The Libyan International Medical University

Faculty of Basic Medical Science

Paget's Disease & Risk Of Spinal Stenosis

Talal Mohamed Belrasali

2145

Supervised by: Esam El-Danaa

Assisted by: Lujin El-Shakmak

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Abstract

Introduction: Paget's bone disease interferes with the usual recycling process in your body, in which new bone tissue is gradually replacing old bone tissue. Over time, the disease can cause the bones affected to become fragile and disfigured. Most commonly Paget's bone disease occurs in the pelvis, skull, spine, and neck.

Materials and Methods: Of the 754 cases, 101 cases were of monostotic or polyostotic spine involvement. There were 29 cases of PDB in the spinal cord (16 males; 13 females) available for histological and histomorphometric analysis.

Results: The most common PDB manifestation site was the lumbar spine (62.2 %), followed by the thoracic spine (29.6 %). The cervical spine was involved in 8.2 % of patients.

Conclusion: In brief, the lumbar stenosis is the most common site in spinal stenosis

Aim of study: In this report you will find a brief introduction of vertebral column and definition of both Paget's disease and spinal stenosis separately. And the relationship between them with a focus on the most common site of spinal and why.

Keywords Paget's disease. Vertebra. Spinal stenosis.

Introduction

Typical vertebrae are a vertebral body, a vertebral spine, and seven processes. As the column goes down, the vertebral bodies increase in size. The vertebral body consists of a trabecular bone containing the red marrow, surrounded by a thin layer of compact bone from the outside. The arch, together with the back side of the body, forms the vertebral (spinal) canal containing the spinal cord.

Paget's disease of bone (PDB) is the second most common type of bone disease after osteoporosis. It is a disorder of the bone remodeling process due to an increase in osteoblastic and osteoclastic activity, that lead to resorb old bone and forms deformed, enlarged, and fragile new bones. There are factors that can increase your risk to get Paget's disease like Age (>40), sex and family history, and one of the complications of Paget's disease is spinal stenosis which is my topic.

Spinal stenosis is narrowing in the spinal canal space, which is lead to put pressure on the nerves that is inside it, that cause several symptoms differ according to the type of spinal stenosis. The 2 types of spinal Stenosis are:

-Cervical stenosis which is narrowing in the spinal canal at level of neck, that leads to upper limp weakness and neck pain.

-Lumbar stenosis which is narrowing in the spinal canal at the lower back and it's the most common type. This type can cause lower limp weakness and back pain.

The relation between the spinal stenosis and Paget's disease that PDB leads to overgrowth of bone, sometimes these overgrowths happen in the vertebrae, mainly in the arch, that lead to narrowing spinal canal which is called spinal stenosis and cause neural damage.

Materials and Methods

The files of the Hamburg Bone Register at the University Medical Center Hamburg-Eppendorf were searched for cases diagnosed as PDB.

Out of the 754 cases, 101 cases presented with monostotic or polyostotic involvement of the spine, those were reviewed in a retrospective study between January 1972 and

March 2010, whereas 29 cases were obtained at the University Medical Center Hamburg-Eppendorf. Details of all patients, including patient history, treatment course and clinical complications, were recorded; sALP levels were determined in units per liter (U/L) at the time of diagnosis. Characteristic histopathologic features were compared with radiologic findings for each patient. A total of 29 spinal PDB cases (16 males; 13 female) were available for histological and histomorphometric analysis. In those cases, a vertebral body biopsy—in addition to an iliac crest bone biopsy—was performed to confirm the diagnosis of a spinal manifestation of Paget disease. This study was carried out according to existing rules and regulations of the University Medical Center Hamburg-Eppendorf. All patients died in accidents or of acute disease. Review of hospital records and whole-body autopsy reports were used to exclude individuals with cancer, diabetes, glucocorticoid medication or donors on other drugs known to affect calcium metabolism. Therefore, patients with severe liver or kidney disease or periods of longer immobilization were excluded.[(1)]

Results

In our cohort of 754 patients, 101 cases involving the spine (13.4%) were recorded. The majority of cases were found in male patients (60.4%), whereas 39.6% were female. A total of 62 patients (61.4%) presented with polyostotic skeletal manifestation, while only 39 patients (38.6%) had monostotic involvement. In contrast to these findings, in the entire cohort of 754 patients more than 75% of patients presented with monostotic disease manifestation. The peak incidence of spinal PDB was found at 65.02 years of age with a small number of cases occurring in the fourth and fifth decade. A steady increase in cases was noted from the sixth decade onward and a peak disease incidence was reached during the eight decade.[(1)]

The lumbar spine was the most common site of PDB manifestation (62.2%), followed by the thoracic spine (29.6%). In 8.2% of patients, the cervical spine was involved. In each case, Here, typical radiological changes of pagetic vertebral bodies consisted of enlargement in width and height and reduced translucency leading to a typical ‘‘ivory vertebra’’ appearance. Serum ALP levels were elevated in 86.0% of patients with an average level of 256.05 U/L. The normal range for serum ALP set by our institution’s laboratory was 50–130 U/L.[(1)]

Discussion

PDB is a chronic, progressive condition with one or more bones involved. PDB skeletal lesions are characterized by increased osteoclastic bone resorption, increased but slightly disorganized bone formation, and increased bone vascularity. The number and size of osteoclasts is increased and may contain more nuclei than normal. The nuclei can contain bodies of inclusion similar to viral particles.[2]

It is suspected that the original lesion is a focal rise in osteoclastic bone resorption. Due to the rapid bone turnover, new collagen fibers are not arranged in an ordered linear fashion, but in a disorganized way. The resulting bone is a technically deficient matrix of woven and lamellar bone with increased risk of fracture or deformation.[2]

PDB is considered an osteo-clast disorder. Bone marrow and osteoclast circulating precursors show increased responsiveness to factors known to promote bone resorption such as 1,25 dihydroxy vitamin D and NF-kB ligand (RANKL) receptor activator. Increased expression and signaling of interleukin-6 (IL-6) may contribute to increased osteo-clastic activity. RANKL (stimulating osteo-clastic differentiation) expression is increased in pagetic marrow cells and high circulating RANKL levels have recently been identified in PDB patients. Nevertheless, osteoblasts are increased in numbers on pagetic sites, they are morphologically regular and are not considered by most authorities to be a key pathophysiological factor in PDB.[3]

In PD, the loss of homeostatic control results in increased osteoblastic and osteoclastic activity and is the background for the three main phases. The initial lytic phase represents a predominantly osteoclastic activity, the late osteoblastic phase is characterized by the formation of new bones, while the intervening mixed phase is seen when osteoblastic and osteoclastic activities are combined. Another phase, "inactive sclerotic phase," characterized by normal or decreased bone activity, was also identified when the stimulation of new formation of osteoblast and osteoclast ceases. Despite its metabolic inactivity, the bone retains a sclerotic coarsened architecture. The early lytic process is radiologically represented in bones with a low trabecular / cortex ratio such as the skull, femur, and humerus by a distinct leading edge at the normal bone interface. For bones with a high trabecular / cortex ratio such

as the vertebra, sacrum and pelvis, the lytic process is generally not detected. The presence of the vertebral body in radiological diagnosis is almost always complete and therefore the leading edge of the other affected bones is not seen in the vertebra. In the same patient and at the same time in various bones, including the vertebral column, these phases can be evident. While disease progression occurs within an infected bone, it is unusual to suddenly appear bone involvement at new skeletal sites years after initial diagnosis.[3]

The increased abnormal osteoblastic activity results in new bone formation (apposition) of periosteal and endosteal. Bone resorption (absorption) results from abnormal osteoclastic activity on the surface of the endosteal. The various combinations of these mechanisms result in four distinct bone remodeling patterns at the periosteum / endosteal interface leading to bone enlargement: periosteal and endosteal apposition; periosteal apposition and endosteal absorption; periosteal apposition with regular endosteal surface; and focal periosteal apposition – the appearance of "pumice stone". [3]

The processes generally responsible for changes in the periosteal and endosteal surfaces of the vertebral body and subsequent neural arches differ. These different mechanisms are not mutually exclusive, but can occur at different boundaries in combination in the same vertebra. In the vertebra involved, usually one of the pathomechanisms predominates. Periosteal apposition and endosteal absorption are the most common mechanism of expansion of the vertebral body. The new bone formation predominates on the periosteal surface and is responsible for the enlargement of the vertebral body, while the absorption on the endosteum results in an increased space for the bone marrow. Two less common remodeling mechanisms are the periosteal / endosteal apposition and periosteal apposition with normal endosteal surface. In both, the periosteal side apposition results in an enlargement of the vertebral body, but the size of the bone marrow is decreased or natural. The least common mechanism of expansion of the vertebral body is the focal periosteal apposition that gives the appearance of "pumice stone." Expansion of the radiologically detected vertebral bodies occurs in 63% of cases. [3]

A combination involving periosteal and endosteal apposition or periosteal apposition and endosteal absorption are the most common mechanisms in neural arch

involvement. The periosteal apposition in both systems causes the size of the spinal canal to decrease, resulting of spinal stenosis.(3)

Average vertebral body heights were measured and associated with the distribution of PDB in the spine in the skeletal-intact individuals. A strong correlation was found here. The height of the vertebral body, the more probable it became that the vertebral body was displaying pagetic changes.[1]

It can be further suggested that the height and size of the vertebral body may play an additional role in PDB incidences. This theory is supported by the fact that, if involvement of the cervical spine was known, C2 presented in this study as one of the primary disease locations. That is because C2 is significantly larger than the other six cervical vertebrae.[1]

Conclusion

In brief, the lumbar and C2 stenosis are the most common sites in spinal stenosis , some researchers said it’s the most common type because it’s the most movable part in the spine, but the other researchers said that when the spine goes down, the vertebral body increase in size and the arch of the vertebra decreases, and the lumbar vertebra has the biggest vertebral body and the smallest vertebral arch, so when the Paget’s disease occurs in the lumbar site, it will be no free space for the spinal cord. Therefore, the new bone will compress the spinal cord that will lead to numbness and pain in the lower limb.

References

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