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Idiopathic hypertrophic pachymeningitis: A case report and review of the literature

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ABSTRACT

We report the treatment and follow-up, including magnetic resonance imaging (MRI) of a patient with idiopathic hypertrophic pachymeningitis and review the literature published in 21th century, with emphasis on the management and clinical outcomes of this rare disorder. Hypertrophic pachymeningitis is extremely rare. It is a fibrosing inflammatory process which involves the dura mater. Numerous pathological entities produce thickening of the pachymeninges. Thus, idiopathic hypertrophic pachymeningitis is diagnosed by exclusion. We present a case of patient with idiopathic hypertrophic pachymeningitis who had varied clinical presentation. Imaging studies revealed diffuse thickening of the pachymeninges.

Keywords: Pachymeningitis, Hypertrophic, Review**,**

INTRODUCTION

Idiopathic hypertrophic pachymeningitis (IHP) is a chronic progressive diffuse inflammatory fibrosis of the dura mater, leading to its diffuse thickening. This rare disorder is usually found intracranially though spinal forms have also been reported [1]. The first was described by Charcot and Joffroy as a "process in which the neighboring leptomeninges always suffers as well becoming opaque and thick, and further united to the dura and cord". MRI is the examination of choice in preliminary diagnosis of IHP. Histopathological examination of a biopsy specimen of the dura mater would finally confirm the diagnosis [2]. The following presents a patient with IHP and discusses the clinical features, radiological and pathological findings of this rare disorder.

CASE PRESENTATION

A 64 year old man (from Amol, Iran), who had a progressive daily headache since October 2014 for a few minutes in the morning and afternoon every day. His headache was sharp on bifrontal and sometimes pulsatile without nausea and vomiting, photophobia, phonophobia or osmophobia with improvement in recumbent position. He developed bilateral facial numbness since April 2015 and gradually bilateral blurred vision. Speech disorder was added to his symptoms since July 2015 and he was hospitalized for severe exacerbation of his headache on August 2015. At the first clinical examination, he had dysarthria due to left hypoglossal nerve palsy, hyposthesia in bilateral V1 cranial nerves territory and bilateral visual acuity of 7/10. To elucidate these findings, MRI was performed, that showed extensive thickening and enhancement of the dura (Fig. 1).The erythrocyte sedimentation rate (ESR) was 80 mm/h (normal 1-20 mm/h), C-reactive protein (CRP) was 51mg/l but other blood investigations include complete blood count (CBC), renal function test (RFT), liver function test (LFT), serum biochemistry, cerebrospinal fluid (CSF), and studies for ADA, ACE, TB PCR, Aspergillosis PCR, Cryptococcus PCR, wright and cytology (three time) all were normal. CSF protein and CSF WBC number were respectively 80 mg/dl and 81 (PMN 30% & monocyte 70%) with normal CSF sugar and CSF pressure. ANA, ds-DNA, c-ANCA, p-ANCA, serum Ig G4 level,

SSA-Ro, SSB-La, VDRL, ACLA, anti phospholipid antibody, lupus anticoagulant, wright , 2ME were within normal limits or negative. Blood borne virus screen including HIV antibody, HBsAg, HCV antibody all were negative too. PPD test and chest CT scan were normal. After all of these evaluations were done, biopsy of the thickened pachymeninge revealed chronic inflammatory cell infiltration with granulomatous reaction without any evidences of the fungus, acid fast bacilli or malignancy. (Fig 2).

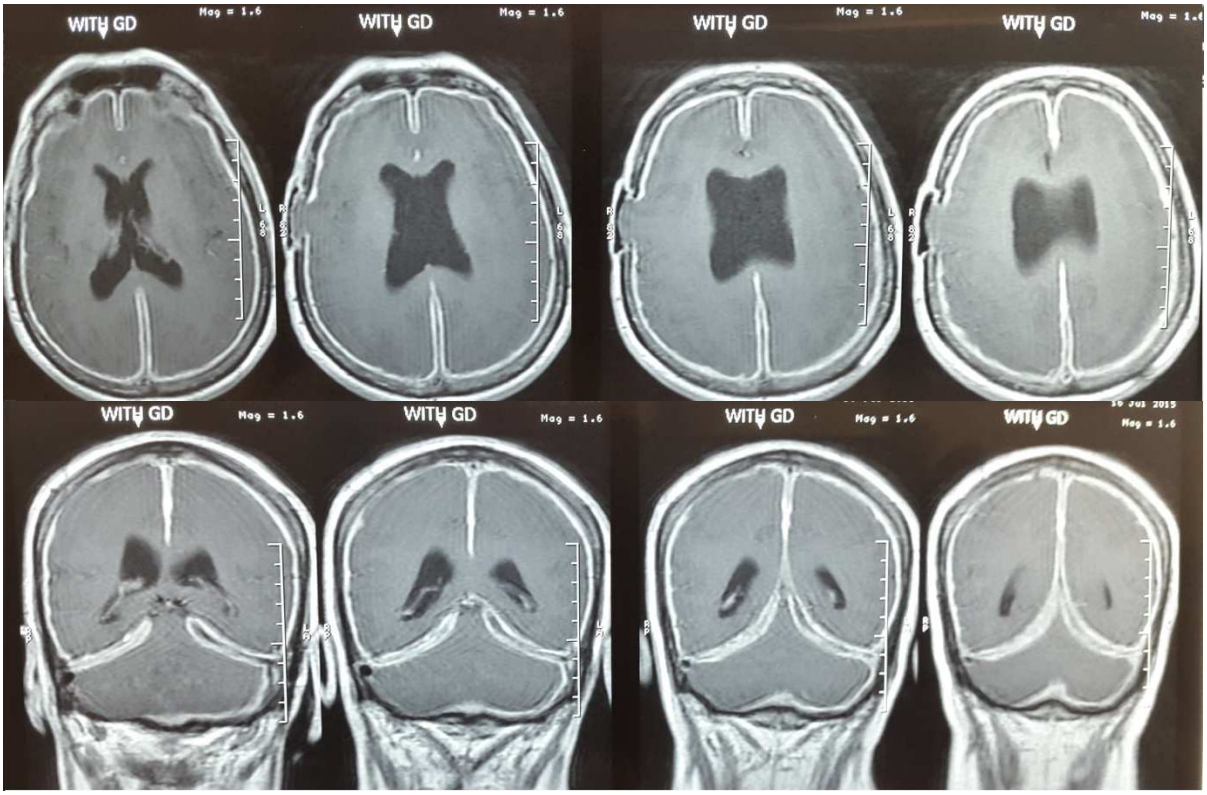

Figure 1: : Brain MRI shows thickening and enhancement of pachymeninge in T1 WI with contrast

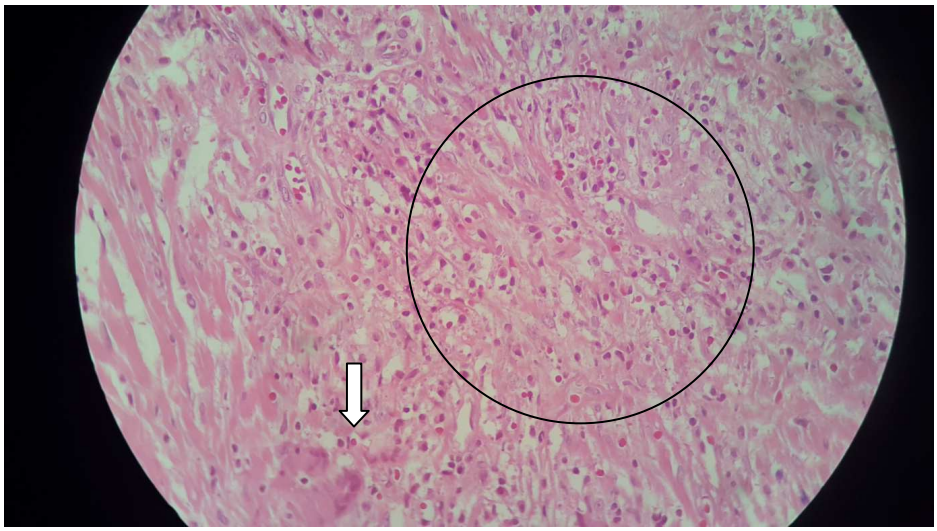

Fig. 2- Histopathological features of a specimen obtained during dural biopsy. A)view showing diffusely thickened, fibrotic dura and marked infiltration by inflammatory cells, with granulomatus reaction (circle) (hematoxylin and eosin; original magnification, ×40). B) higher-magnification view showing lymphocytes, plasma cells, histiocytes and giant PMN (arrow) (hematoxylin and eosin; original magnification, ×100)

After treatment with methylprednisolone 5 gr divided doses in five days, his headache subsided. Treatment continued with 50 mg/day prednisolone. His signs and symptoms except for dysarthria were improved and dural thickening decreased in follow up 4 months later (Fig 3).

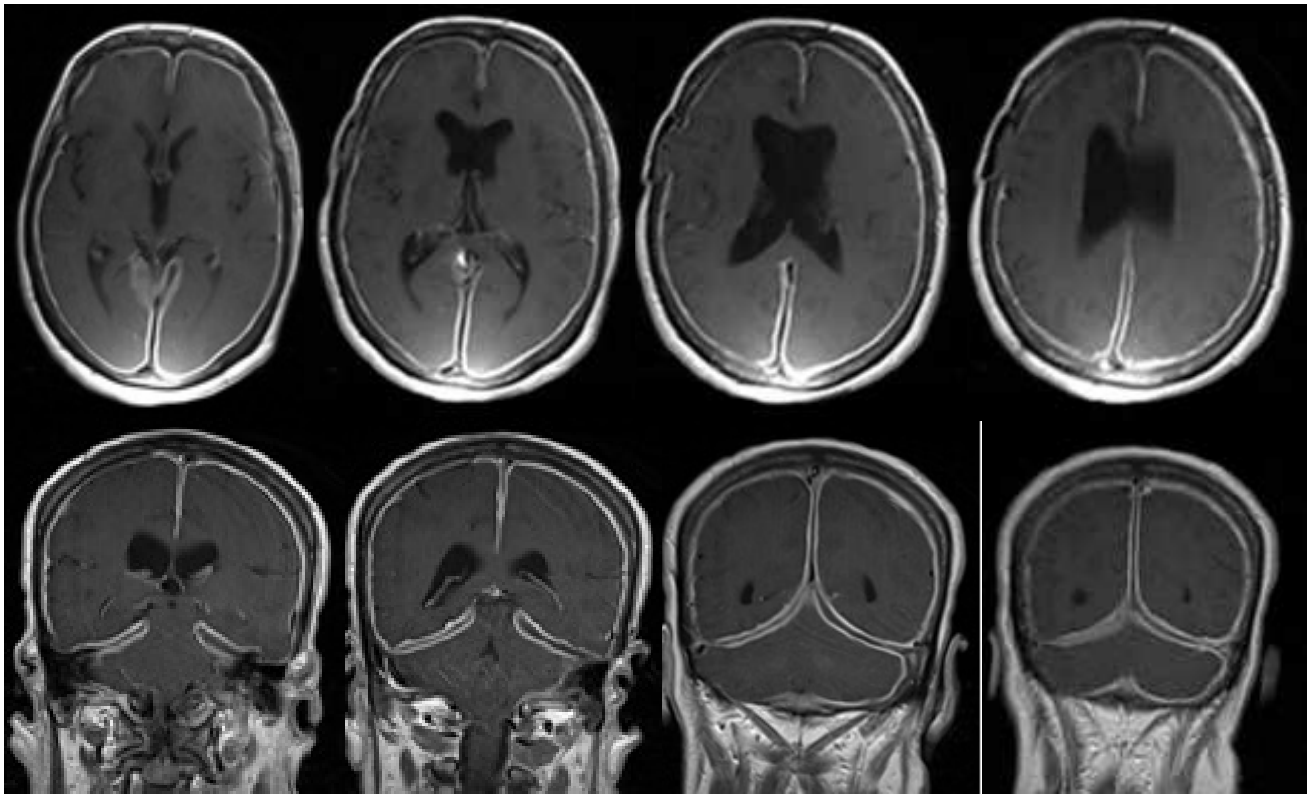

Figure 3- Brain MRI shows a decrease in thickening of pachymeninge in T1 WI with contrast

DISCUSSION

Hypertrophic cranial pachymeningitis is a rare, idiopathic form of granulomatous pachymeningitis. These lesions typically cause progressive cranial nerve palsy, headache and cerebellar dysfunction. It occurs in all age groups but its peak incidence is in the sixth decade of life. Hypertrophic cranial pachymeningitis is best identified by MRI. The diagnosis is established by excluding all other granulomatous and infectious diseases [3]. A dural biopsy is essential to confirm the diagnosis. Pathological findings consist of thick fibrous dura often associated with chronic inflammatory cell infiltration specially lymphocytes and plasma cells [4].

Hypertrophic cranial pachymeningitis is initially responsive to steroid therapy, but in most cases it recurs or progresses despite the treatment. Surgical excision of granulomas is occasionally necessary to alleviate a mass effect [3]. Pachymeningitis is a term used to define the state in which there is localized or diffuse thickening of the dura mater, usually adjacent to an inflammatory focus. The causes of dural thickening include epidural abscess, rheumatoid arthritis, lymphoma, neurosyphilis, sarcoidosis, tuberculosis, and intracranial fibromatosis. IHP may extend to involve the skull base and adjacent tissues. Pathologically, there is diffuse thickening of the dura mater with considerable fibrosis; chronic inflammatory cells including plasma cells and lymphocytes are seen on microscopy. Generally, there is a elevation of the erythrocyte sedimentation rate, and cerebrospinal fluid studies reveal elevated levels of cells and protein [5, 6].

Hypertrophic pachymeningitis (HP) is divided into two types: IHP in which no identifiable cause is found; and secondary HP in which there is an identifiable coexisting cause. Therefore, IHP is a diagnosis of exclusion. In our patient, exhaustive attempts to identify the cause of the dura thickening were unsuccessful.

Clinical manifestations in the majority of patients are similar. The most common symptoms of cranial IHP are headache and cranial nerve palsy [7-9]. IHP may also manifest with chronic headache resembling migraine. Headache, attributed to focal dural inflammation is a universal symptom, and at times may be the only symptom for many years. Intraparenchymal involvement in IHP is rare [10]. The above-mentioned patient presented with headache and multiple cranial nerve involvement.

Dense fibrosis and inflammatory cell infiltration are common histopathological features of IHP [11-13], and in addition to these findings, vasculitis or granulomatous changes have also been reported in several cases [14, 15]. The present case had inflammatory changes in the dura, and as well as epitheloid cells and granulomatous Mikawa et al. subdivided IHP into two groups: first, those with inflammatory signs including fever, increased ESR, leukocytosis, and increased CRP (group P); and second, those without inflammatory signs (group N). They suggested that group P had worse prognosis than group N [16, 17]. Our patient had raised ESR and CRP, so he belongs to group P.

This patient had CSF pleocytosis and increased protein with normal intracranial pressure. Dural inflammation may play a main role in causing headache because in many cases no evidences of raised intracranial pressure are present [18].

MANAGEMENT

The Table 1 summarizes the methods of treatment and outcomes in the patients with hypertrophic pachymeningitis in which MRI or CT documentation were published in 21th century. Most of these patients were idiopathic. Given that management differs for the cranial (ICHP) and spinal (ISHP) forms, we separately address their managements here.

As illustrated in the Table, therapeutic strategies for ICHP include steroids, azathioprine, methotrexate, radiotherapy, surgery and observation. Early treatment is an important point in management of these patients because of its association with improved outcome in terms of neurological recovery [19]. Ventriculoperitoneal shunt and antiepileptic drugs may be needed for symptom therapy. Corticosteroids such as prednisolone could decrease the dural thickening as documented with MRI and resulted in dramatic reduction or even complete remission of symptoms in some patients. Patients may become steroid dependent. Surgical techniques and ventriculoperitoneal shunt have been used with variable success. An empirical treatment with antituberculous drugs may be warranted in selected patients.

Table 1 : Idiopathic hypertrophic pachymeningitis - Response to Treatment

Author [Ref No.]	Age	Sex	Description	Treatment	Outcome
Martin et al [20]	20	F	Intracranial	Corticosteroids, Azathioprine	Improvement at 4 months, asymptomatic at 18 months
	52	F		Corticosteroids, Azathioprine	Progression at 5 years
	58	M		Corticosteroids, Azathioprine	Improvement with diminished tentorial enhancement at 16 months
Terada et al [21]	73	M	Intracranial	Surgery	Improved
Gollogly et al [22]	23	M	Intracranial	Corticosteroids	Asymptomatic at 3 years of follow-up
Lee et al [23]	23	F	Intracranial	Surgery	Improvement, but the disease progression was still uncertain
Riku et al [24]	69	M	Intracranial	Corticosteroids	Improvement at 2 years
Kanzaki et al [25]	70	F	Intracranial	Antibiotics, Surgery	Improved the symptoms
Rossi et al [26]	48	F	Intracranial	Corticosteroids	Psychic symptoms unchanged
	65	F	Intracranial	Corticosteroids	Progressive decreasing at 15 months
	62	M	Intracranial	Corticosteroids	Improved polyneuropathy
	57	M	Intracranial	Corticosteroids	Headache free; seizures free, remitted polyneuropathy at 8 months
Tuncel et al [27]	36	F	Intracranial	Corticosteroids Azathiopyrine Carbamazepine	Improvement of Symptoms at 6 months
Muthukumar et al [28]	60	F	Intracranial	Corticosteroids	Neurologically stable 6 months, but recurrence
Pai et al [29]	47	F	Spinal: T1T6	Laminectomy	spontaneous temporary resolution of symptoms, recurrence after surgery
	68	F	Spinal: C6C7	Corticosteroids, Laminectomy	spontaneous temporary resolution of symptoms, recurrence after surgery
Kesavadas et al [30]	49	F	Intracranial	Corticosteroids	progression in symptoms at 10 years
Bruggemann et al [31]	78	F	Intracranial	Corticosteroids	Resolved aphasia, persistent Headache after 6 months
	76	M	Intracranial	Corticosteroids	Fluctuating and recurrent Symptoms
	83	M	Intracranial	Corticosteroids	Improvement of hemiparesis
	45	M	Intracranial	Cyclophosphamide	Decline of seizure frequency due to antiepileptic drugs
	69	F	Intracranial	Corticosteroids, Cyclophosphamide	Distinct improvement of neurological symptoms
van Toorn et al [32]	10	M	Intracranial	Oral methotrexate	Recurrent after 21 months
Bosman et al [33]	62	M	Intracranial	Oral Corticosteroids oral methotrexate	Improvement within the 6 weeks
Witoonpanich et al [34]	60	F	Intracranial	Steroids, Azathioprine, Cyclophosphamide	Improvement at 7 months
	36	M	Intracranial	Corticosteroids, Azathioprine	Developed central scotoma and optic disc swelling of

					the left eye at 19 months
	19	M	Intracranial	Corticosteroids	Improved, recurred
Lu et al [35]	43	M	Intracranial	Surgery, Corticosteroids	Seizure free
Tao et al [36]	51	F	Intracranial	Corticosteroids	MRI improvement at 2 months
Bhatia et al [37]	23	M	Intracranial	prednisolone and warfarin	Improvement in symptoms at 3 years
	38	M	Intracranial	Corticosteroid	Recurred, asymptomatic within 21 months
Kim et al [38]	37	M	Intracranial	Corticosteroid	Improvement in symptoms
Ito et al [39]	63	F	Catatonia induced	Corticosteroid	Resolving the catatonic symptomatology and EEG abnormality at 1 year
Zhou et al [40]	23	M	Intracranial	Corticosteroid	Improved significantly at 6 months
Takahashi et al [41]	67	M	Spinal: C3C7	Corticosteroid	At the 2-year follow-up, the patient could walk independently
Ranasinghe et al [42]	43	F	Spinal: T1T6	laminectomy, oral Corticosteroids, surgery	Remained neurologically stable and no worsening at 30 months
	77	F	Spinal: T2T3	Laminectomy, Corticosteroids	Dramatic improvement in lower-extremity function and no indication of worsening weakness at 36 months
	65	M	Spinal: T7T8	Laminectomy, Corticosteroids	MRI improvement at 57 months
Hassan et al [43]	24	F	Intracranial	Anti-tubercular, Azathioprine, Corticosteroids	Improvement and no recurrence at 1 year
	56	F	Intracranial	Corticosteroids, Azathioprine	Headache decreased and ophthalmoplegia and MRI improved at 6 months
	24	M	Intracranial	Anti-tubercular, Azathioprine, Corticosteroids	Remained symptom-free over 9 months of follow-up.
Dourado et al [44]	31	M	Intracranial	Prednisone, cyclophos-phamide	Headaches are controlled with prednisone and symptomatic at 7 years
Caldas et al [45]	62	F	Intracranial	Antibiotics	Asymptomatic
Chen et al [46]	49	M	Spinal: T1T3	Laminectomy, Corticosteroids	Neurological function fully recovered within three weeks
Karakasis et al [47]	47	F	Intracranial	--------	Asymptomatic at 8 years
Yasuda et al [48]	28	M	Spinal: T1T4, then L1L3	Corticosteroids laminectomy	Recovered
Chan [49]	50	M	Intracranial	Methylprednisolone	Dramatically decreased the frequency of his pain attacks
	46	M	Intracranial	Oxcarbazepine	Dural enhancement on MRI and his retro-orbital pains were both resolved at 6 months later
Yu et al [50]	40	M	Intracranial	Rehydration	Symptoms improved significantly at 7 days
Lai et al [51]	52	M	Intracranial	Surgery	Seizure stopped and he was smoothly tapered off antiepileptic medication.
Sharma et al [52]	2 year-11month	M	Intracranial	Antitubercular, Corticosteroids	Facial palsy improved completely at 6 months
Sakellariou et al [19]	50	M	Intracranial	Methotrexate, prednisolone	Under follow-up
Senapati et al [53]	50	F	Intracranial	Antitubercular	Walk independently with some spasticity at 15 months
Liu et al [54]	36	F	Intracranial	Surgery	Clinical follow-up at 39 months was unremarkable
Khalil et al [55]	82	M	Intracranial	Prednisolone	Revealed an almost complete remission of the initially abnormal morphological findings at 8 months
Zhu et al [56]	78	M	Intracranial	Corticosteroids	Symptoms improved rapidly
Harshey et al [57]	39	M	Intracranial	Corticosteroids	Improved completely within 3 weeks
Young et al [58]	28	F	Intracranial	Corticosteroids	Remained symptom-free 6 months after biopsy
Lai et al [59]	41	M	Spinal: T2T4	Laminectomy, dura excision	Unavailable
Verma et al [60]	18	F	Intracranial	Corticosteroids	Any residual meningeal thickening at 8 months
The present report	64	M	Intracranial	Corticosteroids	Complete improvement other than dysarthria till writing this report

CONCLUSION

In conclusion, although Idiopathic hypertrophic pachymeningitis (IHP) is a rare disorder, the clinical and radiological findings are characteristic. The most frequently encountered neurological manifestations include headache, cranial nerve palsy and enhanced thickened dura that can easily lead to a diagnosis. The diagnosis of IHP is based on MRI examination and histopathological assessment of dura mater biopsy specimen. The pathologic and laboratory data suggest a close association with granulomatous or connective tissue disease. Therefore, we postulate that IHP might be an autoimmune systemic inflammation, which is localized in the dura mater.

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