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Calvarial Paget's Disease Complicating Severe Platybasia and Basilar Impression: MRI Features

Case Report

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ABSTRACT

We herein describe a case with calvarial Paget's disease associated with basilar impression and platybasia leading to bulbomedullary junction compression. The patient was presented with the symptoms of increasing deafness, loss of balance, dysphagia, and neck pain spreading to the arms and skull. The diagnosis of basilar impression and detailed measurements of the craniocervical junction could be possible by the use of magnetic resonance imaging. MRI should be the preferred technique in such cases.

Key words: *Paget's disease, Platybasia, Basilar impression, MRI*

Şiddetli Platibazi ve Baziler Impresyona Yol Açan Kalvaryal Paget Hastalığı: MRG Özellikleri

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ÖZET

Biz burada platibazi ve bazilar invajinasyon nedeniyle bulbomedüller bileşke basısına neden olan Kalvaryal Paget olgusunu sunduk. Olgu artan işitme azlığı, denge kaybı, kollara ve kafatasına yansıyan boyun ağrısı ile başvurdu. Bu tür olgularda kranioservikal bileşkenin değerlendirilmesi için manyetik rezonans görüntüleme mükemmel bir tanı tekniğidir.

Anahtar Kelimeler: *Paget Hastalığı, Platibazi, Bazilar Etki, MRI*

INTRODUCTION

Paget's disease of bone is a chronic osseous disease characterized by bony enlargement or deformity and bone fragility (1). It is common in males and tends to increase in prevalence with age. Although viral aetiology has been proposed, the exact cause is unknown (2). Majority (>90%) are asymptomatic and are detected incidentally due to raised alkaline phosphatase or when radiographs are done for other reasons. Pain (80%), deformity (15%), fractures (9%), and malignancy (<1%) occur in those with symptoms. Majority, are polyostotic. In some cases the disease may be limited to the calvarial bones presenting with diploic thickening, and signal intensiy changes. One of the most common complication of calvarial Paget's disease is basilar impression associated with secondary platybasia. MRI is an excellent technique in terms of craniocervical junction evaluation and demonstration of brainstem and spinal cord compressions (1,3,4).

CASE REPORT

A 62-year-old female patient known to have Paget's disease restricted to the calvarial bones for 8 years presented with the symptoms of increasing deafness, loss of balance, dysphagia, and neck pain spreading to the arms and skull increasing for one year. Neurological examination revealed mild weakness of extremities.
Cranial MRI examination performed following initial examinations revealed widening of the diploic space with mixed signal intensity on all sequences consistent with Pagetoid involvement of the calvarium. Also paranasal sinuses and mastoid air cells were obliterated due to the involvement of bones. Visual assessment and than measurements of craniovertebral junction on the sagittal T1-weighted image showed basilar impression. The tip of the odontoid process had extended 12 milimeters above McGregor's line leading to compression of the bulbomedullary junction. Increased Welcher basal angle (155°), and a decreased Wackenheim craniovertebral angle (124°) was consistent with marked platybasia.

To relieve the symptoms of the patient, elective suboccipital craniectomy was performed the next week. The postoperative period was uneventful and than the patient was discharged.

DISCUSSION

Paget's disease is a chronic disorder of the skeletal system. Its origin is not definitely known. However the current thinking is that a slow virus may be the initiating factor. The disease affects 3-4% of the population over the age of 40 but it is more frequently found in the elderly. The men are affected slightly more than women. Paget's disease may be monostotic or polyostotic. Monostotic disease is not common (10-20%). Pelvis, sacrum, lumbar spine, thoracic spine, femur and calvarium are the most frequently affected bones. The disease progresses through three phases. First the initial phase of increased osteoclastic activity results in bone resorption. This early phase is osteolytic and is not commonly seen radiologically. It may persist in the skull, as osteoporosis circumscripta. In the mixed phase, increased resorption of bone is followed by increased formation of abnormally coarsened trabeculae of increased volume (mosaic pattern). In the last phase named as sclerotic phase osteoclastic activity declines and osteoblastic activity proceeds. Disorganized new bone of increased density replaces lytic areas. Eventually the disease becomes quiescent (3-5).

Basilar invagination is the protrusion of odontoid process into the foramen magnum (McGregor's line is the line from posterior hard palate to base of occiput: if dens protrudes more than 6 mm then basilar invagination is diagnosed). The causes are condylus tertius, condylar hypoplasia, basiocciput hypoplasia and

atlanto-occipital assimilation. The acquired form of basilar invagination is termed as basilar impression. The causes of the disease are Paget's disease, osteomalacia, rickets, fibrous dysplasia, hyperparathyroidism, Hurler syndrome, osteogenesis imperfecta and skull base infection (1,3,4).

Secondary basilar impression is associated with advanced Paget's disease of the skull quite often. Hearing loss, the most common complication, is due to pressure on the auditory nerve caused by involvement of the petrous bone. Seventh nerve involvement apparently does not occur. Primary basilar impression, usually a skeletal derangement of the craniovertebral junction, is known to present svmptoms of compression of the spinal cord, the brain stem, the cerebellum, the basilar and vertebral arteries, the fourth ventricle and the foramina of Magendie and Luschka (1,6).

The neurologic complications result from pagetic involvement of the skull. Expansion of diseased bone can result in compression of cranial nerves as they exit their bony foramina (hearing loss, blindness, facial palsy). Softening of the skull leads to basilar invagination with compression of the brain stem, cerebellum, and lower cranial nerves. Brain stem compression can cause hydrocephalus (4,5).

In conclusion, MRI is an excellent technique to demonstrate the complications of Paget's disease especially in the evaluation of craniovertebral junction.

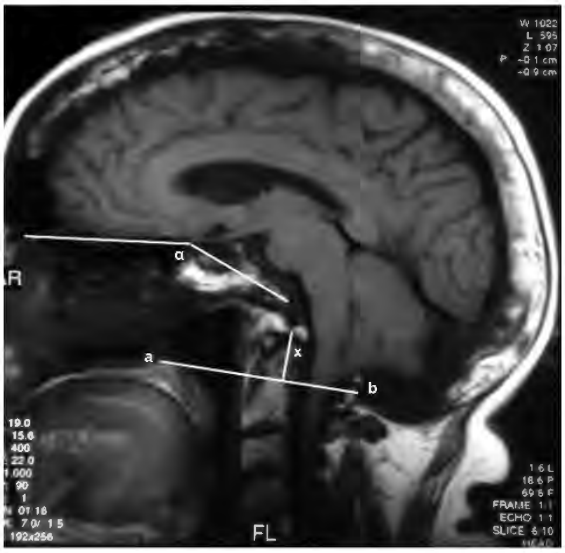

Figure 1: *Sagittal T1-weighted MR image of the craniovertebral junction demonstrates platybasia (increased Welcher basal angle: 155°) and basilar impression (the odontoid tip extends 12 mm(x) above McGregor's line). The image also depicts the compression of bulbomedullary junction with the retroflexed odontoid tip, and the decreased diameter of foramen magnum.*

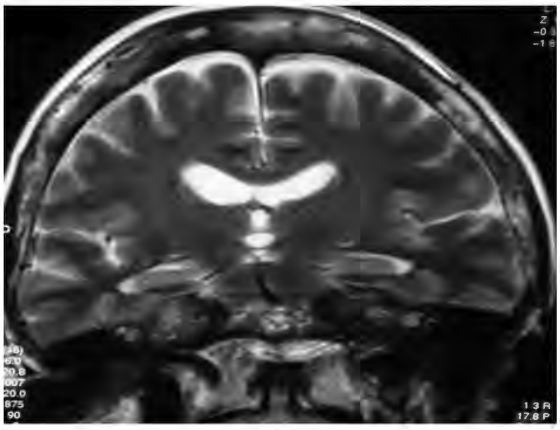

Figure 2: *Coronal T2-weighted MR image of the cranium demonstrates the compression of brainstem and diencephalon.*

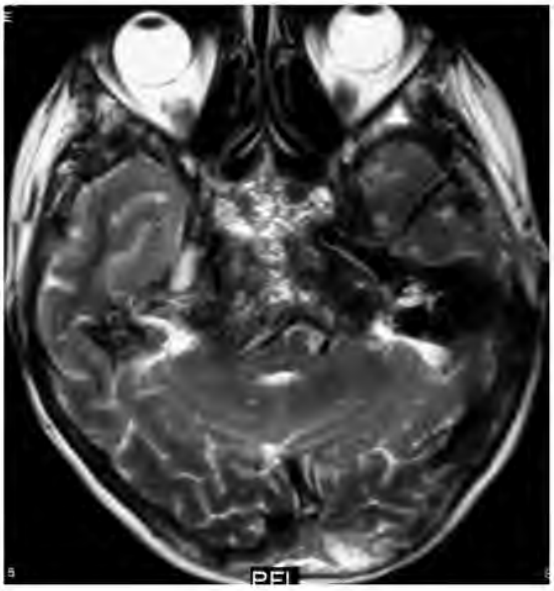

Figure 3: *Axial T2-weighted MR image of the cranium at the level of internal acoustic channels. Normally we should see pons at this level, however in this case we see the bulbus with marked compression from the right side. The internal acoustic channels seem narrowed. Also the calvarial diploic spaces seem heterogeneous and widened.*

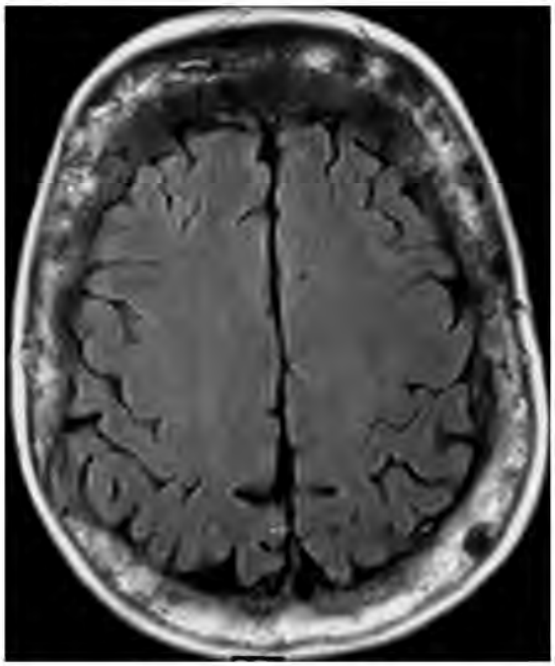

Figure 4: *Axial FLAIR-weighted MR image of the cranium at the level of centrum semiovale reveals irregular inner borders of tabula interna and externas in addition to enlarged and heterogenous diploic space.*

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