, aged 20-80 years. The normal mean value for young men and women was approximately 175 mg/cm. Men, by a linear regression, lost an average of 0.84%/year, and women, by a cubic regression, lost an average of 1.2%/year. Correcting for age-related bone marrow fat changes altered the observed rates of loss only by about 10%.

Cross-sectional studies in 80 perimenopausal women showed an average of 5.6%/year vertebral cancellous-mineral loss by QCT compared with 3%/year peripheral cortical mineral loss by photon absorptiometry and radiogrammetry. Longitudinal studies in 31 women following oophorectomy showed an average 9%/year vertebral cancellous bone loss, and peripheral cortical bone loss was 2-3%. Cross-sectional studies in 25 young women with various forms of amenorrhea showed an average decrement from normal of 30% in vertebral mineral values and 10% in peripheral cortical values.