Doença de Paget Evoluindo com Malformação de Chiari Tipo 1 como Causa de Manifestações Otoneurológicas - Relato de Caso e Revisão de Literatura

Paget's Disease Evolving to Type 1 Chiari Malformation as a Cause of Otoneurological Manifestations - Case Report and Review of Literature

Francis Zavarezzi*, Luciana Matshie Matumoto*, Aracely Fernandes Duarte*, Ulisses José Ribeiro**, Sérgio Bittencourt**, Flávio Augusto Sekeff Sallem.***

* Médico (a) (Residente do Serviço de Otorrinolaringologia do Hospital Nossa Senhora de Lourdes - São Paulo / SP).
** Médico (Chefe do Serviço do Otorrinolaringologia do Hospital Nossa Senhora de Lourdes - São Paulo / SP).
*** Médico (Neurologista do Hospital do Servidor Público Estadual - São Paulo / SP).

Serviço de Otorrinolaringologia do Hospital Nossa Senhora de Lourdes – São Paulo/SP.
Endereço para correspondência: Rua das Perobas, 295/344 04321-120 – São Paulo / SP.

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RESUMO

Introdução: Doença de Paget evoluindo com Malformação de Chiari tipo 1 é caso raro descrito na literatura, constando apenas três relatos. Sintomas otoneurológicos são freqüentes na evolução da doença . A doença de Paget consiste em uma osteíte deformante, autossômica dominante. Em 50% dos casos cursa com perda auditiva. A malformação de Chiari tipo 1 é uma doença rara, predominante em adultos, e é uma das causas de vertigem de origem central. É caracterizada por deslizamento das tonsilas cerebelares através do forame magno.

Relato do Caso: Relatamos o caso de um paciente do sexo masculino, 57 anos, com história de vertigem, tinitus, hipoacusia bilateral, cefaléia e cervicalgia há cerca de 6 anos, evoluindo em cinco meses com disfagia e ataxia de marcha. A fosfatase alcalina sérica apresentava importante aumento (2100Ui/l).

Conclusão: À Ressonância Magnética de ossos temporais, observa-se achados ósseos sugestivos de Doença de Paget e sinais de Malformação de Chiari tipo 1.

Palavras-chave: doença de Paget, malformação de chiari tipo1, manifestações otoneurológicas.

SUMMARY

Introduction: Paget's disease evolving to type 1 Chiari malformation is rarely described in medical literature, and only three case reports have been published so far. Otoneurological symptoms are frequent on disease course. Paget's disease is a dominantly inherited disorder. Hearing loss is seen in 50% of cases. Type 1 Chiari malformation is a disorder which frequency is high in adults, and is one of the causes of central vertigo. It is characterized by tonsil herniation through the foramen magnum.

Case Report: We report the case of a 57-year-old male patient who presented with vertigo, tinnitus, hearing loss, headache and cervical pain for six years, progressing in five months to dysphagia and gait disturbance. Seric levels of alkaline phosphatase were markedly high (2100Ui/l).

Conclusion: At temporal bone magnetic resonance imaging, bone abnormalities which suggested Paget's disease and signs of type 1 Chiari malformation were seen.

Key words: Paget's disease, type 1 chiari malformation, otoneurological manifestations.

Introduction

Paget's disease, also known as deforming osteitis, is a localized chronic disorder that occurs when the body loses control over the normal process of bone remodeling, and is characterized by excessive bone resorption followed by overproduction of new bone (1,2). It may be related to infection by paramyxovirus (1), but its etiology remains unclear (4,5). An autossomal dominant pattern of inheritance has been disclosed (5), predominantly in 40-year-old and over Caucasian males (M:F 1,5:1) (5,6,7). Patients are asymptomatic in most cases; however, Paget's disease may be accounted for axial bone chronic pain, joint pain, skeleton deformities, hearing loss, and compression of cranial nerves (4). Expansion of cranial bones affecting temporal bones may lead to hearing loss because of inner ear lesion (6). Most often Paget's disease is discovered incidentally, when radiographs or lab tests are requested for other reasons. Its diagnosis is made by signs and symptoms, physical examination, imaging procedures and levels of bone metabolism markers, mainly seric alkaline phosphatase (6). Hearing loss occurs in about 50% of the cases, and it occurs due to alterations in bone density, shape and mass, which altogether contribute to the delicate harmony of synchronized movements between the middle and inner ear (8). Symptoms like vertigo and tinnitus can also be seen (1, 7, 8, 9).

Type 1 Chiari malformation is a rare disorder, most seen in adults, and is a cause of vertigo of central origin. It occurs when cerebellar tonsils slide through the foramen magnum into the cervical canal (10, 11). Most commons symptoms are pain, asthenia and headache, and these are often preceded by otoneurologic manifestations. Sensorineural dysacusis, vertigo, nystagmus, ataxia, tinnitus, and other cranial nerve disorders have been reported with this condition (10, 12, 13). Audiologic tests and imaging procedures, especially magnetic resonance imaging (MRI), are of extreme importance because they display alterations in the cerebellopontine angle region (12). The otolaryngologist and the neurologist must understand and recognize type 1 Chiari malformation as part of the differential diagnosis of balance disorders, because this may be the initial manifestation of this disease (13).

We report a case of Paget's disease evolving to type 1 Chiari malformation, its course and prognosis.

Case Report

A 57-year-old Brazilian caucasian male, who worked as a cabinet-maker, came to our otolaryngology outpatient service complaining of tinnitus and bilateral hearing deficit for six years, vertigo and loss of balance for about two years. Personal history disclosed diabetes mellitus (DM) and systemic hypertension (SAH), treated with diet, glibenclamide and hydroclorotiazide.

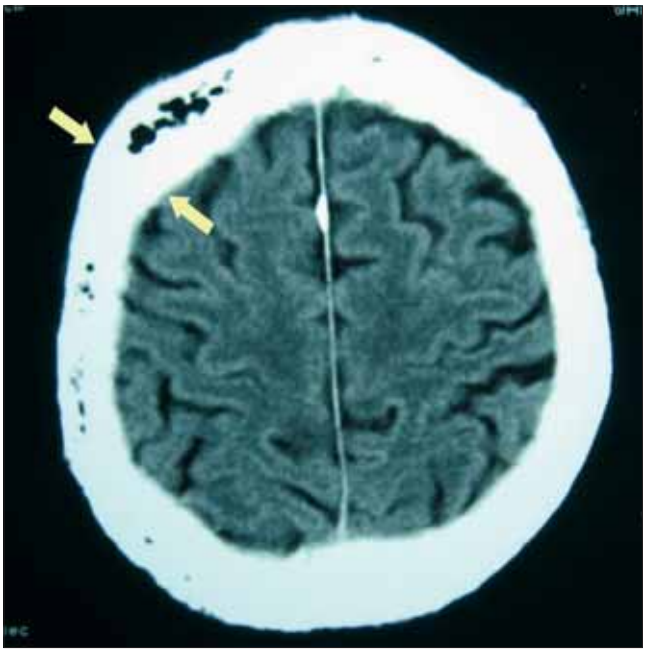

Figura 1. Head CT scan demonstrating focal thickening of the cranial vault.

His father died after progression of symptoms similar to his without final diagnosis. This patient had been requested an otoneurologic evaluation by his neurologist and his orthopedist. At initial examination, oroscopy, rinoscopy, and otoscopy were normal. There was no Romberg sign. There was a right-beating nystagmus present at oblique dislocation of the head. Apendicular coordination was normal. There were no signs suggestive of syringomielia or hydrocephalus.

Head CT scan demonstrated thickening of the skull vault (Figure 1). Electroencephalogram with no evidence of focal activity.

Audiometry demonstrated bilateral mixed dysacusis, 40 dB gap in the right ear and 20 dB in the left ear, with sensorineural hearing decay from 2 kHz in both ear.

Imitanciometry demonstrated normal tympanic membranes and bilateral absence of stapedian reflexes.

Otoneurologic examination disclosed right-sided deficitary peripheral vestibular syndrome.

Auditory brain stem response demonstrated ill-defined waves in right ear; in left ear, the exam disclosed

ill-defined wave I, with prolonged waves III and V latencies. This exam did not rule out retrocochlear disease.

Temporal bones and head MRI disclosed thickened cranial vault with diffuse heterogeneous signal and gross bone trabeculae, suggestive of Paget's disease and signs suggestive of type 1 Chiari malformation (Figure 2).

Seric calcium 9.3 mg/dl (normal range 8.5 - 10.5 mg/dl), urine hydroxiproline 531 UI (normal range 100 UI - 300 UI), seric alkaline phosphatase 2100 UI/l (normal range 110 UI - 360 UI/l).

During the diagnostic workup the patient was on bromazepam by the neurologist, non-steroidal anti-inflammatories (NSAI) by the orthopedist, medications for vertigo, papaverine, quinine and vitamin A for the otoneurologic symptoms, without any improvement, but in the next 5 months, there was worsening of the headache, pain in the legs, widening of the base of support, ataxic gait, and dysphagia.

The diagnosis of Paget's disease evolving to type 1 Chari malformation was made, and the patient was referred back to the neurologist and to an endocrinologist, and conservative treatment was instituted with sodium alendronate, calcium carbonate, amitryptiline, NSAI and analgesics, with improvement of the symptoms.

Discussion

Paget's disease evolving to type 1 Chiari malformation is rarely described in medical literature, and only three case reports have been published so far (13, 14, 15). In the case reported here, we present a 57-year-old male patient who fulfilled the criteria to Paget's disease. In relation to the otoneurological symptoms found in this disorder, mixed hearing loss is commonly seen because the temporal bone is afflicted (8). This patient had, besides mixed hearing loss, tinnitus and vertigo, two other symptoms similarly found in Paget's disease (1,9,10).

Type 1 Chiari malformation occurred because of thickening of the cranial vault due to the bone disorder, and compression of the encephalic structures ensued, forcing the sliding of cerebellopontine structures through the foramen magnum inside the spinal canal. So, besides symptoms frequently seen in type 1 Chiari malformation patients, like neurosensorial dysacusis, vertigo, nystagmus and tinnitus (12, 14, 15), this patient also presented with gait ataxia, dysbasia and dysphagia, the latter due to bulbopontine compression.

In the initial workup, head CT scan already

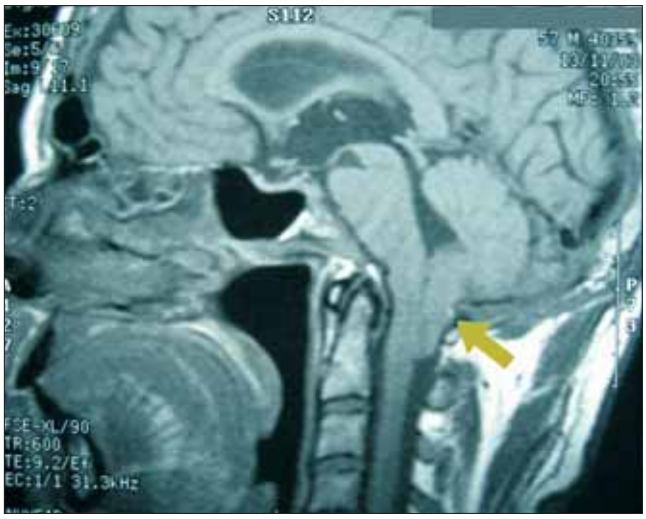

Figura 2. Brain MRI demonstrating sliding of the cerebellar tonsils through foramen magnum, corresponding to Chiari type I malformation.

demonstrated disease of the cranial vault, with important thickening. The audiometric abnormalities demanded further evaluation with more complex imaging procedures. Magnetic resonance imaging (MRI) would be the procedure of choice facing a central vestibular disorder because of brain parenchyma or cochleovestibular system disorders (7), and head MRI was ordered, disclosing vertebrobasilar invagination and signs suggestive of Paget's disease. From this point, bone metabolism markers were ordered, confirming the diagnosis of Paget's disease, with significant raise of seric levels of alkaline phosphatase and urine levels of hydroxiproline. In most Paget's disease cases, seric levels of calcium remain in the normal range (2), as was observed here.

Because of a past personal history of DM and SAH, and in view of Paget's disease with advanced biochemical activity, surgical treatment was not considered, and clinical conservative treatment was the option chosen, with satisfactory control of the otoneurological manifestations and the other symptoms presented by the patient. Conservative treatment was also instituted for other cases presented in literature (13, 14, 15).

Final Remarks

This case illustrates the need to use a differential diagnosis approach in front of a patient presenting with otoneurological manifestations, warning the otolaryngologist and the neurologist to look for less common disorders. With a differential diagnosis in mind, the case can be conducted more satisfactorily towards and adequate and effective treatment, with improvement of the prognosis.

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References

1. Marcos PSM, Montes PJM, Valda RJ, Rubio SMJ: Paget's disease with temporal bone involvement, hypoacusis and vertigo. Acta Otorrinolaringol Esp 1992, 43:232 - 4.

2. Valle LO, Almeida CIR, Alves FRA, Breuel MLF. Perda auditiva progressiva. Em: Campos CAH, Costa HOO (eds). Tratado de Otorrinolaringologia. 1ª ed. São Paulo: Editora Roca; 2003, vol. 2, pp. 348 - 9.

3. Ankrom MA, Shapiro JR: Paget's disease of bone (Osteitis Deformans). JAGS 1998, 46:1025 - 1033.

4. Khetarpal U, Schuknecht HF. In search of pathologic correlates for hearing loss and vertigo in Pagets disease. A clinical and histopathologic study of 26 temporal bones. Ann Otol Rhinol Laryngol 1990, 99(Suppl 145):1-16.

5. Levine SB, Snow Jr JB: Pulsatile tinnitus. Laryngoscope 1987, 97:401 - 6.

6. Melgarejo MPJ, Ruiz GA, Latorre LJF, Palomar GV, Ribera CI: Secondary hearing impairment due to Paget's disease. An Otorrinolaringol Ibero Am 1996, 23:383 - 8.

7. Santa Cruz RS, Munoz HA, Perez PD, Aguirre F, Guillen GV, Figueroa GT: Labyrinth aspects of cranial Paget's disease. Otorrinolaringol Ibero Am 1999, 26:549 - 56.

8. Albers FW, Ingels KJ: Otoneurological manifestations in Chiari-I malformation. J Laryngol Otol. 1993, 107:441 - 3.

9. Bejjani GK: Definition of the adult Chiari malformation: a brief historical overview. Neurosurg Focus 2001, 11:1 - 8.

10. Weissman JL: Hearing loss - state of the art. Radiology 1996, 199:593 - 611.

11. Urban I, Namyslowski G, Morawski K, Wojtacha M: Otoneurologic symptoms associated with Arnold-Chiari syndrome type I. Otolaryngol Pol 2004, 58:281 - 8.

12. Weber PC, Cass SP: Neurotologic manifestations of Chiari 1 malformation. Otolaryngol Head Neck Surg 1993, 109:853 - 60.

13. Iglesias-Osma C, Gomes SJC, Suquia MB, Querol PR, Portugal AJ: Paget's disease of bone and basilar impression associated with an Arnold-Chiari type 1 malformation. An Med Interna 1997, 14:519 - 22.

14. Otsuda F, Inagaki K, Suzuki J, Ogura T, Makino H: Skull Paget's disease developing into Chiari malformation. Endocrine Journal 2004, 3:391 - 2.

15. Richards PS, Bargiota A, Corrall RJM: Paget's disease causing an Arnold-Chiari type 1 malformation. Gerontology 1980, 26:155-9.