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Case Report

Paget's disease of bone presenting with multiple cranial nerve palsies: A case report ☆,☆☆

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ABSTRACT

Paget's disease of bone (PDB) is a progressive monostotic or polyostotic osteopathy with unknown cause. It is associated with the involvement of the nervous system. The cranial nerves, spinal roots, cauda equina, spinal cord, and brain can be affected in PDB due to their close anatomical relation to bone. Hearing loss occurs in 12%-50% of patients with PDB. The optic nerve can be affected at the optic canal. The diagnosis of PDB is radiological by highlighting characteristic lesions like thickening of the cortical bone, hypertrophic and fibrillary bones. Progressive or chronic neurological deficits should be treated with bisphosphonates. We present a rare case of multiple cranial nerve palsies as the first manifestation of PDB.

Introduction

First described by James Paget in 1886, Paget's disease of bone (PDB) is a progressive monostotic or polyostotic osteopathy with unknown cause. It occurs in patients above 50 years old with male predominance and represents the second acquired osteopathy behind osteoporosis [1].

The involvement of the nervous system in PDB is infrequent. The uncontrolled growth of bones can affect the brain, spinal cord, and cranial nerves. Headaches, dizziness, and deafness are the most common neurological signs of the disease [2]. In this report, we present an unusual case of multiple cranial nerve palsies as the first manifestation of PDB.

Case report

A 60-year-old man was admitted to our neurological department for a progressive decrease in visual acuity in the right eye. Three months before admission, he reported hearing loss of the right ear, vertigo, and continuous right hemifacial neuralgia. His medical history included hypertension treated with enalapril.

The neurological examination revealed a conscious patient with a supple neck. Examination of muscle strength and sensitivity was normal. Muscle stretch reflexes were normal. Motor coordination was normal. On ophthalmological examination, visual acuity was 1 of 10 in the right eye and 7 of 10 in

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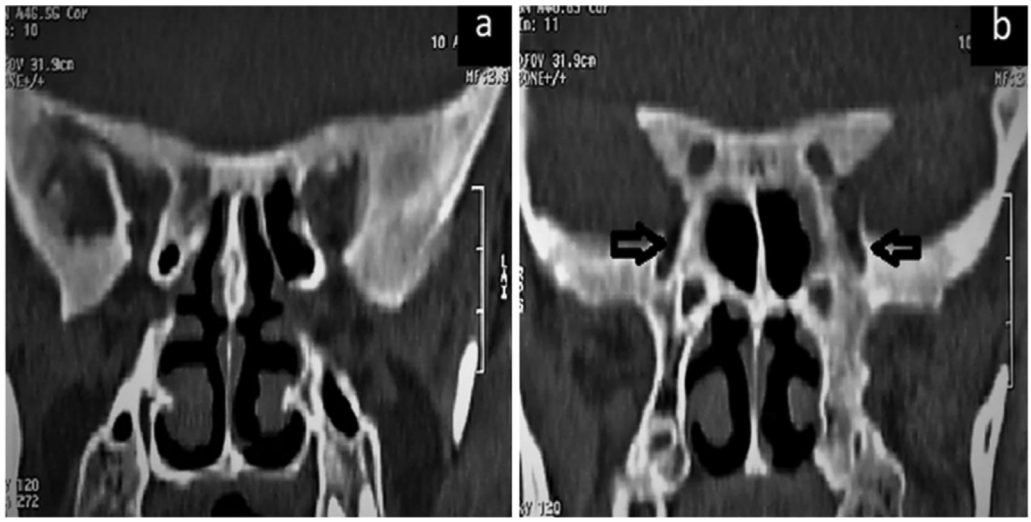

Fig. 1 – Facial CT scan with coronal reconstruction centered on the anterior (A) and medial (B) cranial fossa of the skull base identifies widening of the diploic space with typical "cotton wool appearance", and stenosis of both foramen rotundum more pronounced on the right side (arrows).

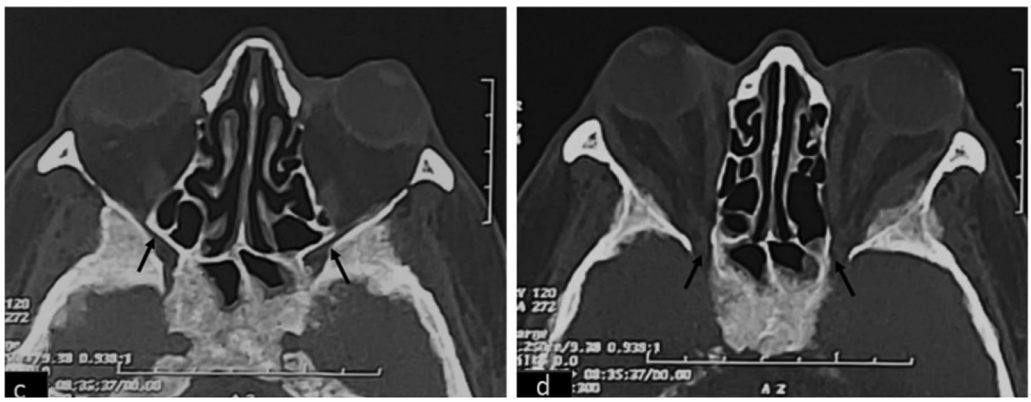

Fig. 2 – Facial CT scan with axial reconstruction centered on the skull base revealing thickened and disorganized trabeculae with narrowing of the inferior orbital fissure (C) and of the optic nerve canals (D) (arrows).

the left eye. The right direct pupillary light reflex and the left consensual pupillary light reflex were absent. A fundus examination revealed papilledema in the right eye. Examination of oculomotor nerves was normal. There was a bilateral V2 and V3 dermatome hypoesthesia without trigger zone. The patient had no facial palsy. There was a right hearing loss of perception. The audiogram revealed that the decrease in air conduction was parallel to the decrease in bone conduction. The lower cranial nerves were intact. The examination of other extra neurological systems was unremarkable.

Facial computed tomography showed on coronal reconstruction an over-riding enlarged bone, widening of the diploic space, typical 'cotton wool appearance' and stenosis of both foramen rotundum more pronounced on the right side (Fig. 1). Axial reconstruction demonstrated thickened and disorganized trabeculae, narrowing of the optic nerve canals and of the inferior orbital fissure (Fig. 2).

Cerebral MRI showed on T2 weighted images cortical thickening, expansion of the sphenoidal bone and of the petrous temporal bones (Fig. 3). CISS sequence through the cerebellopontine angles revealed bilateral asymmetric stenosis of the internal acoustic meat with compression of the right acoustic-facial complex nerves (Fig. 4).

Blood tests revealed an alkaline phosphatase (ALP) level of 2200 U/L (reference range, 32-91 U/L). Large paraclinical tests were performed and all went negative (serum calcium, serum magnesium, lactate dehydrogenase, parathyroid hormone, 25-hydroxy-vitamin D, thyroid hormone, hemogram, serum electrolytes, renal and hepatic tests).

As a result, the patient was diagnosed with multiple cranial nerve palsies caused by PDB. He was immediately treated with risedronate 30 mg/day and carbamazepine. Three months after the treatment, the patient noticed a gradual improvement of the trigeminal neuralgia, while hearing loss and visual acu-

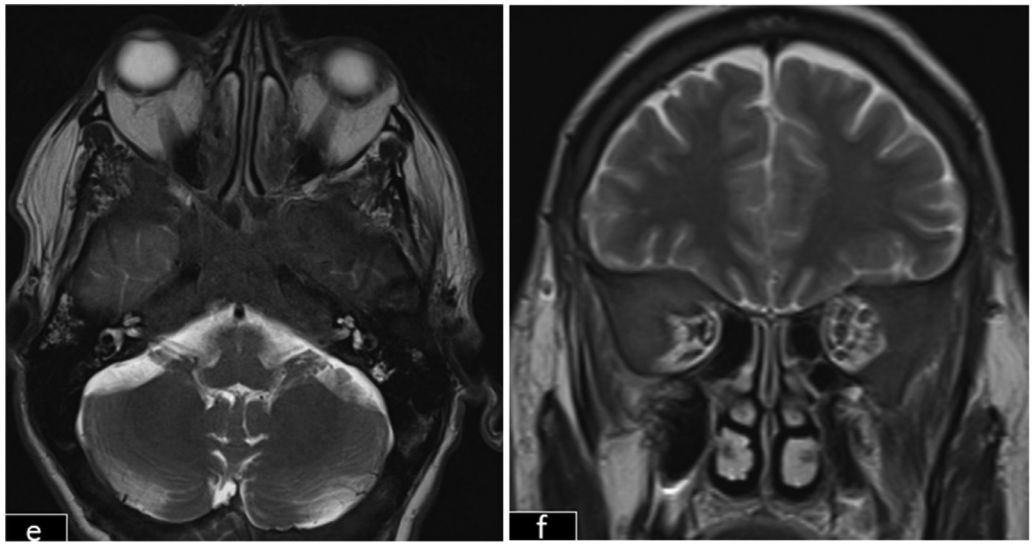

Fig. 3 – Cranial MRI on axial (E) and coronal (F) T2 weighted sequence showing thickened cortex, coarsened trabeculae with an expansion of the sphenoidal bone and of the petrous temporal bone.

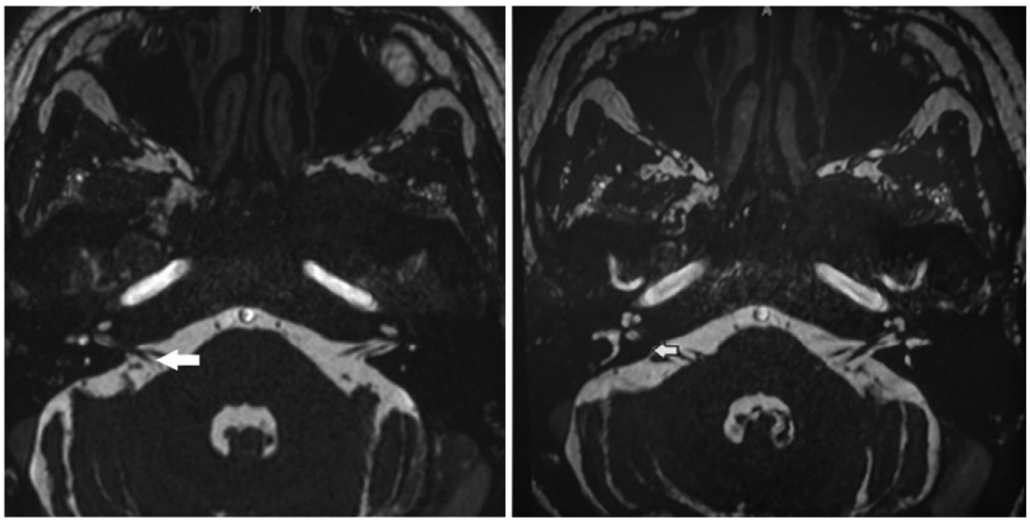

Fig. 4 – CISS sequence through the cerebellopontine angles showing stenosis of the internal acoustic meati more pronounced on the right side with compression of the right acoustic-facial complex nerves (arrows).

ity remained stable. Blood tests showed 40% decline in ALP. We decided the initiation of alendronate sodium 70 mg once a week while risedronate sodium treatment was terminated. A new reexamination of the patient was scheduled after 3 months.

Discussion

PDB is a focalized osteopathy touching single or many bones without including the entire skeleton. The prevalence of PDB represents 2%-3% of patients over 55 years of age [3]. In 40% of cases, PDB is discovered in the asymptomatic phase before the appearance of bone deformities. PDB etiology remains unknown, but virus hypotheses and genetic factors were pronounced [1].

PDB is associated with the involvement of the nervous system. The cranial nerves, spinal roots, cauda equina, spinal cord and brain can be affected in PDB due to their close anatomical relation to bone. Neurological syndromes are myelopathy brainstem, cerebellar dysfunction, cauda equina syndrome, radiculopathies and cranial neuropathies [4].

Apart from auditory involvement, the neurological complications of PDB are rare [5]. In the study by Clarke et al [6], 34%

of 96 patients diagnosed with PDB had cranial nerve involvement. 13 patients had an auditory involvement, one patient had a trigeminal sensory loss, and no case of optic nerve involvement.

Hearing loss occurs in 12%-50% of patients with PDB. It is caused by a lesion of the cochlea or by compression of the eighth cranial nerve in the auditory canal. The optic nerve can be affected by compression of the optic canal causing reduction of vasa nervorum flow. Clinical manifestations are diminished vision, papilledema retinal, optic atrophy, and choroiditis. Trigeminal neuralgia and facial numbness can be caused by a narrowing of the foramens of the branches of the trigeminal nerve [7].

The diagnosis of PDB is radiological. Plain radiographs show characteristic lesions such as thickening of the cortical bone, hypertrophic or fibrillary bones. Body scintigraphy increases the diagnostic sensibility of the disease. MRI scanning is important to demonstrate the compression of neural structures and to exclude other causes [5].

Urine hydroxyproline and serum ALP concentrations are increased in patients with neurological complications in PDB due to the osteoblast hyperactivity [1].

Few cases of cranial nerve palsies in PDB have been previously reported in the literature [8–10]. Our patient had typical radiological lesions of PDB with evidence of multiple cranial nerve palsies.

It is recommended that progressive neurological deficits should initially be treated with bisphosphonates [5]. The therapeutic efficiency in PDB with multiple cranial nerve palsies is not well documented. Bisphosphonates can stabilize hearing loss. Calcitonin with etidronate could stabilize the optic neuropathy [11]. Trigeminal neuralgia improves with carbamazepine. Intravenous biphosphates is an alternative to oral bisphosphonates treatment failure [12].

Conclusion

The neurological complications of PDB are rare and can be severe. Clinicians should consider PDB as a possible cause of neurologic symptoms. MRI scanning, body scintigraphy and serum ALP levels enable the diagnostic of PDB. Therapeutic management of PDB with involvement of the nervous system should be adapted to the clinical presentation and based on bisphosphonates, decompressing of neural structures, and symptomatic treatment.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.03.028.

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