



Une Baisse Visuelle Révélant Une Pachyméningite Hypertrophique Idiopathique. Visual Loss Revealing Idiopathic Hypertrophic Pachymeningitis.

C. Mandour, J. Laaguili, M. Gazzaz, B. E L Mostarchid

Department of Neurosurgery - Military Hospital Mohammed V - Rabat - Morocco.

Correspondance: Cherkaoui Mandour ; email : mandour1978@hotmail.com

Abstract: Idiopathic hypertrophic pachymeningitis is a rare fibrosing inflammatory disorder that causes thickening of the dura matter of the base of skull. We report a case of a 27 year-old man presented with this pathological form causing a visual decrease. The diagnosing is a dural matter biopsy and the main treatment is steroid and immunosuppressive therapy. Idiopathic hypertrophic pachymeningitis is a differential diagnosis of optic neuropathy.

Keywords: Hypertrophic; Idiopathic; Pachymeningitis; Visual.

Résumé : La pachyméningite hypertrophique idiopathique est un trouble inflammatoire fibrosant rare qui provoque un épaississement de la dure mère de la base du crâne. Nous rapportons le cas d'un homme de 27 ans qui a présenté cette forme pathologique entraînant une diminution visuelle. Le diagnostic se fait par une biopsie de dure mère et le traitement est principalement des stéroïdes et des immunosuppresseurs. La pachyméningite hypertrophique idiopathique est un diagnostic différentiel de la neuropathie optique.

Mots-clés: hypertrophique; Idiopathique; Pachyméningite; Visuel.

Hypertrophic pachymeningitis is a rare fibrosing inflammatory disorder that causes thickening of the dura matter of the base of skull, tentorium and falx (1). It can be caused by a variety of diseases, including sarcoidosis, tuberculosis and syphilis. In some cases, even after extensive investigation, no specific cause is found and the process is called idiopathic hypertrophic pachymeningitis (IHP) (2). We report a case of this pathological form causing a visual decrease with a literature review.

Case report:

A 27 year-old man was admitted for assessment of a left progressive visual loss. On examination, her best-corrected visual acuity was 20/20 in the right eye but he had no perception of light in the left eye and ophthalmoscopic examination showed obliteration of cup with slightly hyperemic fundi and papillary atrophy. Rest of the neurological examination revealed involvement of the oculomotricity with hypoaesthesia of the hemiface on the left side. Brain Magnetic Resonance Imaging (MRI) (figure: A ; B ; C) revealed a thickened and contrasting dura of the cerebelli left tentorium, temporal region and cavernous sinus, including the left sheath of the optic nerve. All laboratory tests were negative (HIV, syphilis antibodies, tuberculosis, Torch, TB-PCR, tumor markers, complete blood count test, thyroid function, CRP, ESR, RF, ANA, dsDNA, antineutrophil cytoplasmic antibodies (p-ANCA and c-ANCA), Ro (SS-A), La (SS-B), U1RNP, Jo-1, anti-CCP antibody and IgG4). A biopsy of the temporal dura was performed showed chronic inflammatory changes, lymphoplasmacytic cell infiltration and the diagnosis of idiopathic hypertrophic pachymeningitis was suspected.

Discussion:

Hypertrophic pachymeningitis is a rare disorder of diverse etiology (3) characterized by thickening and fibrosis of the dura mater. The first report was made by Charcot and Joffroy in 1869 and later by Naffziger and Stern (4). IHP is diagnosed if no cause can be identified and the exact etiopathogenesis of this entity is still unknown, but it is speculated to be an autoimmune phenomenon or occur as a direct result of infectious or infiltrative pathology (5).

According to the anatomical location and in order of frequency the cases of hypertrophic pachymeningitis can be subdivided into spinal, cranial and craniospinal (6). The entity especially affects older adults, usually after 50 years, with a slight predominance of males (6).

The symptoms are related to the topography of the lesions and rarely are disease specific. The most frequent symptoms are headache, paralysis of cranial nerves and cerebellar dysfunction (7).

In our case, the acute monocular visual loss was the important symptom, which is considered an unusual clinical presentation (1).

MRI is the most useful radiological investigation, the thickened dura appears isointense to hypointense on both T1 and T2W (8). The thickening is better appreciated on coronal and sagittal images (9).

The gold standard for diagnosing IHP is a dural matter biopsy including the following: densely dispersed mixed inflammatory infiltrates, abundant lymphoplasmacytic cells, exuberant fibroplasia, and focal hyaline degeneration (10).

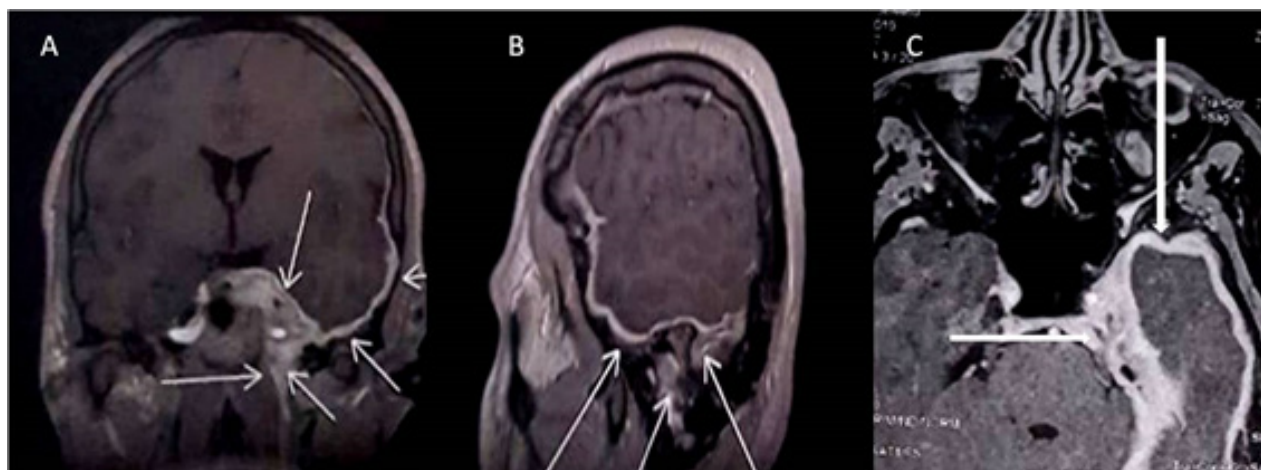


Figure : Brain magnetic resonance imaging (A: Coronal; B: Sagittal; C: Axial) gadolinium enhanced T1-WI revealing thickened and contrast-enhanced dura of the left tentorium cerebelli, temporal area and cavernous sinus, including the left optic nerve (arrows). Imagerie par résonance magnétique cérébrale (A: coronale; B: sagittale; C: axiale) T1 avec injection de gadolinium révélant une dure-mère épaissie avec prise importante de contraste de la tente du cervelet gauche, de la zone temporale et du sinus caverneux, comprenant aussi le nerf optique gauche (flèches).

There is no clear consensus of the treatment of IHP. Although, steroid and immunosuppressive therapy are effective in alleviating symptoms and preventing permanent damage to neural structures.

Conclusion:

IHP is a chronic inflammatory disorder of the dura mater that causes of recurrent cranial neuropathies and headaches; it may be added to the differential diagnosis of optic neuropathy.

Références:

- [1] Rashmi G. Mathew; Kieran M. Hogarth; Andrew Coombes. Idiopathic hypertrophic cranial pachymeningitis presenting as acute painless visual loss. *International Ophthalmology* April 2012, Volume 32, Issue 2, pp 195–197.
- [2] Yuanyuan Huang, Jun Chen, Li Gui. A case of idiopathic hypertrophic pachymeningitis presenting with chronic headache and multiple cranial nerve palsies. *A carry forward. Medicine* 2017; 96:29.
- [3] Hassan K. M., Prabal Deb, and H. S. Bhatoe. Idiopathic hypertrophic cranial pachymeningitis: Three biopsy proven cases including one case with abdominal pseudotumor and review of the literature. *Ann Indian Acad Neurol* 2011; Jul-Sep; 14(3): 189–193.
- [4] Mamelak AN, Kelly WM, Davis RL, Rosenblum ML. Idiopathic hypertrophic cranial pachymeningitis. Report of 3 cases. *J Neurosurg.* 1993; 79:270–6.
- [5] Goyal M, Malik A, Mishra NK, et al. Idiopathic hypertrophic cranial pachymeningitis: Spectrum of disease. *Neuroradiology.* 1997; 39: 619–23.
- [6] Montes Tome A.A, Velasco Casares M, Rizzo Raza S, et al. Idiopathic Hypertrophic Cranial Pachymeningitis associated with an orbital pseudotumor. *EURORAD/CASE.* 2018; ISSN: 1563-4086.
- [7] Karthik S. N, Bhanu K, Sakthi Velayutham, et al. Hypertrophic pachymeningitis. *Ann Indian Acad Neurol.* 2011 Jul-Sep; 14(3): 203–204.

[8] Bang OY, Kim DI, Yoon SR, et al. Idiopathic hypertrophic pachymeningeal lesions: correlation between clinical patterns and neuroimaging characteristics. *Eur Neurol* 1998; 39:49-56.

[9] Friedman DP, Flanders AE. Enhanced MR imaging of hypertrophic pachymeningitis. *AJR* 1997; 169:1425-1428.

[10] Bang JH, Cho KT, Kim EJ. Idiopathic hypertrophic pachymeningitis in the craniocervical junction. *Korean J Spine* 2015; 12:169–72.

Conflits d'intérêts :

Les auteurs ne déclarent aucun conflit d'intérêts.

