Neurological Disorders

Kumar, et al., J Neurol Disord 2015, 3:1
DOI: 10.4172/2329-6895.1000198

Case Report Open Access

Idiopathic Hypertrophic Pachymeningitis – MRI Diagnosis and Follow up

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Rec date: Sep 27, 2014, **Acc date:** Dec 20, 2014, **Pub date:** Dec 22, 2014

Abstract

Idiopathic hypertrophic pachymeningitis is a rare entity, posing challenge to the radiologist for a confident prospective diagnosis. The same is of prime importance as in the absence of a definitive treatment, the clinician has to initiate a presumptive therapy based on radiological diagnosis, histopathology not being the pragmatic option in most cases. We present a case which is interesting not only by virtue of the rarity of this disease but also due to the extent of cranial nerves involved and the drastic response to therapy initiated once suspected on MRI. The emphasize of this report is on being mindful of the possibility and early recognition of signs differentiating it from other close and commoner mimickers.

Keywords: Idiopathic hypertrophic pachymeningitis; Cranial neuropathy; Polycranialis

Introduction

Diseases of cranial nerves are difficult to diagnose and treat. Some of the pathologies presenting clinically with symptomatology, referable to cranial nerves, primarily involve them, while most others cause extraneous compression of these fine structures that traverse through a unique environment of meninges, subarachnoid space, the skull, and its foramina [1]. Idiopathic hypertrophic pachymeningitis (IHP) is one of the rare entities that usually present with cranial nerve involvement in various numbers and combinations. Magnetic resonance imaging plays a pivotal role in diagnosis of IHP [2]. An index of suspicion based on knowledge of this entity enables one to closely scrutinize the potential areas of involvement while imaging the patient. Further an awareness about the imaging appearance leads to an early diagnosis and initiation of a rational management protocol. We present one such case, where both above factors played a key role in enabling us to guide our clinical counterparts towards the diagnosis and a post-therapy confirmation of disease resolution.

Case Report

An 18-year-old female patient presented with 5-month history of progressive headache. Since 2 weeks, drooping of bilateral upper eyelid, drooling of saliva, deviation of the angle of mouth (towards left side), absence of tear production while crying and, diminished taste and smell perception were also noted. She had a gradual but progressive diminution of vision (both eyes). Neurologic examination confirmed a reduction in bilateral visual acuity along with grade 4 right sided facial nerve palsy. Associated bilateral IIIrd, IVth, VIth, VIIIth cranial nerve palsy and left IXth, Xth, XIth and XIIth palsies were also documented. The patient was referred for cranial MR imaging which was performed on a 1.5T superconducting magnet (Magnetom Avanto, Seimens Medical System, Erlangen, Germany) with actively shielded whole body gradients, using a 4 channel bird-cage type qudrature head coil. The imaging revealed lumpy thickening and enhancement of the dura in the region of bilateral cavernous sinus extending along bilateral tentorial edges (Figure 1A and 1B). The lesion further extended along bilateral superior orbital fissure to encase the intracranial and intraorbital optic nerves. An extension along the left mandibular nerve across the foramen ovale was also noted (Figure 1B and 1D). No bone erosion or hyperostosis in the underlying calvaria was evident. No other lesion or invasion of any other structure was noted. Rheumatologic, oncologic, and infectious disease work-ups did not reveal any positive result. The cerebrospinal fluid stains and cultures for fungi and mycobacterium were negative, as was the serology for c-ANCA. Serum angiotensin-converting enzyme (ACE) level was normal and a chest CT performed during the course was unremarkable. 3D-TOF MR angiography, added to the exclusion of the possibility of vasculitis. A diagnosis of idiopathic hypertrophic pachymeningitis was made on the basis of imaging appearance and exclusion of all other common mimicking pathologies. Consent for biopsy could not be obtained because of the patient's reservation against surgical procedure hence a presumptive (yet evidence based) management was started. Oral corticosteroid (three-day course of methyl prednisolone 1gm/day followed by maintenance of oral prednisolone 60 mg/day) was administered under supervision. Drastic recovery of cranial nerve palsy was noted over the next two days after starting the therapy. The patient recovered completely on day 7 hence a repeat MRI was proposed. Contrast enhanced MRI showed significant decrease in thickness of the meninges in the previously affected areas, no new lesions were noted (Figure 1C and 1D). The treatment was stopped after 15 days and a monthly clinical follow-up was done for six months, whereby the patient remained asymptomatic. An imaging done at 8 months did not reveal any residual meningeal thickening at the site.

Discussion

Disorders of multiple cranial nerve alteration can be attributed to various pathological possibilities, because of complex anatomy and close proximity of vascular, dural and osseous structures. Keane (2005) retrospectively studied 979 cases of multiple cranial nerve palsies and found that tumors (30%), vascular disease (12%), trauma (12%), infection (10%), inflammatory processes (5%), and diabetes mellitus (2%) were the most frequent causes [2].

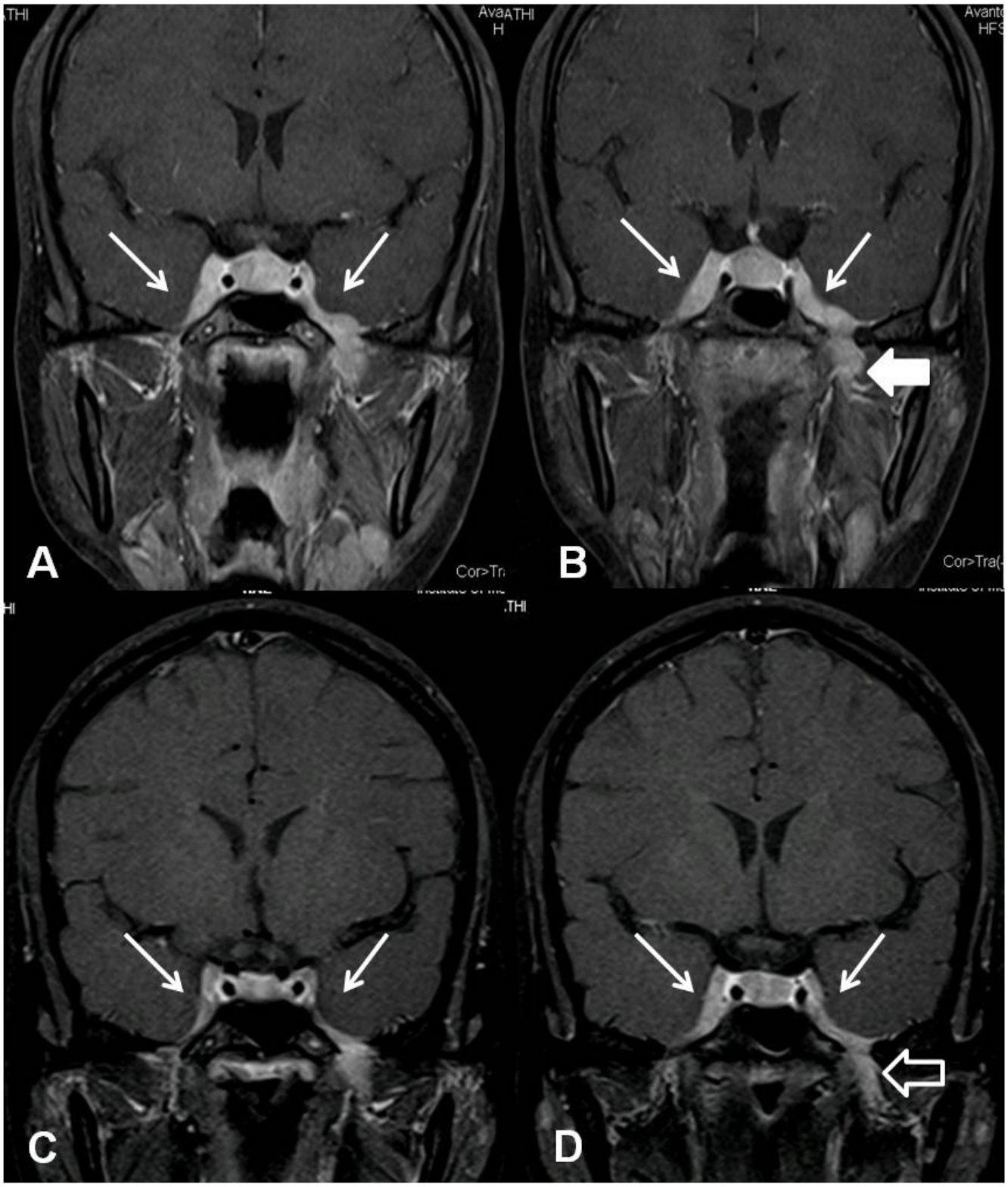

Figure 1: Coronal post contrast fat suppressed T1W images showing meningeal thickening in the region of bilateral cavernous sinus. Pre-treatment images (A,B) showing more lumpy thickening (straight arrows) as compared to the post-treatment images (C,D). Note the meningeal thickening which extends out of the left foramen ovale, along the mandibular nerve (solid arrow in C) and the reduction in bulk of this component as well in post-treatment images (hollow arrow in D).

Idiopathic hypertrophic pachymeningitis, a rare but possibly underdiagnosed condition, often present with multiple craniopathies. It is characterized by pronounced hypertrophy of dura mater with consequent compression of adjacent structures [3]. Exact etiopathogenesis of this entity is still unclear but an autoimmune phenomenon is postulated. Associations with infectious agent, mucopolysaccharidosis, intrathecal toxin, fibrosclerotic disease all have been speculated [4]. The disease can be classified into cranial, spinal and combined craniospinal forms based on the site of involvement [3]. The cranial form of the disease commonly presents with a progressive migraine like headache which is probably caused by dural inflammation as many cases are not associated with raised intracranial pressure [5,6]. However in present study the pattern of headache was non throbbing progressive global headache. Other common presentation of this disease is cranial nerve involvement, either involving cavernous sinus and superior orbital fissures or affecting falx, tentorium and posterior fossa dural linings [7]. Cranial nerve VIII is the most commonly involved cranial nerve, followed by CN V, VII, IX, X, and XII, with equal frequency [4]. However on extensive MEDLINE search we could not find a case of IHP with involvement of all the cranial nerves as was the case in present study. Garcin syndrome is a rare condition affecting the base of the skull that results in impairment of all or nearly all CNs. However, the involvement in the above syndrome is always unilateral [8]. Other clinical presentations of IHP include diabetic insipidus with hypophysitis, cerebellar ataxia, and dural sinus thrombosis [4,9]. The case in present study was associated with compression of the pituitary gland but with no clinical symptoms of diabeted insipidus. IHP has also been associated with Tolosa-Hunt syndrome (cavernous sinus inflammation with subsequent painful ophthalmoplegia), multifocal fibrosclerosis (retroperitoneal fibrosis, Riedel thyroiditis, sclerosing cholangitis, and pseudotumor oculi) [10]. The reported imaging findings of IHP include tumefactive or linear dural thickening, dural sinus thrombosis and associated venous infarct. T1-weighted images typically show a markedly thickened dura which shows strong enhancement on administration of Gadolinium contrast, periphery enhancing more than the center. The lesion shows hypointense fibrotic core on T2-weighted images with hyperintense peripheral active inflammatory front [3]. Such an appearance is in contrast to lymphoma at this site, which shows a totally hypointense lesion due to dense cellularity [3]. Further global enhancement of lesion is seen with no difference between the center and periphery [3]. In the present case, MR imaging findings were typical for IHP, showing linear plaque like thickening of dura mater involving bilateral cavernous sinus, sella, bilateral superior orbital fissure, tentorium cerebella and posterior fossa. It should be noted that such focal thickening of the dura mater is encountered in numerous other diseases such as neurosarcoid, tuberculosis, syphilis, fungal infections, en plaque meningioma, dural metastasis, melanoma, wegener's disease, vasculitis, long-term dialysis patients and continued administration of medicines directly to the cerebrospinal fluid [4,9]. IHP is a diagnosis of exclusion and can only be made if an exhaustive neoplastic, rheumatological and infectious disease work-up fails to procure the diagnosis. Sarcoidosis and tuberculosis are more commonly associated with heterogenous contrast enhancement and T2W intensity pattern with brain parenchymal involvement [3]. Meningioma en plaque shows hypervascular stain both on MRA and conventional angiography in contrast with avascular thickening of IHP [4].

Based on presence of inflammatory signs (fever, increased ESR, leukocytosis, and elevated CRP) IHP has been classified into two groups: group P (with inflammatory signs) carries worse prognosis than group N (without these inflammatory signs) [11]. Our patient belongs to group N and understandably showed robust response with the treatment, undergoing near complete recovery. Corticosteroid therapy has been the mainstay of treatment and has shown effectiveness in alleviating symptoms and in arrest of clinical progression of IHP. The patient usually responds in 2-3 weeks and the therapy may be stopped once an adequate clinical reposnce has been achieved [4-8]. In refractory cases, various immunomodulators such as azathioprine and methotraxate have been used on the assumption that IHP is an autoimmune disease [3,6]. Azathiopine has also been used in steroid dependent patients to taper the corticosteroid dose. A recent study demonstrated the successful use of intraventricular cytarabine in a patient unresponsive to steroids or immunomodulating agents [10,12]. Our case was successfully treated with steroid with complete resolution of clinical symptoms with 8 months follow-up. Radiotherapy remains another treatment option but is usually not preferred due to chances of developing post-therapy progressive multiple cranial nerve palsy [3]. Surgical decompression might be indicated in cases of spinal pachymeningitis. MRI is considered to be of limited value in followup as most reports show insignificant change in the size of the lesions [13]. However in our case, an progressive decrease in thickness of the dural lesion was documented which showed near total resolution in an 8 month follow up. Further a decrease in the peripheral enhancement during the early phase confirms response to therapy [13].

Though a rare condition, a diagnosis of IHP can be made by being mindful of the possibility. MRI signs of IHP are specific and play an important role in differentiating it from other differentials. The modality with intravenous contrast agent contrast should hence be used as a routine while evaluating a case of IHP.

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