The Osteodystrophies

A brief Review with Case Report

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THERE is a group of general diseases of bone, the pathology of which is obscure and which is characterized by a disturbance of calcium metabolism and of ossification. Boyd mentions among these: Osteitis Fibrosa, Osteitis Deformans, Osteomalacia, Rickets, Osteogenesis Imperfecta, Achondroplasia, Hereditary Chondrodysplasia and Marble Bone. Some writers consider these conditions to be closely related, particularly Osteitis Fibrosa, Osteitis Deformans and Osteomalacia and state that the different manifestations are caused by the age, vitality, etc. of the person affected.

Of the group, those which are liable to cause difficulty in diagnosis are: Rickets, Osteitis Deformans, Osteitis Fibrosa, local and general, and, rarely, Osteomalacia.

Bone tumors, particularly malignant bone tumors, must also be considered. However, they run fairly "true to form", and have certain general identifying characteristics which are easily borne in mind.

The Giant Cell Tumor develops in children and young adults, principally at the ends of long bones. Its gross appearance, feel and X-ray picture are characteristic. It is usually single. Osteogenic Sarcoma, a disease of the second and third decades, occurs at the end of long bones in the metaphysis. The X-ray appearance is characteristic. Ewing's tumor occurs at the age of 5-15 years. It is found in the shafts of long bones. X-ray shows diffuse involvement of the greater part of the shaft. Multiple myeloma occurs usually over the age of 40. Pain is a constant feature. The multiplicity of lesions and occurrence in flat bones is typical.

In healed Rickets with deformity the X-ray appearance will show bones deformed but otherwise normal. Blood examination is normal, i.e. calcium, phosphorus and "phosphatase reaction". "Plasma phosphatase" is an enzyme which hydrolyses phosphoric esters to inorganic phosphates, leading to a precipitation of calcium phosphate in bone. The plasma phosphatase is determined by the amount of inorganic phosphate converted from sodium glycero-phosphate in 48 hours at 38 degrees certigrade and a pH of 7.6 by the action of 1 c.c. of plasma. The normal is 0.15 mgm. but higher values are found in the bones of growing children. In the following diseases of bone the plasma phosphatose is increased ten to twenty times above normal: Osteitis Deformans (Paget's Disease); generalized Osteitis Fibrosa (in localized Osteitis Fibrosa the plasma phosphatase is normal); Malignant Metastases in bone and in Rickets.

Osteomalacia usually occurs in pregnant women but occasionally is seen in males. There is general decalcification of bones with softening, deformity and marked asthenia. Blood calcium is low, 5-8 mgm. per 100 c.c., the normal being 9-11 mgm. Phosphorus is low, phosphatase is high but, like calcium, the value tends to return to normal after giving calcium and Vitamine D. Calcium excretion in the urine is high.

Generalized Osteitis Fibrosa involves many bones. There are multiple cystic areas with little sclerosis and diminished density of bone shadows. Blood calcium is high, usually over 13 mgm. per 100 c.c. Phosphorus is low and phosphatase is high. Calcium and phosphorus are excreted in the urine in large quantities. It usually occurs in early adult life and later, and is associated with a tumor of the parathyroids.

Local Osteitis Fibrosa affects only one bone. Calcium phosphorus and phosphatase of the blood are normal. There is no change in the urine and the disease occurs chiefly in adolescence.

Osteitis Deformans occurs at a later period of life than Osteitis Fibrosa, usually between the ages of 40 and 65. There is first softening, then overgrowth of bone. A number of bones are usually affected but it may be confined to one bone and usually begins in the legs. In the advanced stage the appearance is highly characteristic. "The short, squat figure, with bent shoulders, curved back, sunken chest and great head hanging forward, as it waddles along, with bowed legs, out-turned toes, and the aid of a stick, is a living justification for the name Osteitis Deformans." The X-ray picture is characteristic. Blood calcium and phosphorus are normal while phosphatase may be high. Calcium and phosphorus excretion in the urine is normal.

Cases which present several or all of the individual characteristics of a given disease are easy to diagnose but nature does not always present the picture so clearly, as the following case report illustrates.

Mr. A. B., age 41, was admitted to hospital complaining of a "sore wrist" and "lumps on his legs".

He had first noticed five years previously that his right elbow was becoming stiff. The stiffness was progressive until in October, 1938, the elbow was flexed at 90 degrees and movement was limited to a few degrees. He also complained of small lumps on his right forearm and both legs. These also were first notice in 1933 and have progressively increased in size and number. They were not painful or tender.

History, except that incidental to the present disease, was negative. Except for "lumps" and elbow as described by the patient, physical examination revealed nothing. Of the extremities, only the left arm and forearm were normal. X-ray showed absence of lower portion of right ulna. There was a circumscribed area of rarefaction

with some expansion of cortex. The shaft of the radius was thickened and moderate inbowing was present. There was marked decalcification of carpal bones. Examination of skull and cervical, dorsal lumbar spine did not show any indication of Paget's Disease. Kahn and Hinton tests were negative. Blood picture was normal. Blood calcium, phosphorus and phosphatse were normal and there was no undue excretion of calcium or phosphorus in the urine. The parathyroids were explored at operation and no tumor was found, thus excluding a diagnosis of Osteitis Fibrosa. Biopsy showed "subperiosteal thickening or exostosis and in addition extensive resorption of bone, with the formation of a soft fibrous marrow, the marrow being composed of vascular fibrous tissue, and showing evidence of production of soft bone-like tissue—opinion a somewhat atypical Paget's Disease".

Thus, the final diagnosis was Osteitis Deformans. The subsequent progress of this patient will be watched with interest.

1. Boyd William: Text Book of Pathology.
2. Romans and Mitchner: Text Book of Surgery.
3. Nicholson: Laboratory Medicine.
4. Wright: Applied Physiology.
5. Hunter D.: Hyperparathyroidism. Generalised Osteitis Fibrosa. Brit. Jour. Surg. 1931. 19. 203.

"It may be remarked in passing that doctors are sometimes called silly because they spend their lives inventing ways of controlling disease, thereby lessening their business. That is a stupid view. By preventing the epidemics we keep the kids alive until they reach old age, opulence, and achieve rheumatism and high blood pressure. One rheumatic patient will produce more revenue than a whole epidemic of measles. I can prove it by my books."

—From "The Horse and Buggy Doctor"
by Arthur E. Hertzler, M.D.