

42. PAGET'S DISEASE OF BONE- A LITERATURE REVIEW

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Introduction. Paget's disease of bone (PDB) accounts for 2-4% of adults over 57, and is more frequent in males than females. PDB is the most common metabolic bone disease after osteoporosis. The most likely etiology is a slow paramyxovirus infection.

Aim of study. Paget's disease of bone proved a family history genetic susceptibility in 10-20% of cases. Bone damage occurs due to focal excessive bone resorption, abundant formation of the new bone structure and developed over time bone marrow replacement with vascular and fibrous tissue. Often, patients are diagnosed incidentally on X-ray, the most sensitive test being bone nuclear scintigraphy. Thus, the most common clinical presentations are bone pain, bowing and deformity of long bones, fractures, enlarged skull, hearing loss etc. It can affect a single bone (monostotic) or many bones (polyostotic). The main differential diagnosis is with sclerotic or lytic metastases.

Methods and materials. The nature, type and design of the research is narrative descriptive with the extensive systematic review of international and in-country academic and grey published literature with the main focus on Medline, PubMed, Google scholar and Google searching of websites to identify the range of the spectrum as well as most common forms of various clinical presentations of PDB, review of diagnostic tests and pharmacological and non-pharmacological treatment options. The desk research of published articles in peer-reviewed journals was supplemented by documentary analysis of published reports, key publications (including aide memories) and health care statistics provided by the WHO, NCBI, Directory of Open Access (DOAB) and online researches using Google scholar with the focus of the recent publications from the year 2000 and up to present. The initial study selection identified 88 articles and sources with the following inclusion criteria: 1.Be publically available in a published journal article or website; 2.Have a clear focus on good description of clinical presentation of PDB and treatment options; 3.Include a specific set of aspects to follow such as review of the most common clinical types of presentation, evolution pattern, differential diagnosis, various correlations, etc. The excluded articles were the ones which were limited to one specific aspect of PDB; focused on specific medical practice guidelines, clinical techniques and/or based on limited experience. The applied inclusion and exclusion criteria narrowed the list of the relevant articles to 47.

Results. According to the analyzed literature, the most common clinical manifestations of PDB are musculoskeletal, such as bone deformity and pain, fractures and spinal stenosis. The neurological complications are also common and include deafness, tinnitus, spinal stenosis with cauda equina syndrome, vascular steel, cerebellar dysfunction, obstructive hydrocephalus and various cranial nerve palsies. Para- or quadriplegia rarely occurs. Cardiovascular complications are uncommon. The most described in the literature are increased congestive cardiac failure, aortic stenosis and endocardial calcification. Hypercalcemia, hyperuricemia and immobilization hypercalciuria with nephrolithiasis count for metabolic complications of PDB. Very rare is described the neoplastic transformation of PDB in osteosarcoma, chondro- or fibrosarcoma.

Conclusion. Paget's disease of bone is the second commonest metabolic bone condition after osteoporosis. The follow-up monitoring of the patients for clinical relapses or complications development is mandatory. Common clinical manifestations include bone pain. Bone transformation results in enlargement leading to deformity, fragility, compression and, very rare malignant transformation. An elevated serum alkaline phosphatase reveals the disease activity. The long-term biochemical remission in Pagetic bone activity can be easily controlled by the intravenous bisphosphonate. For the difficulties resulting from deformity and abnormal biomechanics physiotherapy and occupational therapy proved helpful. However, patients can still have pain or problems from complications (e.g. secondary osteoarthritis). Orthopedic surgery for correction of deformity, joint total hip or knee replacements and nerve decompression are recommended for patients with PDB who develop osteoarthritis. However, hearing loss does not seem to respond to treatment of the PDB. Malignant transformation to an osteosarcoma has a poor prognosis.