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Idiopathic hypertrophic cranial pachymeningitis: a rare but treatable cause of headache and facial pain

INTRODUCTION

Idiopathic Hypertrophic Cranial Pachymeningitis (IHCP) is a rare disease with pain and compression related cranial nerve dysfunction as main clinical features. The leading diagnostic finding of IHCP consists of diffuse or localised thickening of the dura, which demands appropriate imaging and image interpretation. This case description aims at increasing the awareness for the clinical symptoms and imaging findings of this rare disease to allow prompt diagnosis and treatment initiation.

CASE DESCRIPTION

An 82-year-old man presented with recurrent left sided headache and worsening facial pain, which had begun more than 1 year ago. Neurological examination at presentation revealed ptosis of the left eye and gaze-induced nystagmus when looking to the left; visual function was intact and no other neurological signs or symptoms were noted. Pre-existing medical conditions included atrial arrhythmia requiring treatment with oral anticoagulants and arterial hypertension. External brain MRI at 1.5T showed diffuse thickening of the dura mater with contrast enhancement over the left frontotemporal region, extending to the left optic nerve and into the orbita. Differential diagnosis included neoplasm (meningeoma en plaque) and inflammatory conditions. Biopsy of the dura was suggested to the patient to obtain diagnostic certainty, but rejected when weighing risks versus benefits. A repeat brain MRI at 3T using an optimised protocol supported the suspicion of IHCP as the most likely aetiology (figure 1A–F) because of the gestalt of dural thickening and the absence of evidence for HCP due to bacterial sinusitis or chronic otitis media. Consequently, the patient was put on oral corticosteroids with prednisolone 75 mg once a day. Thereafter, the patient's clinical symptoms dissolved gradually and a follow-up MRI performed 3 months after initiation of corticosteroid therapy showed a significant reduction of the initially abnormal

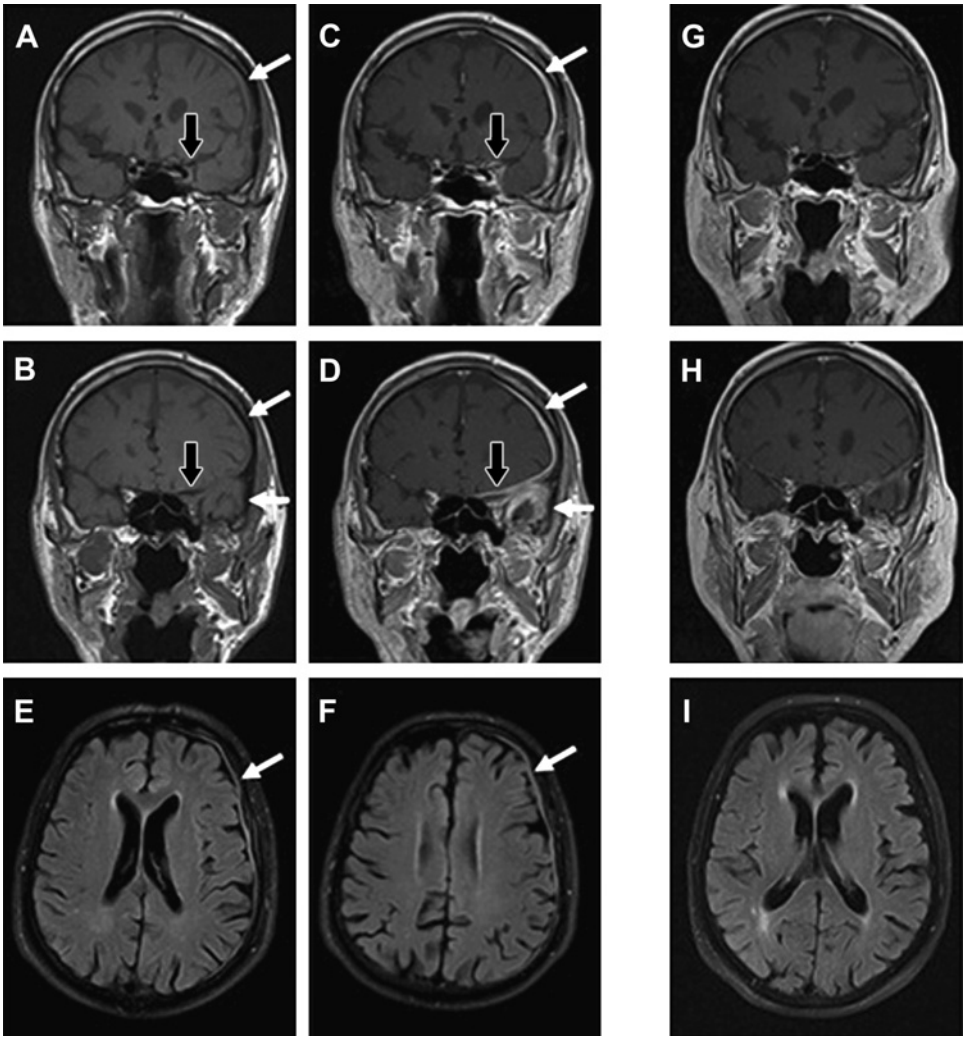

Figure 1 MRI at 3T performed before (A–F) and 8 months after initiation of corticosteroid therapy (G–I). Coronal native T1-weighted scans (A) show hypointensity and bulging of the left cavernous sinus extending into the orbital apex (B, black arrows) and diffuse broadening of the meningeal spaces of the left hemisphere (white arrows). All the hypointense structures are densely enhanced following application of contrast material (C, D) especially in the temporal fossa (D). Diffuse meningeal thickening is seen almost over the entire left hemisphere (E, F). Follow-up MRI shows marked regression of tissue masses and contrast enhancement (G, H) and signal hyperintensity of the meninges over the left hemisphere has disappeared (I).

morphological findings. At that time, the patient was free of complaints and showed no abnormal neurological signs and prednisolone therapy was then tapered to a maintenance dosage of 7.5 mg once a day. A recently performed follow-up MRI performed 8 months after initiation of corticosteroid therapy (figure 1G–I) revealed an almost complete remission of the initially abnormal morphological findings.

COMMENT

IHCP is a rare disease of unknown aetiology, characterised by diffuse or localised thickening of the dura mater and optionally associated with inflammation[1] that recently has gained more attention probably because of the wider application of MRI.[2 3] Clinical symptoms can include headache, facial pain, vision loss, cranial nerve palsy and cerebellar ataxia.[1–3] During diagnostic work-up, IHCP may be suspected from smooth thickening of discrete portions of the dura mater with contrast enhancement.[1 4] Differential diagnosis includes infectious hypertrophic cranial pachymeningitis related to bacterial sphenoid and ethmoid sinusitis and chronic otitis media mostly in immunosuppressed patients, other inflammatory conditions like sarcoidosis and tuberculosis and neoplasms like meningioma en plaque and lymphoma. While thus biopsy is usually performed to confirm the diagnosis, our case illustrates that a characteristic gestalt of MRI findings and their response to treatment may support the diagnosis of IHCP in specific instances, especially when biopsy is denied or deemed too risky. IHCP is primarily treated with corticosteroids.[5] In case of insufficient treatment response, other immunosuppressive agents like methotrexate may be effective, also as add-on therapy to reduce corticosteroid dosage (online material).[5]

►Multiple choice questions to this paper are published online only. To view these files please visit the journal online (http://dx.doi.org/10.1136/jnnp-2012-303295).

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Competing interests None.

Ethics approval All data for this report were obtained during routine diagnostic procedures. The MRI images presented are anonymised, and this report does not contain any information that may identify the patient.

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Notes