## Ivory vertebrae: definition and differential diagnosis

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Learning objectives

The increase in density of the vertebral bodies is not a rare finding in imaging tests of the spine. For a proper interpretation of this finding on X-Ray (XR) and Computed Tomography (CT), it is important to understand the normal anatomy and histology, as well as the main pathologic basis of the diseases that may cause this finding. Thus this presentation aims to:

• Present the anatomy and histology of normal vertebral bodies as well as the factors influencing their appearance on imaging studies.
• Define the concept of an Ivory vertebra
• Present the main differential diagnoses that cause the Ivory vertebra, as well as the factors that influence its imaging appearances.
• Emphasize the radiological specificities of each pathology, that may assist radiologists in establishing the differential diagnosis.
Background

There is continuous expansion of access to diagnostic imaging in all parts of the world. Thus, common complaints in doctors' offices are being increasingly being investigated. One of these complaints is back pain. The first exam in the investigation is, in general, the XR and later proceeds to CT and MRI (magnetic resonance imaging). On the imaging analysis the radiologist may encounter vertebra whose density are greatly increased. These become much whiter than their usual color, resulting in a white comparable to that found on ivory, hence its name.

In order to correctly interpret the appearance of a vertebra seen on a radiography or CT, and to distinguish normal from abnormal, it is important to comprehend first the normal components and composition of the vertebra, and then understand the pathophysiology of the main diseases that can generate the Ivory Vertebra.

1 - Physiology and composition vertebral bodies

The vertebral column consists of 33 vertebrae, and has many important roles, in the support of the body, movement, and posture as well as in the protection of the spinal cord and nerve roots. The vertebrae are composed of a body, pedicles, lamina, spinous and transverse processes. The vertebral body is composed of thin outer layer of hard cortical bone on the outside, which coats a trabecular bone (cancellous bone) and less dense cancellous bone on the inside. The vertebral body reveals the presence of trabecular bone (cancellous bone) vascularized coated with a thin outer layer of compact bone, with the interstices being occupied by the trabecular bone marrow red and yellow. In figure 1 it is observed the typical aspect of a vertebra with their usual density.

The vertebral body is composed of a vascularized, trabecular (cancellous) bone, which has its interstium filled with red and yellow bone marrow, and is coated by a thin outside layer of cortical bone.

Figure 1 shows the typical aspect of the vertebrae, with its usual density.

When analyzing vertebrae images, the radiologist may find conditions which cause an decrease or an increase in mineral density. Ivory Vertebra refers to an increase in density of a vertebral body, which retains its size and shape, without any associated change in the opacity and color of adjacent intervertebral discs, at some stage in certain diseases. The increased opacity can be diffusely and homogeneous, involving most or all of the vertebral body, giving it a whitish appearance, in contrast to the normal appearance of the spine, or the possibly darker appearance of the osteoporotic spine. (1). Below are
presented the radiological characteristics of each of the major pathologies that cause the Ivory Vertebra, this could aid the radiologist in establishing the differential diagnosis between these pathologies.

2 - PATHOLOGIES (characteristics)

2.1 - Paget's disease of the bone (PDB) (Fig. 2 and 3)

Paget's disease (PD), also known as osteitis deformans, is a hypermetabolic bone disease that affects one (monostotic) or more (polyostotic) bones, and is characterized by areas of increased bone resorption mediated by osteoclasts, followed by disorganized osteoblastic bone healing. As a result of this process, there is disruption of the architecture of the bone tissues involved, which results in bone expansion and increased bone fragility (2).

It can affect almost any segment of the skeleton, but has a preference for the skull, spine, pelvis and long bones of the lower limbs. (2).

Paget's disease of the bone (PDB) is a very common finding in the elderly, with estimates ranging from 2.3 to 9% in older patients. Its onset is usually after the age of 55, with a slight predominance in males. (3).

Most patients with Paget's disease of the bone are asymptomatic, but some manifest symptoms such as pain due to the Paget's bone lesions, or as a consequence of the bone expansion and deformities, that may lead to osteoarthritis, nerve invasion, pain and tumor.

In one series, only seven of the 24 patients recognized as having Paget's disease were diagnosed during their lifetimes (29%) (4).

Histology: osteoclasts in patients with Paget's disease are bizarre in appearance, multinucleated and excessive in number. As a consequence, there is abnormal deposition of lamellar bone interspersed with normal bone tissue. The bone appears disorganized, and presents thickening of the trabeculae, which is surrounded by numerous osteoblasts, forming the mosaic pattern (most characteristic).

Features that support the diagnosis:

It should be noted that during the evolutionary phase of Paget's disease there are three distinct phases, as cited by Greenspan (5):
1. Lytic or hot phase: active bone resorption with radiolucent wedge or elongated area with sharp edges that destroy cortical and cancellous bone.

1. Intermediate phase: bone destruction is accompanied by neo-osteogenesis, with the latter process usually predominating. Diffuse enhancement of the opacity with sclerosis most significant in the periphery and a radiolucent center.

2. Cold or sclerotic phase: diffuse increase in bone density occurs along with the increase and expansion of the bone. There is prominent cortical associated with blurring of the transition point between cortical and cancellous bone. These features, in a way, limit the inclusion of Paget's disease in the definition of an Ivory Vertebra.

Ivory vertebra is observed in the cold or sclerotic phase.

- Trabecular bone is thickened, and there is an increase in the anteroposterior and lateral diameters, resulting in increased vertebral size, which may lead to complications such as spinal stenosis and nerve root compression.

- Homogeneous vertebral body involvement.

- More than one affected site (polyostotic).

- An Ivory Vertebra for which a etiology is not found and remains unchanged over time, may be due to asymptomatic Paget disease of the bone.

2.2 - LYMPHOMA

Lymphomas constitute a heterogeneous group of primary malignancies of the lymphoid tissue. Characteristically the malignant lymphocytes accumulate in the lymph nodes and produce the typical clinical picture of lymphadenopathy. Occasionally, they may invade the blood ("leukemic phase") or infiltrate organs outside the lymphoid tissue. The lymphomas are divided into Hodgkin's disease and non-Hodgkin lymphoma, based on the presence of Reed-Stenberg cells (Hodgkin lymphoma).

Approximately 40% -50% of the cases of lymphoma envolve the skeletal system, usually through metastatic disease. This finding, however, is frequently observed only at the autopsy study and not in radiographs. (6)

Pathophysiology:

The lymphomatous deposits can initiate a marked osteoblastic response in cancellous bone, leading to the formation of new bone, which results in an diffusely sclerotic
appearance. Linfomatous focis involve the vertebral body through hematogenous spread or contiguous invasion from adjacent lymph nodes.

Radiological Features:

Such involvement results in osteolysis or osteosclerosis, or a combination of both. Lytic lesions are more common. Osteoblastic involvement is rare, and usually secondary to Hodgkin lymphoma. In their study, Granger and Whitaker (7) found that of the 210 lesions column for Hodgkin's disease, only 13 had the ivory appearance.

Thus some features may help establish the diagnosis as shown in figure 4:
- Increase in the size and density of the trabecular bone, resulting in trabecular pattern loss and in some cases (13%) the appearance of the Ivory Vertebra.
- Associated paraspinal masses.
- Anterior and lateral margins of the vertebral body may show erosions caused by the surrounding mass or the involved para-aortic lymph node.

2.3 - Blastic Metastases

Blastic metastatic lesions are most commonly derived from prostate or breast neoplasms, however, other metastatic tumors, including lymphomas must be considered; along with rarer lesions such as plasmacytoma, chordoma, or primary bone sarcomas (1).

Pathophysiology:

In blastic metastatic lesions there is stimulation of osteoblasts, resulting in an irregular replacement of the vertebral cancellous bone tissue with a dense and amorphous masses, that may become confluent.

Radiological features:

Some features help your diagnosis:
- They are more common in patients with breast or prostate cancer.
- But, in the elderly, they may also represent metastasis of lung, breast, kidney or colon cancers. In children they may be secondary to neuroblastoma.
- In most cases there is involvement of multiple vertebral levels. Other bones may also be affected.

Such characteristics can be observed in a 62 year old patient with prostate cancer. During his staging, osteoblastic lesions were diagnosed at different levels of the lumbar spine and hip (Figs. 5, 6 and 7).

A radiopaque vertebral body, on one or more vertebral levels, in an elderly with metastatic disease, until proven otherwise is the result of prostate carcinoma.

2.4 - Osteomyelitis (Fig. 12)

Osteomyelitis is an infection that results in progressive inflammatory bone destruction, followed by formation of new bone.

It is difficult to obtain reliable information on the overall incidence of vertebral osteomyelitis. Two studies published in 1979 and 2001, estimated the incidence of vertebral osteomyelitis to be 1:250,000 [3] and 1:450,000, respectively (8).

Osteomyelitis can be divided into acute, when it develops over several days or weeks, and chronic when it is prolonged.

Pathophysiology

The pathogens can reach the spinal bones by three basic forms.

- hematogenous spread from a distant infectious site
- direct inoculation by trauma or spinal surgery
- contiguous spread of infection from adjacent soft tissues

Acute osteomyelitis features suppurative inflammatory cells, accompanied by edema, vascular congestion, and thrombosis of small vessels. In the initial acute disease, the vascular supply to the bone is impaired by the infection, which is spreading to the surrounding tissue. When both medullary and periosteal blood supplies are committed, large areas of dead bone (Sequestrum) can form.

Due to loss of blood supply, the dead appears whiter than normal bone. The cancellous bone is absorbed rapidly, and can be completely destroyed or sequestered in two to three weeks, but the cortical bone may require two weeks to six months to become necrotic. After the complete separation, the dead bone is slowly eroded by granulation tissue and absorbed (9)
Radiological features

- Osteomyelitis produces sclerosis in a vertebra during the healing phase, but this condition rarely involves a single vertebra.

- Normally, the erosive changes are present in the margins of the intervertebral disc, so this helps to guide diagnosis.

- Patients should be asked about possible predisposing factors or events, including underlying diseases, hospitalization, invasive procedures, intravenous drug use, and travel.

Increased erythrocyte sedimentation rate (ESR), may also be a clue.
Fig. 1

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**Fig. 2:** Fig 2 e 3: Patient age 67 years, asymptomatic. XR in the AP profile demonstrates increased density of the L4 vertebral body. This aspect remained unchanged with time.

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Fig. 3: Patient age 67 years, asymptomatic. XR in the AP profile demonstrates increased density of the L4 vertebral body. This aspect remained unchanged with time.

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Fig. 4: Fig 4 and 5: Patient, 62 years, with lymphoma and back pain. XR in the PA and LAT shows diffuse increase in bone density of the vertebral body of L4, characterizing an Ivory Vertebra.

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**Fig. 5:** Fig 4 and 5: Patient, 62 years, with lymphoma and back pain. XR in the PA and LAT shows diffuse increase in bone density of the vertebral body of L4, characterizing an Ivory Vertebra.

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Fig. 6: Figs. 6, 7, 8, 9, 10 and 11: XR and CT images, where osteoblastic lesions were observed scattered all vertebral bodies and also found in bones of the pelvis

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**Fig. 7:** Figs. 6, 7, 8, 9, 10 and 11: XR and CT images, where osteoblastic lesions were observed scattered all vertebral bodies and also found in bones of the pelvis.
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Fig. 9: Figs. 6, 7, 8, 9, 10 and 11: XR and CT images, where osteoblastic lesions were observed scattered all vertebral bodies and also found in bones of the pelvis

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Fig. 10: Figs. 6, 7, 8, 9, 10 and 11: XR and CT images, where osteoblastic lesions were observed scattered all vertebral bodies and also found in bones of the pelvis

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Fig. 11: Figs. 6, 7, 8, 9, 10 and 11: XR and CT images, where osteoblastic lesions were observed scattered all vertebral bodies and also found in bones of the pelvis

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Fig. 12

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Imaging findings OR Procedure details

Images acquired with incidences of RX PA and profile and CT 16-channel axial, coronal and sagittal.
Conclusion

Because the Ivory Vertebrae is, in some cases, the first finding observed in common pathologies (such as prostate carcinoma, lymphoma and Paget’s disease), it is important for the radiologist to know its radiographic characteristics. Knowing this entity, he may help the attending physician formulating the differential diagnosis, and could also perform an active search for other injuries that may be associated with it.
References


