



PIC QUESTION OF THE WEEK: 11/08/10

Q: What is Paget's Disease?

A: Paget's disease (also known as osteitis deformans) is a rare bone disorder characterized by altered osteoclastic and osteoblastic activity that results in generation of deformed bone. In this condition, osteoblastic activity appears to exceed the rate of bone resorption, resulting in thickening and softening of bone in various skeletal sites including the skull, arms, legs, pelvis, spine, and collar bone. Osteoclasts enlarge and develop multiple nuclei; however, vascular supply remains intact. This bone disorder must be distinguished from Paget's disease of the *breast*, a cutaneous lesion associated with ductal adenocarcinoma. Paget's disease of bone occurs in approximately two to three percent of Caucasians and with almost equal frequency in men and women. The etiology of Paget's disease is unknown, but it has been hypothesized that the disease may be associated with viral illness and, as yet, unknown genetic factors. Infections that may have a positive correlation with Paget's disease include measles, pulmonary disorders associated with respiratory syncytial virus, and canine distemper viruses. The most common symptom associated with Paget's disease is severe and persistent bone pain. Other symptoms include joint pain and stiffness, bowing of the legs, headache, enlarged head, fractures, hearing loss, height reduction, and skull deformities. A large number of patients present with no additional symptoms. Enhanced osteoblastic activity accompanied by bone destruction result in dramatic increases in alkaline phosphatase levels. Radiologic findings include irregular patterns of resorption and formation in the diseased bones. Symptomatic relief may be provided by administration of NSAIDs. Although calcitonin is labeled for the treatment of Paget's disease, bisphosphonates are currently considered the treatment of choice. Bisphosphonates include etidronate (Didronel; the first labeled bisphosphonates for Paget's disease), alendronate (Fosamax), risedronate (Actonel), pamidronate (Aredia), and zoledronic acid (Reclast). There is little evidence that one bisphosphonate is clearly superior to another in the management of Paget's disease. Surgical options include joint replacement, osteotomy for extreme deformity, open reduction of fractures, and decompression procedures for the skull and spinal column.

References

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