Idiopathic Hypertrophic Cranial Pachymeningitis associated with an orbital pseudotumor.

Published on 30.10.2018

DOI: 10.1594/EURORAD/CASE.16205
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain Head and neck
Procedure: eLearning
Procedure: Education
Technique: CT
Technique: MR
Special Focus: Hyperplasia / Hypertrophy Inflammation
Case Type: Clinical Cases
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Patient: 65 years, male

Clinical History:
A 65-year-old woman with a chronic headache was submitted to CT and MRI was then performed. A lumbar puncture was performed. The search for infectious agents by microscopy and culture, immunological tests and antibody test were normal. Meningeal biopsy revealed collagenus fibrosis and a chronic non-specific inflammatory process.

Imaging Findings:
Brain CT (computed tomography) and magnetic resonance (MRI) studies were performed (Fig 1 and 2), finding a diffuse supratentorial meningeal thickening in the retroclival region and in the tentorium of the cerebellum. She was then started on oral corticosteroids and showed good improvement with therapy. During five years radiological controls continued without changes. Until the patient presented ocular pain, in addition to proptosis and discrete restriction to the movement of the right eye. An MRI study was performed in which an injury to the soft tissue was observed in the posterior part of the right orbit, which affects the external rectus muscle including its incersion. (Fig 3) A biopsy of the orbital lesion was performed, being negative for tumor cells and for IgG4. The patient was treated with corticosteroids and immunosuppressants (methotrexate), presenting significant clinical improvement and disappearance of orbital involvement.

Discussion:
Background
Hypertrophic cranial pachymeningitis is a clinical disorder due to localised or diffuse thickening of the dura mater, with or without an associated inflammation, of idiopathic or secondary etiology. The first report was made by Charcot
and Joffroy in 1869, and is attributed to syphilis. Malignancy and infectious or autoimmune disorders such as tuberculosis, bacterial meningitis, neurosyphilis, IgG4-related hypertrophic pachymeningitis and human T-lymphotropic virus type I, rheumatoid arthritis, granulomatosis with polyangiitis, Wegener granulomatosis, and Behçet disease are recognised causes. When a cause is not identified, it is called idiopathic hypertrophic pachymeningitis (IHP). The (IHP) is a fibrosante and infrequent chronic inflammatory process, which causes thickening of the dura, although it also associates leptomeninges by contiguity. According to the anatomical location and in order of frequency, the cases of hypertrophic pachymeningitis can be subdivided into spinal, cranial and craniospinal. The gold standard for diagnosing (IHP) is a dural matter biopsy. The histopathological study shows a thickened dura which, under a microscope, is observed composed of dense fibrous tissue and by cellular infiltration of lymphocytes and, by plasmatic cells and macrophages. [1, 2, 3]

Clinical Perspective; The most frequent symptoms are headache, paralysis of cranial nerves and cerebellar dysfunction, however the clinical presentation is variable and depends on the structures that are compromised (convulsions, blindness). The entity especially affects older adults, usually after 50 years, with a slight predominance of males. A variety of syndromes are associated: Tolosa-Hunt syndrome, diabetes insipidus and pseudotumor oculi. [1, 2, 3, 4] In our case, ocular involvement is associated with secondary orbital Pseudotumor, which is considered an unusual involvement, although in some cases it may be primary involvement with intracranial extension (Fig 3).

Image Perspective:
In CT, a focal or diffuse thickening of the dura can be seen with marked enhancement after the administration of intravenous contrast. However, the test of choice for the characterization of leptomeningeal pathology is undoubtedly MRI. Being hypointense in T1 and T2 weighted images, with important enhancement after the administration of gadolinium contrast. The peripheral increase of the signal in T2 suggests activity of the disease.[1, 3, 4] When associated with inflammatory pseudotumors, they are usually isointense to hypointense in T1-weighted images, with a relatively hypointense T2 signal compared to most other tumors.(Fig 1,2,3)

Treatment: It is based on high-dose corticosteroids, immunosuppressants and decompressive surgery is used in localized forms that affect.[1, 2, 3, 4]

"Written informed patient consent for publication has been obtained"

Differential Diagnosis List: Idiopathic Hypertrophic Cranial Pachymeningitis associated with an orbital pseudotumor., Intracraneal Hypotension, Infection Disease (neurosyphilis), Systemic autoimmune desease and Vasculitides (IgG4-related hypertrophic pachymeningitis), Malignancy (Dural carcinomatosis), Meningioma

Final Diagnosis: Idiopathic Hypertrophic Cranial Pachymeningitis associated with an orbital pseudotumor.

References:
Moura FC1, Pereira IC, Gonçalves AC, Marchiori PE, Monteiro ML. (2005) [Cranial idiopathic hypertrophic pachymeningitis associated with orbital pseudotumor: case report]. Arq Neuropsiquiatr;63(3B):885-8
Description: Axial CT. A marked thickening and enhancement of the interhemispheric fissure is observed. Origin: Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
Description: Magnetic resonance imaging (MRI) Axial gadolinium-enhanced T1WI showing thick meningeal enhancement of the dura mater involving tentorium, falx and basal meninges which enhance intensely on contrast administration. Origin: Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
Description: Magnetic resonance imaging (MRI) coronal gadolinium-enhanced T1WI showing thick meningeal enhancement of the dura mater involving tentorium, falx and basal meninges which enhance intensely on contrast administration. Origin: Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
Description: Magnetic resonance imaging (MRI) Sagital gadolinium-enhanced T1WI showing thick meningeal enhancement of the dura mater involving tentorium, falx and basal meninges which enhance intensely on contrast administration. Origin: Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
**Description:** Axial T1 SATWI sequence. Soft tissue tumor in the right orbit that encompasses the lateral rectus muscle, with marked thickening in all its extension, with significant enhancement after the administration of intravenous contrast. **Origin:** Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.