Journal of Frailty, Sarcopenia and Falls

Case Report Article

Vertigo as the first manifestation of paget's disease: a case report

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Abstract

Paget's disease of bone is characterized by the dysfunction of the bone architecture due to the extreme increase of the osteoclast and osteoblast activity which results in the production of pathological bone. The particular disease afflicts any skeleton bone. Its sources have not been specified yet. There is evidence for the existence of strong genetic background as well as the effect of environmental factors. The neurological complications are relatively rare and are related to the central and peripheral nervous system (headache, ataxia, vertigo, loss of hearing, smell etc.). In our case, we present a patient with vertigo as the first manifestation of Paget's disease.

Keywords: Paget disease, Vertigo, Bone disease, Neurological complications, Osteitis deformans

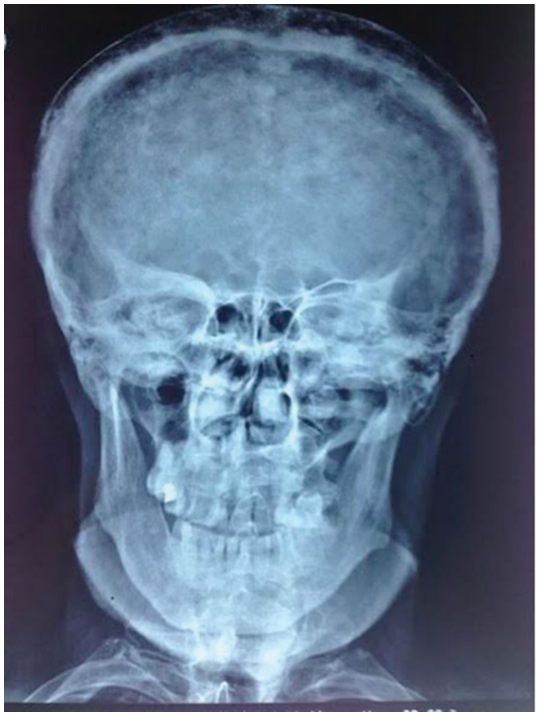

Figure 1. Paget's disease by the 'cotton wool' appearance in the x-ray film.

Introduction

Paget's disease of bone is a rare disorder that occurs in patients under 40 years old[1,2]. Neurologic manifestations of Paget's disease before the radiographic appearance of the bone changes is an uncommon presentation of the disorder[3]. Early neurological symptoms include headache[4], dizziness, deafness, tinnitus and pain due to radicular compression[3]. The uncontrolled growth of bones results in pressure imposed on the conterminous soft tissues, such as brain[5], spinal cord and particular peripheral nerves[3,5,6]. In our case, we report a patient presented at the emergency department with vertigo and finally diagnosed with Paget's disease.

Case report

Treatment was conducted in compliance with the international biomedical studies stipulations, with reference

The authors have no conflict of interest.

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Edited by: *George Lyritis*

Accepted *25 February 2017*

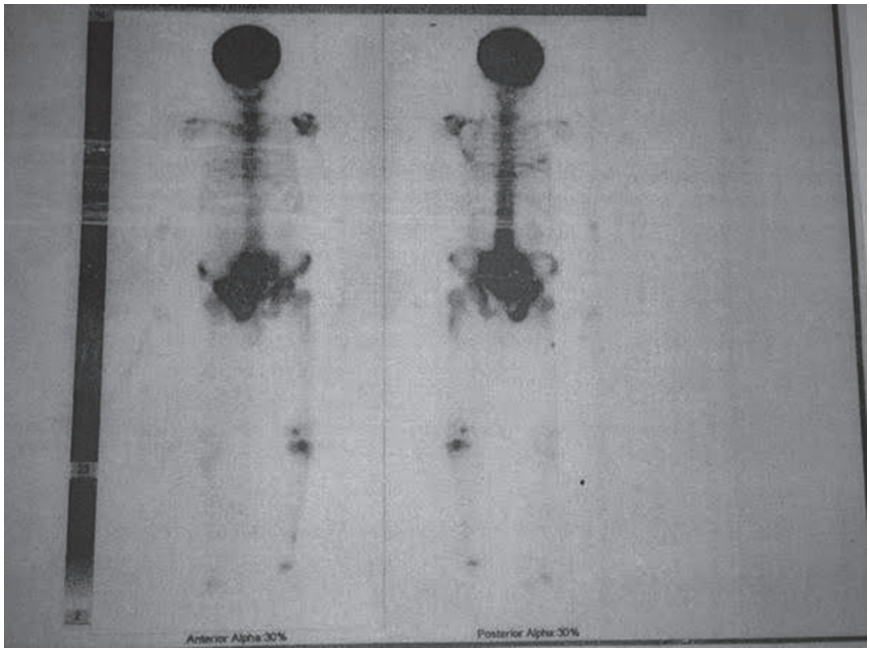

Figure 2. Full body scintigraphy compatible with multifocal Paget's disease

to the Declaration of Helsinki of the World Medical Association. No personal data of the patient were kept, while it was impossible to trace back to any personal data from the data collected for the present study. The present study was approved by the Department of Internal Medicine at the General Hospital of Athens "*Elpis*".

A 70-year-old man was admitted at the emergency department of our hospital complaining for vertigo and nausea for about an hour ago, before admittance. His medical history included coronary artery disease (coronary artery bypass graft surgery 15 years ago), hypertension and epileptic crisis. His medication included clopidogrel, atenolol, enalapril, hydrochlorothiazide and valproic acid. The clinical examination did not reveal pathological findings.

The blood chemistries showed increased levels of alkaline phospatase (ALP): 616 IU/ml (normal values: 50-136), gamma-glutamyl transferase (γGT): 173 IU/l, creatinine: 0,74 mg/dl urea: 76 mg/dl, serum calcium: 9,1 mg/dl, serum magnesium: 1,6 mg/dl and lactate dehydrogenase (LDH): 275IU/l. Additional blood tests included: parathyroid hormone (PTH): 55,5 pg/ml (normal values 15.0-65.0) (>30: adecuacy), thyroid stimulating hormone (TSH): 0.463 (0.35-4.94), 25-hydroxy-vitamin D (VIT 25-OH-D): 18.60 (10-30: Deficiency). The rest blood exams did not reveal pathological findings.

A brain computed tomography (CT scan) revealed disperse bone-diluting and bone-thickness skull areas, an image which is compatible with Paget's disease of bone. The skull X-ray imaging revealed high and low-density areas (Figure 1), while the pelvis X-ray imaging revealed multifocal Paget's disease on the ilium (fattening of iliac crest) and the pubic bone.

The full body bone scintigraphy revealed increased fixation to the bony skull, pelvic bones, inferior cervical spine, body of T9 vertebra, the inferior lumbar spine and along left tibia and left shoulder blade (Figure 2). As a result, the patient was diagnosed with multifocal Paget's disease. The patient was immediately treated with risedronate 30 mg o.d. for 2 months and the patient was scheduled for re-examination.

During re-examination the blood chemistries depicted 50% decline of ALP: 238 IU/l, PTH: 189, 5 pg/ml, serum calcium: 9.25 mg/dl, VIT 25-OH-D:14.50 mg/ml. The patient did not report any neurological symptoms. We decided the termination of risedronate sodium treatment while aledronate sodium 70mg once a week was initiated. A new re-examination of the patient was scheduled after 6 months.

Discussion

Paget's disease of bone or *osteitis deformans* is a chronic bone abnormality, which may affect a single, several, or many bones, but never involves the entire skeleton[7]. Paget's disease affects both men and women, with an apparent small male predominance. It rarely manifests itself clinically before the age of 40 years while the frequency of the condition

increases with advancing age. The prevalence is estimated to be approximately 2-3% among individuals over the age of 55 years[8]. Paget's disease of bone is associated with involvement of the central and peripheral nervous system. Lesions of the central and peripheral nervous systems are due to the frequency in which the disorder affects the skull and spine and the close anatomic relationship to neural tissues. Neurological syndromes include headache, dementia, brain stem and cerebellar dysfunction, cranial neuropathies, myelopathy, *cauda equina* syndrome, and radiculopathies[6,9].

Neurologic complications of the spine occur in 4-10% of the patients[10,11], while the neurological complications of cranial disease, excluding auditory involvement are rare[12]. In advanced cranial disease there may occur an anatomic lowering of the softened base of the skull onto the upper cervical vertebrae resulting in brain stem and cerebellar compression (basilar invagination)[13]. This may cause obstructive hydrocephalus and compression of the cerebellum, lower cranial nerves, pyramidal tract, and upper cervical nerves. The severity ranges from asymptomatic to acute tonsillar herniation and death. In general, there is a slowly progressive neurologic syndrome, advancing over a period of 1-5 years. Symptoms and signs include occipital headache, ataxia, vertigo, tinnitus, dysphagia, dysarthria, and progressive cerebellar and corticospinal tract signs. Basilar impingement may contribute to vertebrobasilar insufficiency and to obstruction of venous return[14].

Serum ALP concentrations and urine hydroxyproline are usually increased in patients with neurologic complications. Plain radiographs and radionuclide scans are important to localize disease activity and the former can often show pathologic fractures. Magnetic resonance imaging scanning is crucial to determine compression of neural structures and to exclude other causes. It is recommended that asymptomatic disease affecting the skull or spine must be treated with bisphosphonates. One of the newer bisphosphonates such as pamidronate, alendronate, risedronate, or tiludronate is recommended as they do not cause the mineralization defects seen with etidronate[12,15]. Chronic, slowly progressive neurologic deficits should initially be treated with bisphosphonates[12].

Conclusions

In conclusion, although the neurological complications of the Paget's disease are rare, clinicians should be aware of the disease because early recognition and appropriate treatment are the cornerstones for achieving a better outcome.

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