Special Review

Immune-Mediated Hypertrophic Pachymeningitis and its Mimickers: Magnetic Resonance Imaging Findings

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Hypertrophic pachymeningitis (HP) is a rare and chronic inflammatory disorder presenting as localized or diffuse thickening of the dura mater. It can be idiopathic or an unusual manifestation of immune-mediated, infectious, and neoplastic conditions. Although some cases may remain asymptomatic, HP can lead to progressive headaches, cranial nerve palsies, hydrocephalus, and other neurological complications, which makes its recognition a fundamental step for prompt treatment. Regarding the diagnosis workup, enhanced MRI is the most useful imaging method to evaluate dural thickening. This article addresses the MR imaging patterns of immune-mediated HP, including immunoglobulin G4-related disease, neurosarcoidosis, granulomatosis with polyangiitis, rheumatoid pachymeningitis, and idiopathic HP. The main infectious and neoplastic mimicking entities are also discussed with reference to conventional and advanced MR sequences.

Key Words: Meningitis; Dura mater; Magnetic resonance imaging; Immunoglobulin G4-related disease; Neurosarcoidosis; Granulomatosis with polyangiitis.

Abbreviations: HP Hypertrophic Pachymeningitis, **CT** Computed Tomography, **MRI** Magnetic Resonance Imaging, **WI** Weighted Imaging, **IgG4-RD** Immunoglobulin G4-Related Disease, **NS** Neurosarcoidosis, **GPA** Granulomatosis with Polyangiitis, **RP** Rheumatoid pachymeningitis, **IH** Intracranial Hypotension, **CNS** Central nervous system, **PNS** Peripheral nervous system, **RA** Rheumatoid Arthritis, **CSF** Cerebrospinal fluid, **VDRL** Venereal Disease Research Laboratory, **FTA-ABS** Fluorescent Treponemal Antibody Absorption

INTRODUCTION

Hypertrophic pachymeningitis (HP) consists of a chronic inflammatory process marked by localized or diffuse enlargement of the dura mater. Although some cases remain asymptomatic, HP may lead to chronic and refractory headache, cranial nerve palsies, intracranial hypertension, hydrocephalus, visual field loss, papilledema, cerebellar ataxia, and seizures (1). Therefore, its rapid recognition is a fundamental step towards prompt treatment, in order to restore quality of life and avoid sequelae.

In the context of immune-mediated diseases, HP may be observed as a manifestation of immunoglobulin G4-related disease (IgG4-RD), neurosarcoidosis (NS), and granulomatosis with polyangiitis (GPA). There are also some descriptions of HP associated with rheumatoid arthritis, known as rheumatoid pachymeningitis (RP), and idiopathic cases, when there is no identifiable cause for the dural thickening (1). The present article reviews and illustrates the imaging patterns of immune-mediated HP and presents its main differential diagnoses (intracranial hypotension, infectious diseases and neoplastic conditions) with reference to conventional and advanced MR sequences.

Dura mater anatomy

Dura mater is a thick, rigid membrane formed by fibrous and elastic connective tissue which is the most superficial of the three meningeal layers involving the central nervous system (CNS). It has two parts: the endosteal layer, hardly attached to the calvarium, which has the blood vessels responsible for

Acad Radiol 2023; 30:2696–2706

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https://doi.org/10.1016/j.acra.2023.01.017

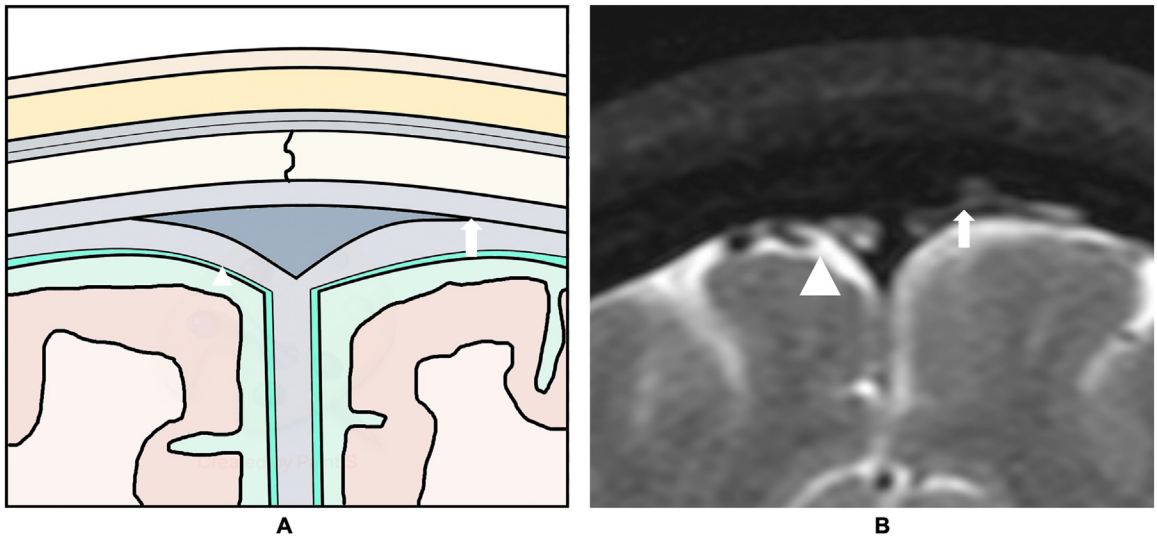

Figure 1. (A) Schematic drawing showing the dura mater layers. The arrowhead points to the meningeal layer and the arrow to the endosteal layer. (B) T2-weighted MRI image showing the same dural layers. (Color version of figure is available online.)

its vascularization; and the meningeal layer, superficial to the arachnoid (Fig 1). These dural parts are fused except in specific locations where they are separated to contain the dural venous sinuses. There are also reflections of dura mater that extend into the intracranial cavity (*diaphragm sellae*, *tentorium cerebelli*, *falx cerebelli,* and *falx cerebri*) (1,2).

IMMUNE-MEDIATED HYPERTROPHIC PACHYMENINGITIS

Immunoglobulin G4-Related Disease

IgG4-related disease is an immune-mediated entity that was first recognized as a distinct disease by Japanese investigators in 2003 (3). It affects men and women of any age, but it seems to be more frequent in men in their fifth and sixth decades (4). The disease causes fibroinflammatory lesions that can lead to organ damage, organ failure, and even the patient's death (4). Although it can occur at almost any organ, there are some predilection organs, including the major salivary and lacrimal glands, orbits, pancreas, biliary tree, lungs, kidneys, aorta, retroperitoneum, thyroid gland, and meninges (5-8).

The most classic findings of biopsy samples are: dense lymphocytic infiltrate, storiform fibrosis, obliterative phlebitis, high IgG4/IgG rate, and an elevated number of IgG4+ cells per high-power field (8-10). The disease tends to be insidious and responsive to glucocorticoids. In this sense, rapidly progressive lesions (significant radiologic worsening within a 4-6 week period) and lack of objective response to corticosteroids are unexpected features and considered as exclusion criteria in the 2019 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) Classification Criteria (8,11).

In addition to HP, infundibulo-hypophysitis and peripheral nerve disease are other conditions of neurologic interest that can be caused by the lymphoplasmacytic infiltrations (9). The prevalence of HP in patients with IgG4-RD is around 2% of the cases. When this presentation is isolated, meningeal biopsy may be required to rule out alternative diagnoses such as infections and neoplastic conditions (9, 12-14). IgG4-RD HP is seen as diffuse linear dural thickening or focal nodular lesions. The distribution varies from the convexities to the base of the skull. Lesions may circumferentially involve the cranial nerves like nerve sheath meningiomas. They usually show intense contrast enhancement, hypointensity on T2 WI, and restricted diffusion (frequent pattern of fibrous tissue) (15-18) (Fig 2**)**.

Neurosarcoidosis

Sarcoidosis is classically described as an idiopathic systemic inflammatory disease histologically characterized by noncaseating granulomas. This condition affects virtually all age groups, however, most cases have been reported in patients aged between 25 and 45 years (19,20). NS symptoms may be present in 5-15% of patients with sarcoidosis. The clinical presentation is heterogeneous and may include cranial nerve palsies (mainly facial and optic neuropathy), chronic headache, nucal rigidity, parenchymal nodular lesions, myelopathy, peripheral neuropathy, and neuroendocrine dysfunction due to hypothalamic/pituitary involvement (21-25).

Leptomeningeal disease is the most common imaging manifestation of CNS sarcoidosis (approximately 40% of cases), typically found in the basilar meninges. Not rarely, contrast enhancement is the only imaging abnormality of cranial nerve involvement. Regarding optic neuropathy, it can be secondary to different mechanisms: subacute optic neuritis, optic perineuritis (thickening and enhancement of the optic nerve sheath), chiasmal damage due to basal leptomeningitis, and compressive optic neuropathy caused by inflammatory

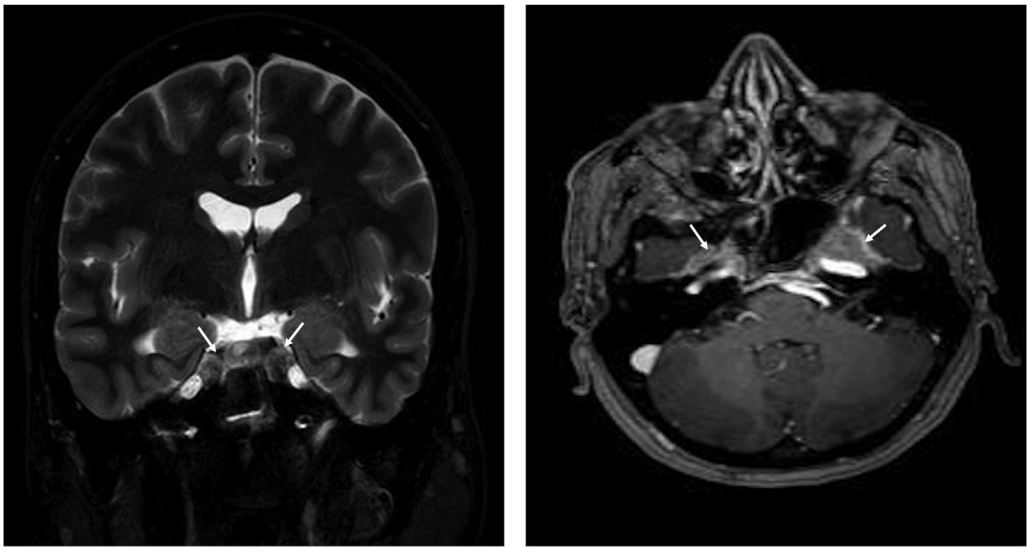

Figure 2. MRI images from a patient with confirmed IgG4-related hypertrophic pachymeningitis. (A) T2-weighted image showing markedly hypointense bilateral lesions on cavernous sinuses and Meckel cava (arrows). (B) Lesions showing subtle contrast enhancement (arrows).

tissue at the ortibal apex. When there is hypothalamic/pituitary involvement, MRI may reveal pituitary stalk thickening and gland enlargement. Loss of the hyperintense signal of the posterior pituitary on T1-WI may be observed in some cases (23,24) (Fig 3).

Pachymeningeal involvement is characterized by focal dural masses or diffuse dural thickening, with hypointensity on T2-WI, irregular/nodular margins, and intense contrast enhancement. Restricted diffusion may be present as in other granulomatous inflammatory processes (e.g., mycobacterial

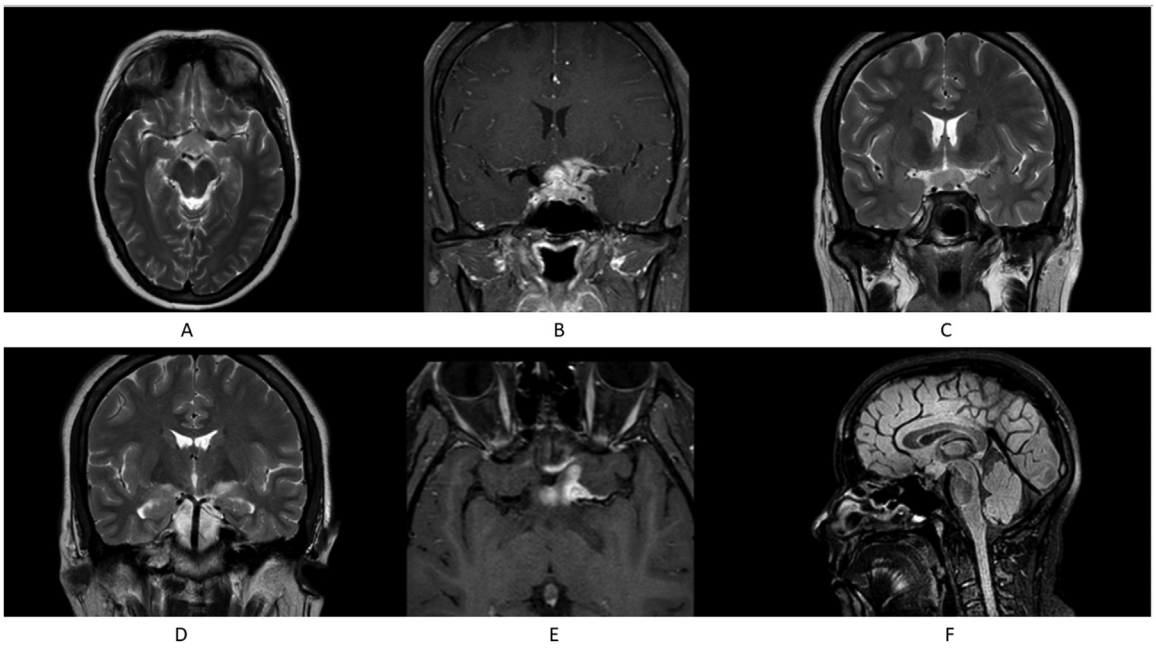

Figure 3. MRI images from a patient with confirmed sarcoidosis. (A) Axial T2 weighted image demonstrating isointense thickening of the optic chiasm and optic tracts. (B) Coronal T1 after contrast:abnormal contrast enhancement and swelling of the pituitary gland and infundibulum, with extension to the left cavernous sinus. There is also leptomeningeal enhancement in the base of the brain, involving the left sylvian fissure. (C) Coronal T2 weighted image showing thickening of the optic chiasm and infundibulum. (D) Coronal T2 weighted image demonstrating parenchymal hyperintensity in the base of the brain. (E) Axial T1 after contrast:abnormal contrast depicting leptomeningeal enhancement in the base of the brain. (F) Sagittal FLAIR demonstrating thickening of the pituitary gland, stalk, and involvement of other structures on the floor of the third ventricle.

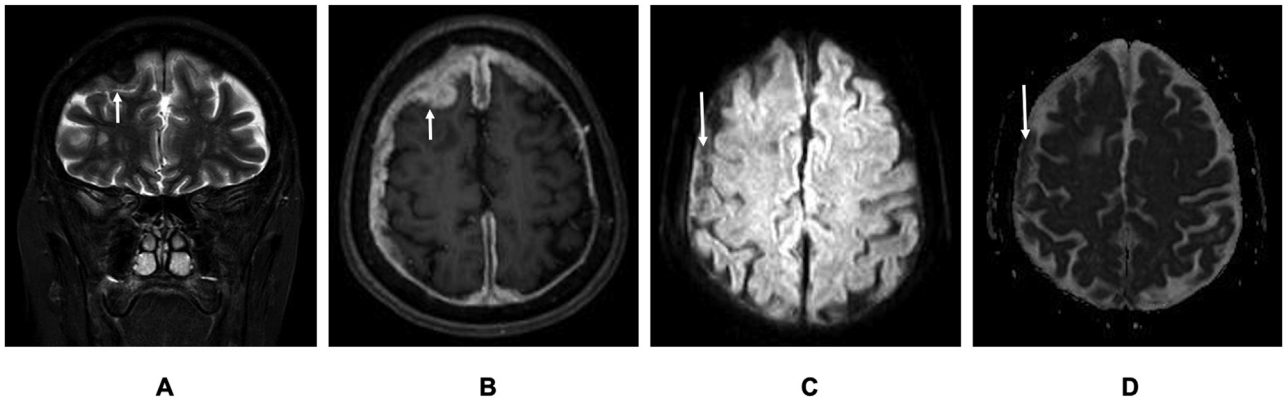

Figure 4. MRI images from a patient with confirmed neurosarcoidosis. (A) T2-weighted coronal image showing an extra-axial low signal lesion (arrow). (B) Axial T1-weighted post-contrast image, showing a diffuse pachymeningeal thickening with strong enhancement (arrow). (C and D) Diffusion-weighted image (DWI) showing high signal in between the pachymeningeal thickening areas, with correspondence low signal in ADC map, compatible with diffusion restriction of water molecules (arrows).

infections) (19,20,26,27) **(**Fig 4**)**. When dural involvement occurs, it is found in distinct areas in relation to the leptomeningeal findings due to the barrier in the outer portion of the arachnoid mater formed by the disease, which prevents its dissemination to the subarachnoid space (20).

Contrast enhancement on T1-WI is a marker of active neurosarcoid lesions and its resolution is an indicator of remission. On the other hand, areas of hyperintensity on T2/FLAIR WI tend to reflect previous but not current inflammatory activity. Recurrence tends to occur in anatomical sites previously affected by the disease, as opposed to demyelinating processes such as multiple sclerosis or neuromyelitis optica spectrum disorder (28,29).

Granulomatosis with polyangiitis

GPA is a rare disease that belongs to the group of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis. It can be localized or manifest as a multiorgan disease (30). Incidence is similar in males and females, and the first clinical manifestations usually occur in individuals aged 45-65 years (30). The disease is characterized by small vessel necrotizing granulomatous vasculitis with few or no immune deposits (30,31,32). The upper airway involvement includes sinonasal lesions with mucosal thickening and osseous erosions; pulmonary involvement may present as parenchymal nodules/cavities and alveolar hemorrhage; kidney disease is marked by pauci-immune glomerulonephritis; and ocular manifestations range from mild conjunctivitis to severe keratitis, scleritis, uveitis, and retinal vasculitis (31,33-35).

Neurological involvement is estimated to occur in 22-54% of patients, with peripheral nervous system (PNS) disease being the most common manifestation. PNS disease may present as polyneuropathy or mononeuritis, which tends to evolve into mononeuritis multiplex due to vasa nervorum inflammation and ischemia (33,36,37). CNS involvement has been reported in three forms: vasculitis of the small vessels of the brain and spinal cord, contiguous invasion of granulomas from extracranial sites, and isolated intracranial lesions (33).

The meningeal disease is mainly characterized by areas of dural thickening (75% of the cases) with two patterns of distribution: diffuse abnormal pachymeningitis unrelated to sinus or orbital disease, and focal dural thickening with enhancement adjacent to sinus or orbital disease (30,31) (Fig 5). Radiologically, the thickened dura with central non-enhanced area (fibrosis) and peripheral nodular enhancement (active inflammation) in the posterior falx and tentorium configures the "Eiffel-by-night" sign (30,31). Isolated leptomeningeal involvement is less commonly observed (25% of the remaining cases) and favors alternative diagnoses (such as infectious and neurosarcoid meningitis). Cerebral vasculitis with white matter lesions showing hyperintensity on T2 WI and vascular distribution (periventricular, subcortical, basal ganglia, mesencephalon, and pons) may also be present (30,31).

Rheumatoid Pachymeningitis

Rheumatoid Arthritis (RA) is a chronic inflammatory disease primarily involving joints, although extra-articular involvement is frequently observed. The disease is more prevalent in women and has an incidence peak between 30 and 55 years. Neurologic clinical findings are more commonly associated with musculoskeletal involvement due to synovitis or articular subluxation compressing peripheral nerves (38).

Direct CNS involvement is uncommon and may be represented by parenchymal and meningeal vasculitis, rheumatoid nodules, and both pachymeningitis and leptomeningitis. The diagnosis of rheumatoid meningitis is supported by a former clinical diagnosis of RA and exclusion of other plausible causes of leptomeningitis and pachymeningits (neoplastic, inflammatory, and infectious conditions). Laboratory analysis of cerebrospinal fluid (CSF) usually reveals elevated protein levels, pleocytosis and reduced glucose level (38,39).

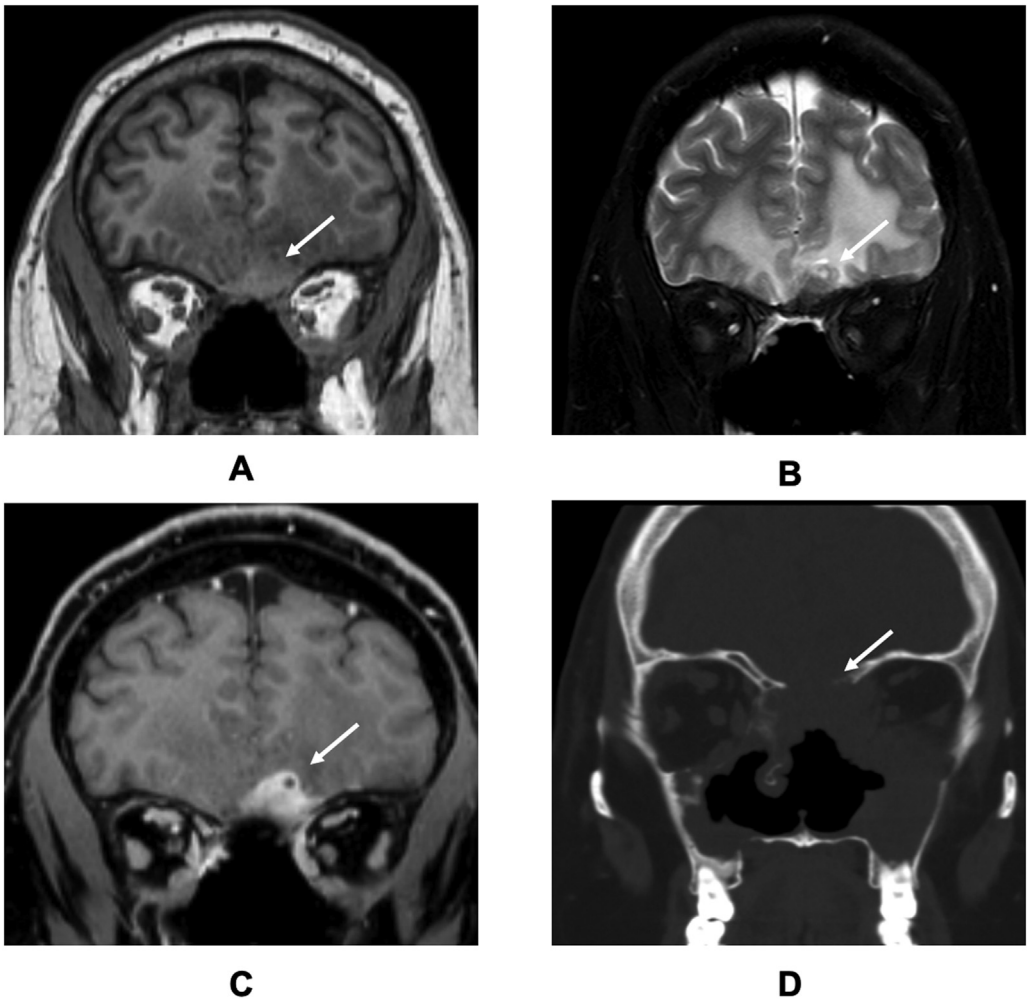

Figure 5. MRI images from a patient with confirmed Granulomatosis with Polyangiitis. (A and B) Coronal images showing expansive extra-axial lesions crossing through the ethmoid bone presenting with isointense signal on T1 weighted image, and hypointense on T2 weighted image (arrows). (C) Coronal T1 weighted images, after contrast, demonstrating intense and irregular enhancement (arrow). (D) CT scan demonstrating adjacent bone erosion (arrow).

Other immune-mediated causes of HP

Rosai-Dorfman disease is histologically characterized by lymph node sinuses dilatation due to histiocytic infiltration, and S-100 immunoreactivity (40,41). Cervical lymph nodes are the most common site of involvement, but extranodal disease is found in up to 43% of the patients (40-42). CNS involvement is rare and is mainly represented by dural-based lesions, which make differential diagnosis with meningiomas (42). Rosai-Dorfman lesions are not associated with calcifications, bony erosion, hyperostosis, or restricted diffusion of water molecules (41). They usually demonstrate hypointensity on T2 WI, while meningiomas vary from low to high signal according to histological type. Contrast enhancement is usually homogeneous (similar to meningiomas) and the most frequent locations are the cerebral convexities, cavernous sinuses, suprasellar, and petroclival regions (40,41) (Fig 6).

There are also some reports in the literature of HP in association with other systemic immune-mediated conditions, including systemic lupus erythematosus, primary Sjögren syndrome, large vessel vasculitis (Takayasu arteritis and giant-cell arteritis), relapsing polychondritis and neuro-Behçet's syndrome (43-50).

Idiopathic Hypertrophic Pachymeningitis

Idiopathic HP is considered an exclusion diagnosis, when no identifiable cause explains the chronic inflammatory tissue proliferation of the spinal and/or cerebral dura mater (51,52). The histopathological findings are non-specific chronic inflammation, with infiltration of lymphocytes, plasmocytes, and some degree of fibrosis (51). Clinical presentation is also nonspecific and associated with long-term headache and cranial nerves involvement (51,53). Some imaging features have been suggested to be more common in idiopathic HP than in secondary HP: lower frequency of anterior and middle fossae involvement and higher frequency of homogeneous enhancement of the dural edge in postcontrast sequences (51) (Fig 7).

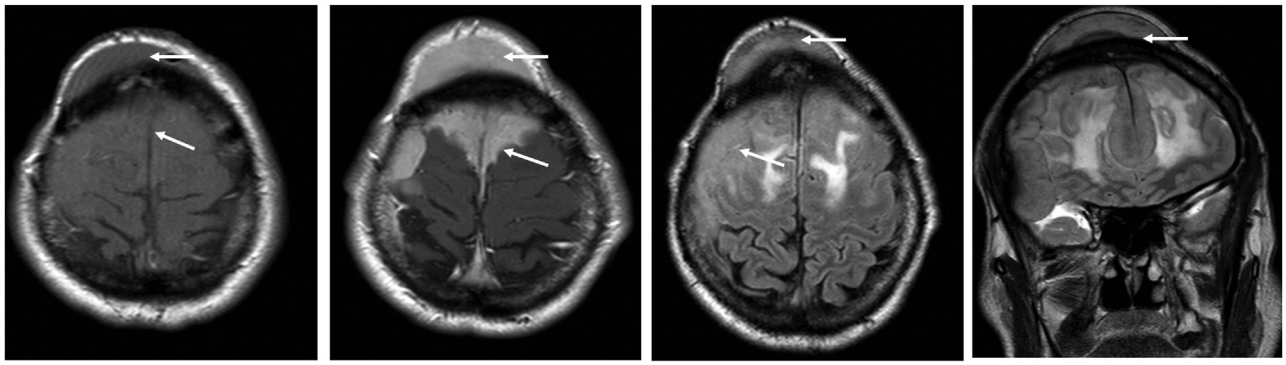

Figure 6. MRI images from a patient with confirmed Rosai-Dorfman Disease. (A and B) Lesion isointense on T1 with strong homogeneous enhancement and dural tail sign (arrows). Iso to hypointense signal on FLAIR axial weighted image (C), and coronal (D) without calcifications or adjacent hyperostosis (arrows).

MIMICKING ENTITIES OF IMMUNE-MEDIATED HP

Intracranial hypotension

IH is a condition associated with low CSF pressure (<60 mm H_2O), frequently due to leakage (54). Spontaneous IH is caused by CSF volume depletion due to degenerative dural dehiscence/tears, while secondary IH is a consequence of dura mater injuries related to cranial/spinal surgery or lumbar puncture (54). The Monro-Kellie doctrine is a principle of intracranial hemodynamics, which states that the combined volume of brain, blood, and CSF is constant. Therefore, any increase or decrease in one of these compartments leads to an opposite change in the others. In the case of CSF hypovolemia, the resulting dural venous engorgement leads to the diffuse dural thickening and enhancement (55).

Neuroimaging findings of IH consist of non-nodular pachymeningeal thickening with hyperintensity on T2 WI (subdural fluid collections), contrast-enhancement on postcontrast sequences, enlarged appearance of the pituitary gland, and sagging of the brain. Reduced mamilopontine distance (<5.5 mm) and cerebellopontine angle inferior to 50° strengthen the qualitative MRI findings (54,56) (Fig 8).

Infectious mimickers of Immune-Mediated HP

Tuberculosis

Tuberculous meningitis is caused by the acid-fast bacillus *Mycobacterium tuberculosis*. The classic dissemination pathway is hematogenic; the tuberculous bacteria reach the central nervous system through blood vessels and establish *foci* in the subpial and subependymal regions of the brain. Later, these *foci* may rupture, spreading the bacteria through the subarachnoid space, which leads to an inflammatory process in the leptomeningeal compartment, in the brain parenchyma (forming tuberculous abscesses/tuberculomas), and, less frequently, in the pachymeningeal compartment (15,57,58). When present, pachymeningeal lesions demonstrate iso or hypointensity on T1 and T2 WI, and intense and homogeneous contrast enhancement. The distribution pattern can be focal or diffuse, presenting with an *en plaque* configuration and associated with vasculitis, cranial nerve palsy, and ischemia (15) (Fig 9).

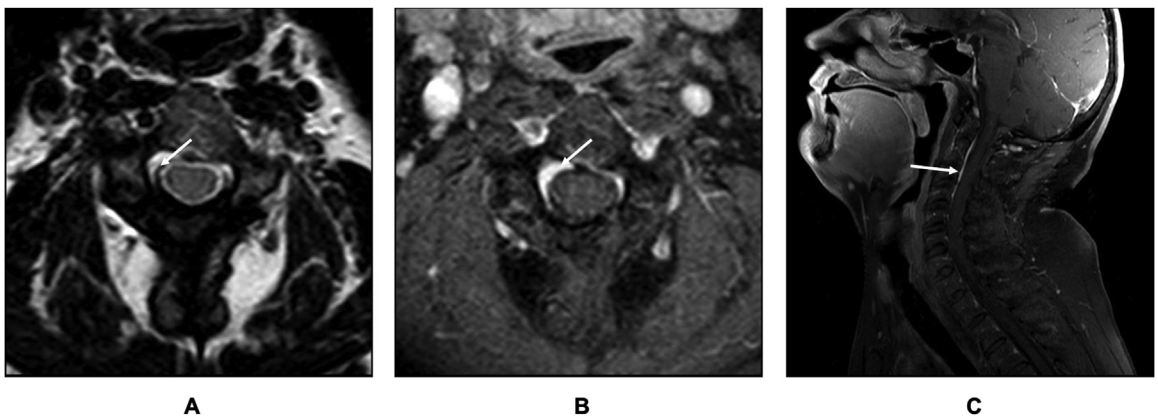

Figure 7. MRI images from a patient with idiopathic hypertrophic pachymeningitis. (A) T2-weighted axial image at C3 level, showing a dural lesion with marked low signal (arrow). (B and C) axial and sagittal T1 post-contrast images showing intense pachymeningeal enhancement (arrows).

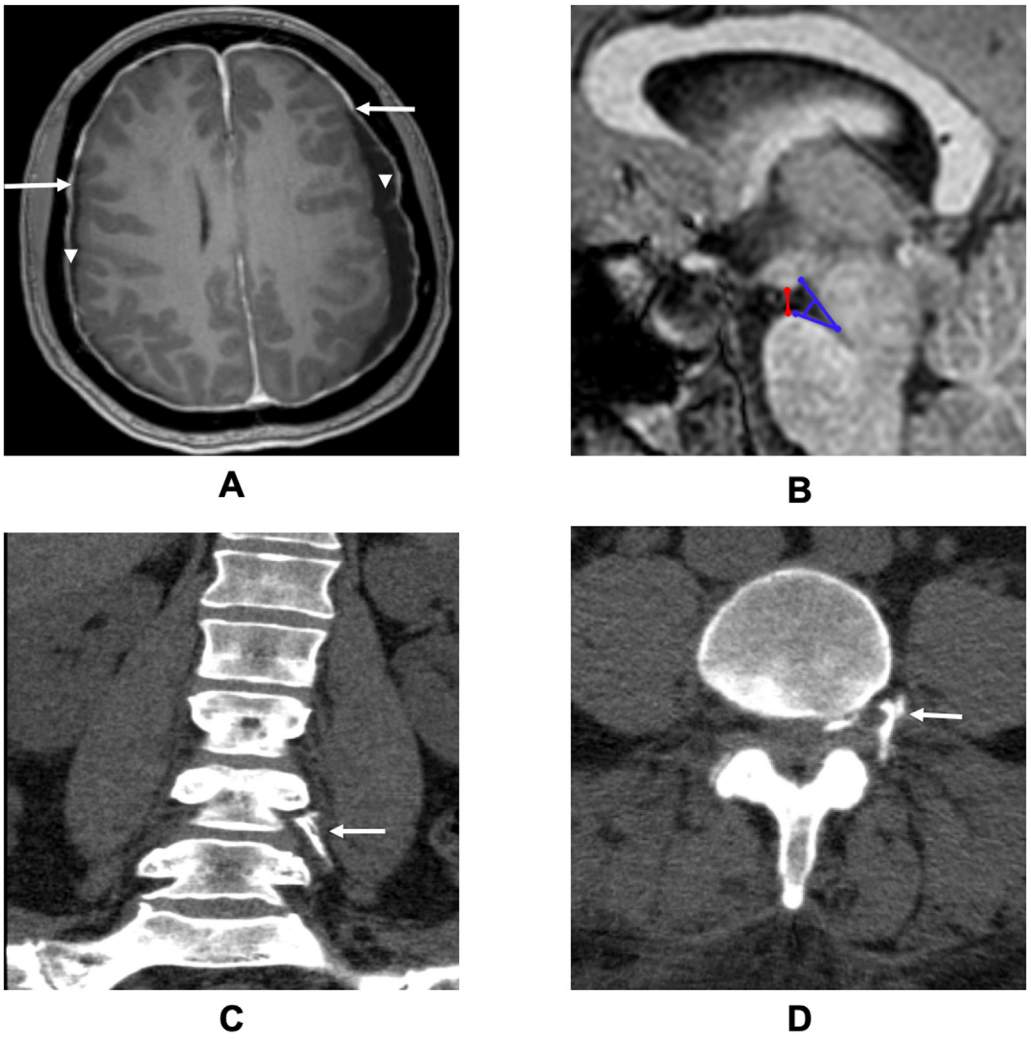

Figure 8. MRI images from a 53 year-old patient with confirmed intracranial hypotension. (A) Axial T1 post-contrast administration showing diffuse regular pachymeningeal enhancement (arrows) and bilateral subdural collections (arrowheads); (B) The red trace indicates the reduced mamillopontine distance estimated in 5 mm (reference > 5,5 mm) and reduced pontomesencephalic angle, estimated in 38° (reference > 55°); (C and D) Computed Tomography myelography scan showing contrast leak related to a dural defect (arrows). (Color version of figure is available online.)

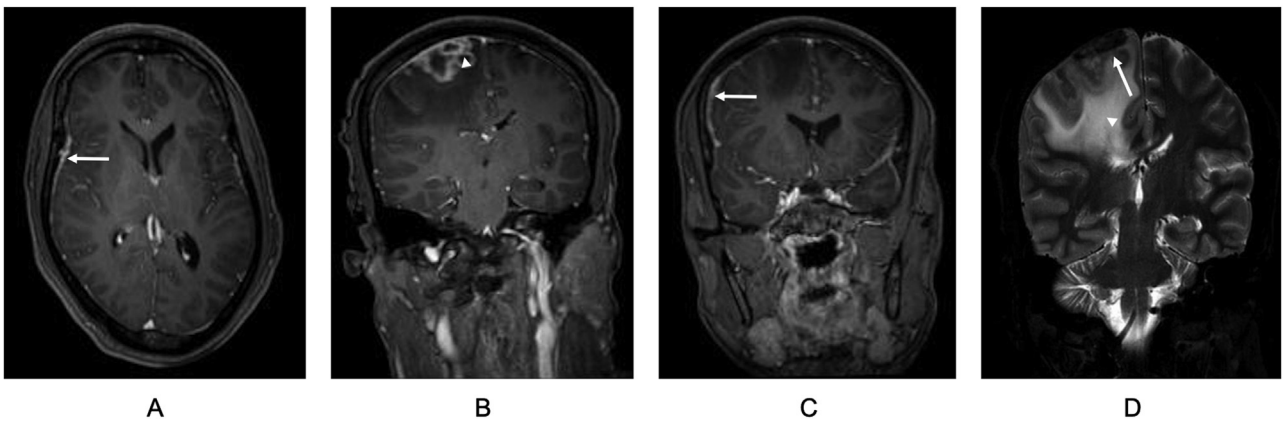

Figure 9. MRI images from a patient with tuberculosis presenting meningeal disease. (A, B and C) T1-weighted post-contrast images in axial (A) and coronal plane (B and C) demonstrating pachymeningeal thickening with intense contrast enhancement (arrows) adjacent to granulomas in the brain parenchyma (arrowhead). (D) T2-weighted image showing hypointense lesions corresponding to granulomas (arrow) and intense perilesional vasogenic edema, greater than expected in meningiomas (arrowhead).

Syphilitic cerebral hypertrophic pachymeningitis

Late (tertiary) syphilis-related HP has rarely been described in the literature. Clinical presentation may be similar to other cases of HP: chronic headache, vertigo, ataxia, and cranial nerve palsies (59,60). The diagnosis of neurosyphilis is based on neurological signs and symptoms and CSF analysis: CSF cell count, protein levels, non-treponemal test (CSF-VDRL) and treponemal test (FTA-ABS). Reactive CSF-VDRL is the gold standard, but due to limited sensitivity, a negative result does not exclude the possibility of neurosyphilis. When there are reactive serological tests (nontreponemal and treponemal) and a high clinical suspicion for neurosyphilis, but a negative CSF-VDRL, the presence of lymphocytic pleocytosis and elevated protein levels in the CSF can be considered in the decision for empirical treatment (61,62).

Paracoccidiodomycosis

Paracoccidioidomycosis is an endemic fungal infection in South America caused by the dimorphic fungus *Paracocciodiodes spp*, which primarily affects the lungs (63-66). The fungus reaches the central nervous system by hematologic or lymphatic dissemination (67,68). Two kinds of lesions may be found in the brain: granulomatous lesions and meningeal disease. Granulomatous lesions are more frequent and present as multiple nodules with supratentorial distribution, while meningeal disease appears as pachymeningeal thickening (63,64,67,68).

The dural involvement is represented by extra-axial lesions that might permeate the adjacent skull, showing peripheral irregular or homogeneous contrast enhancement and low intensity on T2 WI. DWI shows restricted diffusion of water molecules in up to 47% of patients and spectroscopy reveals high lipid and lactate peaks (65,67,69).

Neoplastic mimickers of Immune-Mediated HP

En plaque *meningioma*

En plaque meningiomas are rare primary meningothelial tumors with morphological configuration of flatter carpet-

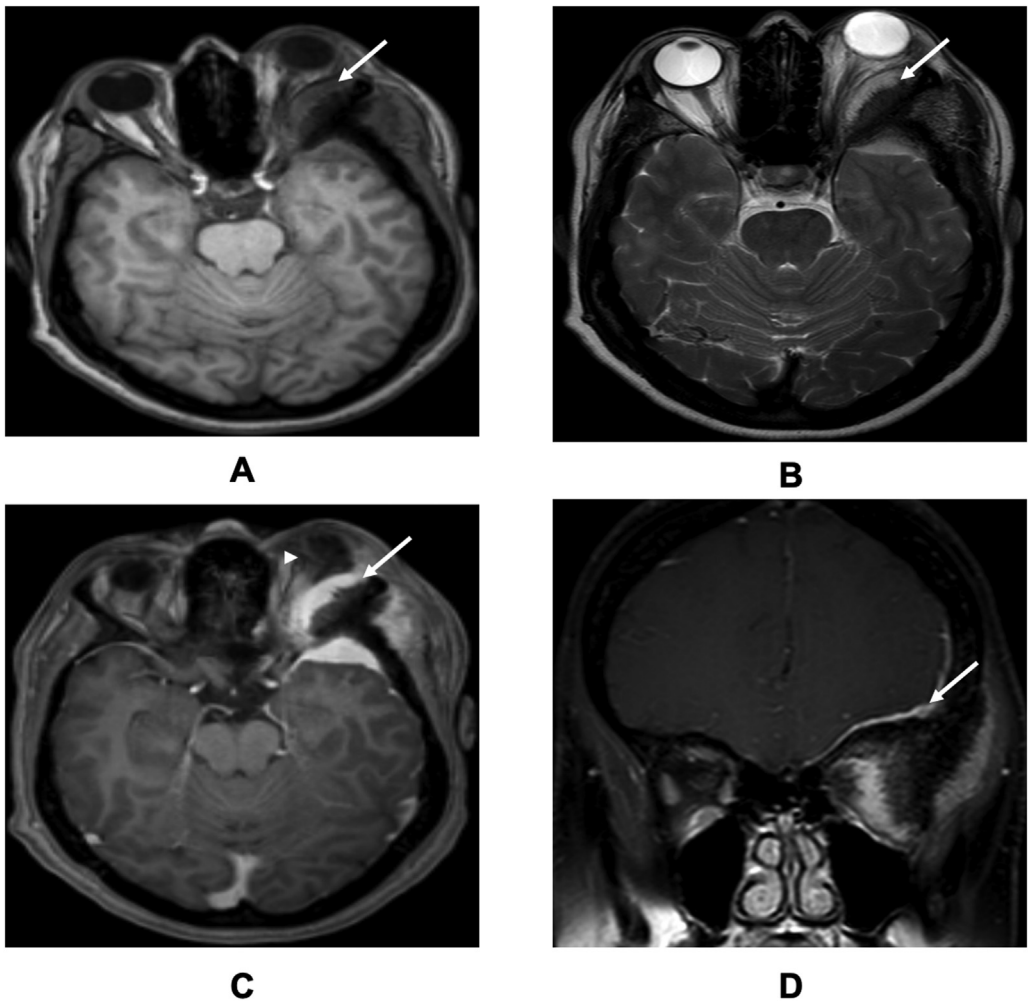

Figure 10. MRI images from a patient with confirmed "en plaque" meningioma. (A and B) Extra-axial left sphenoid wing lesion with isointense signal on T1 and high signal on T2 (arrows). (C and D) axial and coronal T1 post contrast demonstrating intense enhancement (arrow). There is also proptosis of the left eye (arrowhead).

like lesions that infiltrate the dura and may invade the underlying bone (70).These extra-axial lesions are slow growing, and may be focal or extend over large areas of the pachymeninges. The most common locations are the sphenoid ridge and the convexity (70-72). Regarding neuroimaging features, *en plaque* meningiomas present prominent signs of hyperostosis (more than expected for a typical meningioma), with isointensity on T1 WI, hypointensity on T2 WI, and intense contrast enhancement (70-72) (Fig 10).

Pachymeningeal Burkitt Lymphoma

Burkitt Lymphoma consists of a rare non-Hodgkin's highly proliferative neoplasm from undifferentiated lymphocytic cells, which accounts for approximately 50% of all non-Hodgkin's lymphomas in children and 2-3% in immunocompetent adults (73-75). There are also associations with Epstein-Barr virus biomarkers and HIV infection (76). Intracranial involvement occurs mainly due to the dissemination of a primary lesion from other sites, such as the facial bone and abdomen (74). The most frequent presentations are pachymeningeal and cranial nerve infiltrations, with a slightly low signal on T2/FLAIR, and intense enhancement on post-contrast T1-weighted images (73,75).

Primary Dural Lymphoma

Lymphomas originating primarily from meningeal structures are uncommon and usually characterized as low-grade B-cell marginal zone lymphomas (77-80). They are hypercellular lesions with high density on CT, hypointensity on T2 WI, restricted diffusion on DWI, and intense contrast enhancement with a dural tail sign, which may also be found in meningiomas (78). However, more lytic bone areas, more adjacent vasogenic edema, and lower relative cerebral blood volume are expected for primary dural lymphomas when compared to meningiomas (15,77,81) (Fig 11).

Pachymeningeal Metastasis

The most frequent dural neoplastic lesions are originated from breast, prostate, and lung tumors (15,73). Neuroimaging characteristics may vary according to the origin of the tumor,

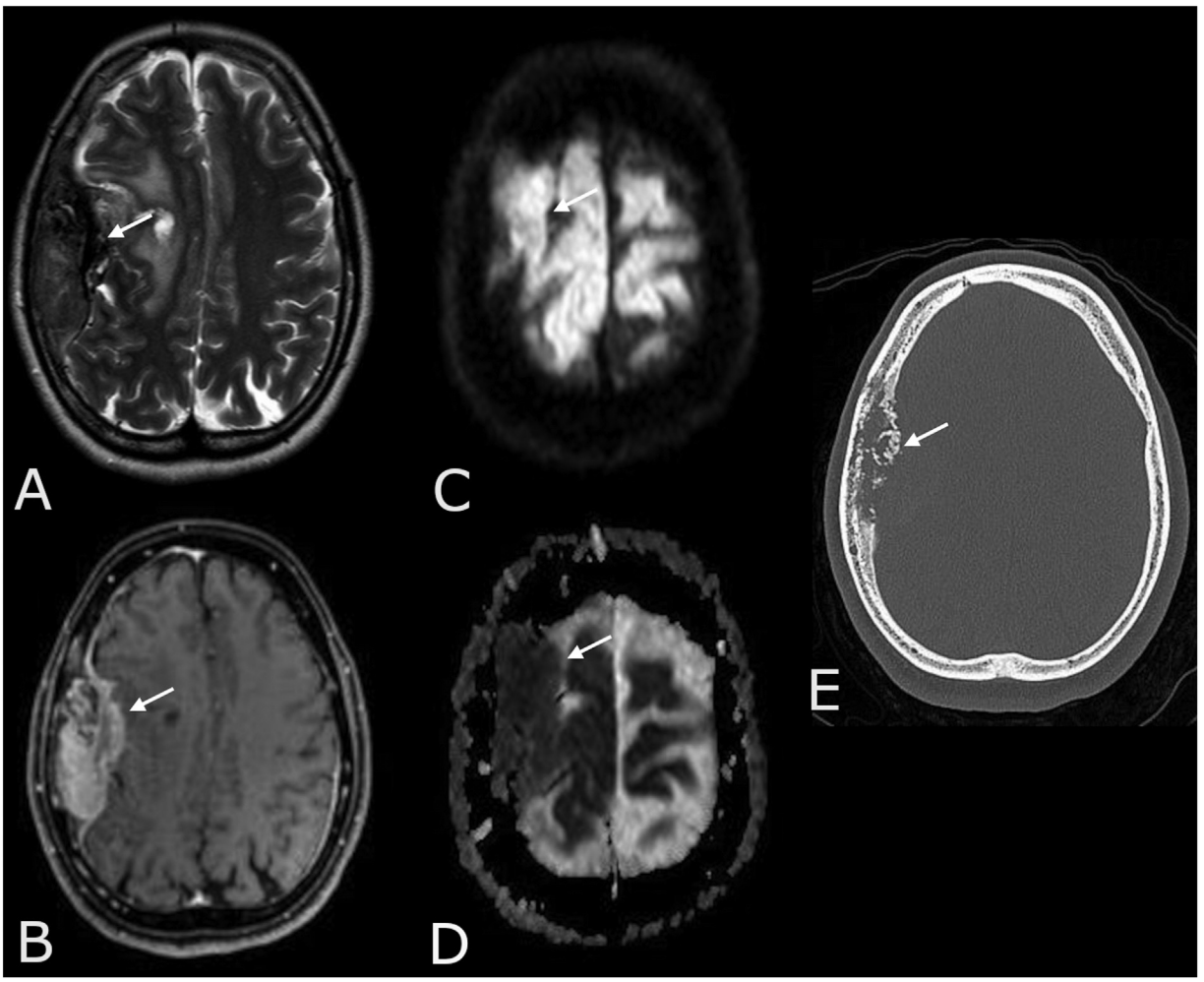

Figure 11. MRI images from a patient with confirmed dural lymphoma. (A and B) An extra-axial dural-based lesion with low signal on T2 weighted images (A), intense contrast enhancement (B) (arrows), and dural tail sign.Diffusion-weighted image shows hyperintensity (C) with low ADC (D), indicating restricted diffusion.The bone lytic adjacent lesions observed on CT (E) are not expected in meningiomas.

although some findings are common to most of them: intense contrast enhancement (associated with neoangiogenesis and lack of blood-brain barrier), dural tail sign, and MR spectroscopy with low NAA/Creatine, high lipid/Creatine ratios and lack of alanine peak (also frequent in meningothelial lesions) (2,15). Other imaging features may resemble the primary tumor, such as the high perfusion in renal carcinomas and metastatic melanoma; the latter may also show a peculiar hyperintensity on T1-WI (15,82,83).

KEY MESSAGES

- HP is a chronic inflammatory disorder presenting as localized or diffuse thickening of the dura mater with contrast enhancement and hypointensity on T2 WI.
- HP can be a manifestation of immune-mediated diseases, such as immunoglobulin G4-related disease, neurosarcoidosis, and granulomatosis with polyangiitis.
- Neuroimaging findings of intracranial hypotension show pachymeningeal thickening and contrast enhancement, and subdural effusions (hyperintense on T2 WI).
- The investigation of HP should also contemplate infectious diseases and neoplastic processes that involve the dura mater.

DECLARATION OF COMPETING INTEREST

None.

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