Paget's Disease of Bone

Paget's Disease of Bone - not to be confused with Paget's disease of breast or with extramammary Paget's disease - is also known as osteitis deformans. It is one of several disorders of bone metabolism that involves inappropriate activation of osteoclasts and osteoblasts resulting in rapid bone remodeling and poor structure of affected bone. Unlike other systemic diseases, Paget's often localizes to certain bones, most commonly the skull, lower vertebrae, pelvis, and femurs. It also predisposes to secondary osteosarcoma (osteogenic sarcoma) and fibrosarcoma.



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Pathophysiology

Men > Age 40
Male-sex-symbol with Greater-than (40) Ounce
Paget's disease of bone has the highest prevalence in patients over the age of 55 with some cases seen over the age of 40. Paget's disease of the bone has a slightly higher prevalence in men than women.

Increased Osteoblast and Osteoclast activity
Up-arrow Ostrich-blast and Ostrich-claws
Initial bone resorption by overactive osteoclasts produces lytic lesions throughout bones and compensatory but disorganized bone remodeling by osteoblasts produces thick, sclerotic bone. Rapid turnover causes a decline in bone structure and strength.

Mosaic Pattern
Mosaic Pattern
Common findings on histology include increased vascularity, bone marrow fibrosis, osteocytes in lacunae, and absent Haversian systems.

Signs and Symptoms

Increased Skull Diameter
Up-arrow Skull Measuring-tape
As the calvarium thickens, patients with Paget’s may mention their hat size is increasing.

Hearing Loss
Plugged Ears
Problems that arise in Paget’s disease are most often in close proximity to the affected bone, so hearing loss in these patients only occurs when pagetic bone affects the skull. Pagetic hearing loss can occur due to auditory foramen narrowing in addition to nearby bone thickening with compression on the cochlea that is closely related to loss of bone mineral density in the cochlear capsule. Treating the underlying Paget’s disease may slow the progression of hearing loss.

Fractures
Fractured bone
Although the bone is thicker, it is poorly constructed, and thus is prone to fractures. “Chalkstick” fractures will occur perpendicular to the long axis of the bone, akin to breaking a piece of chalk in half.

Bone Ache and Joint Pain
Bone and Joint with Pain-bolt
Microfractures of weak bone will manifest with complaints of bone pain.

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High Output Heart Failure

High Output Dead Heart

High-output heart failure occurs when an otherwise normal heart cannot keep up with an unusually high demand for blood to one or more organs. Although the heart may be working well, it cannot pump out enough blood to keep up with the increased demand. Patients with Paget’s disease often have mosaic bone which contain numerous arteriovenous malformations or shunts, which collectively contribute to venous overload and eventual heart failure. High-output heart failure can also be caused by anemia, hyperthyroidism, pregnancy and beriberi.

Diagnosis

Bone Scan

Bone Scanner

Bone scintigraphy or bone scans allow for determination of bone metabolism. Radioactively labeled material is administered to the patient intravenously while the scan is occurring; more metabolically active areas of bone will show greater uptake of the material, and these areas are called “hot spots.”

Normal Parathyroid hormone (PTH) and Calcium

Normal Para-thigh-droid and Calcium-cow

Most patients with Paget’s disease of bone have normal calcium and PTH levels. Although patients can develop hypercalcemia due to osteoclastic bone resorption, particularly if they are immobile. Conversely, increased bone formation and calcium requirements in a person with Paget’s disease can lead to secondary hyperparathyroidism; if this need is not matched by an increase in calcium intake, hypocalcemia may occur.

Elevated ALP

Up-arrow ALP-skier

Osteoblasts utilize an enzyme called alkaline phosphatase (ALP) to produce phosphate used in bone mineralization. An elevated alkaline phosphatase level in these patients is a key lab finding. If serum ALP is not elevated but clinical suspicion remains high, bone specific ALP and urinary pyridinolines can be measured. ALP is also a tumor marker elevated when cancer metastasizes to bone or liver, as well as in seminomas (placental ALP).

Treatment

Bisphosphonate

Bike-phosphate-P

Such as alendronate, risedronate (and other -dronates) are pyrophosphate analogs that bind hydroxyapatite in bone and therefore inhibit osteoclast-mediated bone resorption. Patients taking bisphosphonates are advised to take the medication with water and remain upright for 30 minutes to avoid developing corrosive esophagitis. Another common side effect of some bisphosphonates is osteonecrosis of the jaw.

Calcitonin

Cow-throne

More of a second-line drug, calcitonin is used in patients who cannot tolerate bisphosphonates because of kidney disease or intolerable side effects. It also acts by inhibiting osteoclast function. Calcitonin can reduce pain and is somewhat effective in reducing new bone growth.

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