Management of Paget's Disease of the Spine: Systematic Review and Case Series.

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Disclosures: Addisu Mesfin, Consultant Globus, Consultant GS Medical, Speaking fees Depuy Synthes, Research Support: Medacta, Globus, OREF, Kuros. The other authors have no disclosures.

INTRODUCTION: Paget's Disease of Bone (PDB) is the second most common metabolic bone disease. It is characterized by abnormal bone remodeling and deformity in the affected regions. The spine is the second most common location impacted by PDB. Decreasing rates and severity of PDB have reduced clinician experience with this condition. As a result, many do not recognize symptoms early, leading to delayed management and risk of suboptimal outcomes for patients with spinal PDB. This study is a systematic review focusing on the management of spinal PDB. Additionally, we included case presentations of patients treated for spinal PDB. Study Design/Setting: Systematic Review and Case Series.

METHODS: This systematic review was conducted using the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) guidelines. A retrospective protocol was used to search Medline (via PubMed) to identify all studies focused on spinal PDB. We did not include abstracts or non-published articles. Case reports and smaller case series (<8 patients) were not included. We analyzed all clinical studies available from 1/1969 to 12/2024 on PubMed and available through the ILLiad. On PubMed we identified articles with Key words using "Paget's Disease of Bone" and "Spine."

RESULTS SECTION: Originally 1209 unique citations were found between EMBASE and MEDLINE. Excluding articles that were not specific to spinal PDB resulted in a total of 15 papers representing 14 studies. 51 case studies on PDB of the spine were identified, and 55 patients were included in the analysis. The average age of patients in the cases was 59.6 years old (+/- 15.52 years), 42 were male and 31 were female. 12.7% had PDB in the cervical spine, 38.2% had PDB of the thoracic spine, and 47.2% had PDB in the lumbar spine. Only 10 cases had normal serum alkaline phosphatase at presentation. 43.6% of patients underwent surgery, 49.1% took bisphosphonate as part of their treatment, and 21.8% took calcitonin as part of their treatment.

DISCUSSION: X-ray, bone scans, and bone scintigraphy were the mainstay of imaging used to diagnose Paget's of the spine; vertebral DXA and QCT z-scores are higher in those with PDB of the spine, and CT scans may aid in determining the location and degree of disease and the extent of involvement of the spinal canal. Markers and imaging included Alkaline Phosphatase, Urine Hydroxyproline, serum Calcium, X-rays, Scintigraphs, and 99-Tmc, though some patients presented with normal serum alkaline phosphatase. Treatment is initiated only in those who are determined to be symptomatic from the disease. Management typically focuses on the use of calcitonin or bisphosphonates, and a portion of patients underwent surgery. The use of both surgical management and post-operative bisphosphonates resulted in better long-term neurologic recovery versus the use of only one or the other. Decreasing rates and severity of PDB have reduced clinician experience with this condition, leading to delayed recognition and risk of suboptimal outcomes. Variability in diagnostic pathways and the predominance of smaller case series limit generalizability.

SIGNIFICANCE/CLINICAL RELEVANCE: With the prevalence of PDB decreasing over time, spine surgeons are increasingly unlikely to be familiar with its presentation and management in the spine (6). However, failure to identify and properly manage this condition can lead to unnecessary suffering and unnecessary surgical intervention in these patients.

IMAGES AND TABLES: