PAGET'S DISEASE REVEALED BY MULTIPLE NEUROSENSORY COMPLICATIONS: A CASE REPORT

MULTIPLES COMPLICATIONS NEUROSENSORIELLES RÉVÉLANT UNE MALADIE DE PAGET

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ABSTRACT:

Background: Paget's disease is an osteodystrophy characterized by anarchic remodeling. The combination of neurosensory and neurological complications is an exceptional disease's discovery circumstance.
The objective is to study different aspects of this complicated form.
Here a case revealed by deafness which was combined with multiple neurological and neurosensory complications.
Observation: A 43-year-old man complained of hypoacusis evolving for three years, combined to tinnitus, headaches, vertigo and scalp dysesthesia. Examination revealed an increased head circumference and prominent frontal bosses. Examination showed a normal otoscopy, multidirectional nystagmus, a right pyramidal syndrome and a static cerebellar syndrome. Radio-biological examination was in favour of Paget's disease. During his exploration, the patient had developed axonal optic neuropathy. He had biphosphonates treatment. The evolution was favorable with a follow-up of 24 months.
Conclusion: Combined neurosensory and neurological complications is an exceptional and serious situation of Paget's disease revelation.
Key-words: Paget's disease - Optic Neuropathy- Sensoryneural Deafness

RÉSUMÉ

Introduction: La maladie de Paget est une ostéodystrophie acquise caractérisée par un remaniement osseux excessif et anarchique. La survenue de complications neurosensorielles est un mode exceptionnel de révélation de la maladie.
Le but est de présenter les différents aspects de cette forme compliquée de la maladie.
Observation: Un homme âgé de 43 ans a consulté pour une hypoacousie droite évoluant depuis trois ans, associée à des acouphènes. Il présentait par ailleurs des céphalées holocrâniennes et un vertige rotatoire. L'examen craniofacial a objectivé une augmentation du périmètre crânien et une proéminence des bosses frontales. L'examen vestibulaire a objectivé un nystagmus multidirectionnel. L'examen neurologique a montré un syndrome pyramidal droit et un syndrome cérébelleux statique, le tout orientant vers une maladie de Paget. Au cours de l'évolution, il a développé une neuropathie optique axonale. Il a reçu des cures de biphosphonates. L'évolution était favorable avec un recul de 24 mois.
Conclusion: Les complications neurosensorielles et neurologiques représentent exceptionnellement un mode de révélation sévère de la maladie de Paget.
Mots-clés: Maladie de Paget - Surdité neuro-sensorielle - Neuropathie optique

INTRODUCTION:

Paget's disease (PDB) is a benign acquired osteodystrophy characterized by excessive and anarchic bone remodeling [1]. Clinical presentation is dominated by bone pain, deformities and pathological fractures. Neurosensory complications are rare. According to review articles published in 2014 and 2022 [2, 3], deafness prevalence varied from 2,4 to 11% in PDB. However, optic nerve involvement is far rarer than deafness, six cases had been reported based on a literature review article [4]. The association of neurosensory complications is an exceptional situation [3].
We report an exceptional case of PDB revealed by a combination of neurological and neurosensory complications including deafness.

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OBSERVATION:

A 43-year-old man, with no previous pathological history, complained of progressive right-sided hypoacousia that had been developing for 3 years, associated with annoying whistling-like tinnitus. The patient also reported holocranial headaches, non-positional rotational vertigo lasting a few seconds and scalp dysesthesia. The examination revealed an increase in head circumference and a more pronounced prominence of the frontal bumps on the right (figure 1). Otoscopy was normal. Vestibular examination revealed spontaneous multidirectional nystagmus. The neurological examination showed a right pyramidal syndrome and also a static cerebellar syndrome without intracranial hypertension. Ophthalmological examination was initially normal. Pure-tone audiometry revealed right sensorineural hearing loss (threshold at 50 dB). The standard x-ray of the skull showed an appearance of cotton-like spots in typical mosaic of PDB (figure 2). Non-enhanced computed tomography (CT) of the brain and facial area showed the appearance of bone dysplasia affecting the entire vault and base of the skull, as well as the maxillary bones (figure 3). Alkaline phosphatase levels were significantly elevated. Taking into account these data, the diagnosis of a cranial PDB was made. The extension assessment based on technitium scintigraphy was negative. During his exploration, the patient complained of a rapid decline in vision. Control ophthalmological examination revealed a decrease in visual acuity, fundoscopic examination was normal. Orbital-cerebral magnetic resonance imaging (MRI) did not show optic nerve compression. Visual evoked potentials concluded that there was axonal optic neuropathy. The patient had bisphosphonates treatment. Evolution had been favorable with a stability of the neurosensory lesions. The patient has been followed for 24 months since the treatement's end.

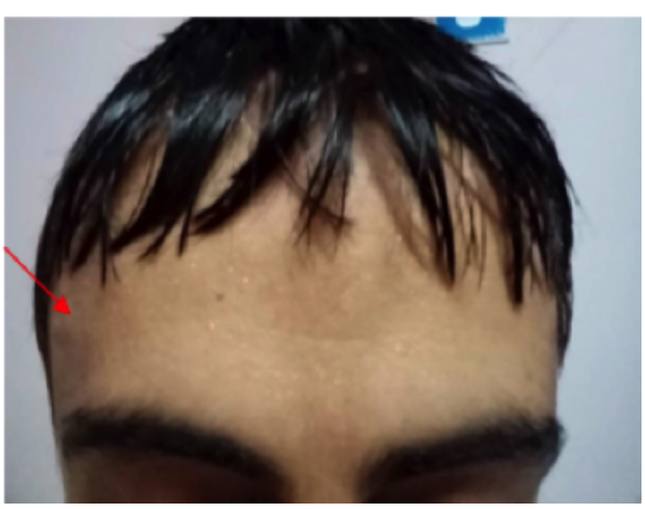

Figure 1: prominence of the frontal bumps more marked on the right (red arrow)

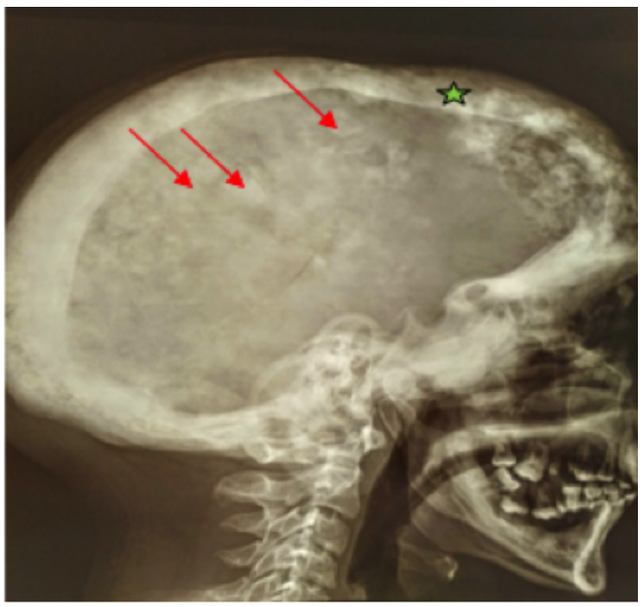

Figure 2: X-ray of the skull in profile showing a cottony spotted appearance (red arrows) and thickening of the calvarium (green star)

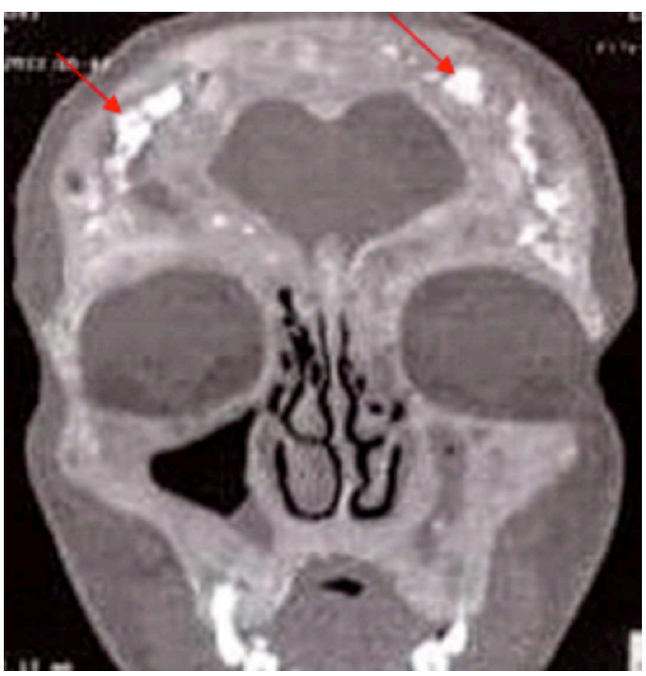

Figure 3: cerebral and facial CT in coronal section showing an appearance of bone dysplasia (red arrows)

PDB represents an imbalance between osteoformation and osteoresorption leading to the formation of fragile and deformable bone [5]. It is predominant elderly subjects' pathology which results in mechanical bone pain, bone deformities and pathological fractures [6]. However, it can be asymptomatic in more than 50% of cases [7]. Neurosensory complications of PDB are rare and occur late [7]. The originality in our case is the rich association of multiple neurological and neurosensory complications constituting an exceptional mode of revelation of PDB. Deafness is the most common neurosensory complication affecting 2,4 to 11% of patients with PBD [2, 8]. Pagetic deafness is classically a unilateral and progressive deafness of mixed type.

More rarely, it may be sensorineural hearing loss as our patient. Pure conductive hearing loss in PDB is extremely rare. The pathogenesis of pagetic deafness is complex: stapedo-vestibular ankylosis, damage of Corti organ, compression of the cochlear nerve in the internal auditory canal, elongation of the cochlear nerve in the posterior fossa, compression of the cochlear vestibular nuclei at the brainstem and finally vascular disorders particularly in Corti organ [9]. Evolution is towards stability or decline. Cophosis is exceptional. Optic neuropathy is the second neurosensory complication of PDB that also our patient presented. The origin is most often a compression of the optic canal leading to a reduction in the flow of the vasa nervorum. Some authors implicate a vascular rather than mechanical factor in the pathogenesis of this neuropathy, particularly in patients with a normal MRI as was the case of our patient [10]. Clinical manifestations are decreased vision, papillary retinal edema, optic atrophy and choroiditis. Furthermore, the oculomotor nerves may be compressed at the superior orbital fissure, causing paralysis of the oculomotor muscles and diplopia. Neurological complications result from bones pagetic damage of the skull and spine. Skull expansion compresses the cerebrum, cerebellum, and brainstem [11], this would be the cause of the headaches, cerebellar syndrome, central vestibular syndrome and pyramidal syndrome presented by our patient. Moreover, rare cases of cranial nerve palsy have been described due to nerve compression in the skull base foramina [8]. In addition, PDB can leads to ischemic myelopathy, cauda equine syndrome or radiculoneuritis due to spinal hypertrophy [11]. PDB treatment is based on bisphosphonates and calcitonin. Complicated forms, as our patient, often require multi-disciplinary treatment. Control of pagetic pathology (normal alkaline phosphatase levels) is not correlated with resolution or improvement of neurosensory and neurological complications. Treatment for pagetic deafness can be medical, surgical or with hearing aids. The use of calcitonin to treat hearing loss in Paget's disease is controversial. Although it provided hearing stability in some patients, it induced ototoxicity in others [12]. Hearing gain can be obtained by a hearing aid or by otological surgery. The surgical procedure may consist, depending on the case, of a stapedectomy, a stapedotomy, an ossiculoplasty or nerve decompression. However, the long-term functional results of surgery are uncertain and depend on the stabilization of the pagetic process. Thus, faced with an evolving PDB, a prosthetic solution seems more reasonable. Faced with nerve damage, medical treatment based on bisphosphonates or calcitonin is the first-line treatment [13]. Etidronate calcitonin could stabilize optic neuropathy [14]. Trigeminal neuralgia is sensitive to carbamazepine. However, surgical nerve decompression is possible as a last resort [14]. In cases of hydrocephalus, ventricular bypass may be useful. Acute compression of the brainstem, spinal cord, or nerve roots requires urgent surgical decompression.

CONCLUSION:

Paget's disease of bone revealed by neurosensory and neurological complications represents an exceptional situation which deserves to be known. Indeed, ignorance of it can leads to a diagnosis error and therapeutic delay. Complications evolve on their own independently of the stability of the PBM, therefore, the earlier the initiation of their treatment the better their prognosis.

Conflict of interests

There are no conflicts of interest.

REFERENCES:

1. Muschitz C, Feichtinger X, Haschka X, Kocijan R. Diagnosis and treatment of Paget's disease of bone: a clinical practice guideline. Wien Med Wochenschr. 2017 Feb;167(1_2):18-24.
2. Tan A, Ralston SH. Clinical Presentation of Paget's Disease: Evaluation of a Contemporary Cohort and Systematic Review. Calcif Tissue Int. 2014 Nov;95:385–92
3. Choi YJ , Sohn YB , Chung YS. Updates on Paget's Disease of Bone. Endocrinol Metab (Seoul). 2022 Oct;37(5):732-43
4. Kerr NM, Cassinelli HR, DiMeglio LA, Tau C, Tüysüz B, Cundy T,et al. Ocular Manifestations of Juvenile Paget Disease. Arch ophthalmol. 2010 June;128(6):698-703
5. Banaganapalli B, Fallatah I, Alsubhi F, Shetty PJ, Awan Z, Elango R,et al. Paget's disease: a review of the epidemiology, etiology, genetics, and treatment. Frontiers in Genetics [Internet]. 2023;14.
6. Alaya R, Alaya Z, Nang M, Bouajina E. Maladie osseuse de Paget : actualités diagnostiques et thérapeutiques. Rev Med Interne. 2018 Mars;39(3):18591.
7. Audran M, Baslé MF. La maladie osseuse de Paget. Diagnostic. Suivi évolutif. Rev Rhum. 2003 Sept;70(8):6347.
8. Hamid M, Touarsa F, Satte A, Bourazza A. Paget's disease of bone presenting with multiple cranial nerve palsies: A case report. Radiology Case Reports. 2022 Juin 1;17(6):18703.
9. Calvet J, Coll J, Dussert J, Pelletier L. Les Surdites Dans la Maladie de Paget. International Audiology. 1967 Janv 1;6(1):6871.
10. Isasi C, Sanz J, Hijós M, Vaquero M, Saucedo G, Andreu JL. Successful treatment of optic neuropathy in osteitis deformans. Rheumatology. 2002 Août 1;41(8):948-50
11. SFO | SFO Rapport Site Generation [Internet]. [cité 30 mars 2023]. Disponible sur: https://www.em-consulte.com/em/SFO/2020/sforender/B97822947638300000049.html
12. Rubin DJ, Levin RM. Neurologic complications of Paget disease of bone. Endocr Pract. 2009 Mars;15(2):15866.
13. Cotten A, Silva J, Moisan S, Cortet B, Boutry N. Place du scanner et de l'IRM dans la maladie de Paget et la dysplasie fibreuse. Rev Rhum. 2003 Sept;70(8):66677.
14. Poncelet A. The neurologic complications of paget's disease. J Bone Miner Res. 1999 Oct;14 Suppl 2:88-91.