Idiopathic hypertrophic cranial pachymenigitis –
A long follow-up needed

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ABSTRACT
Idiopathic hypertrophic cranial pachymenigitis is a rare clinical condition caused by localized or diffuse inflammatory
thickening of dura matter. Described here is a person having diffuse thickening of dura matter of base of skull and he was
on follow-up treatment for 5 years with us. Diagnosis was done by excluding other conditions and with biopsy. The patient
responded to steroid and the MRI picture, which is given serially, shows improvement.

Key words: Idiopathic hypertrophic cranial pachymenigitis, magnetic resonance imaging

Introduction
Idiopathic hypertrophic cranial pachymenigitis is a focal or
diffuse swelling of dura matter of brain or spine. Usually males
are involved. The patient presents as headache, cranial nerve
palsy, cerebellar ataxia, neuro-ophthalmic problems etc. These
signs and symptoms are due to entrapment of cranial nerve
or Cerebro-spinal fluid (CSF) flow interruption.¹² Diagnosis is
based on exclusion of a large number of inflammatory causes
like tubercular, fungal, syphilis, etc. Other conditions like
collagen vascular disease, sarcoidosis, mucopolysaccharidosis,
and neoplastic conditions like lymphomas and meningiomas
are excluded.¹⁰ MRI scan is very helpful and it can depict the
extent and thickening of the dura and its follow-up.¹³

Case Report
The patient was from Medical College and Hospitals. The
patient had presented in 2004 with headache and diplopia.¹⁴ He
also had a H/O of vocal cord paralysis. The study was done on
1.5 tesla GE signa and contrast Gadodiamide (Omniscan) was
used.

His MRI scan had shown thickened dura of base of skull
[Figures 1 and 2]. He was put on full course of antitubercular
medicine. In 2006, the patient came back with not much relief
of his symptoms. MRI did not reveal much change [Figure 3].
His Polymerase chain reaction (PCR) for tuberculosis and
Elisa test for HIV were negative. A CSF study revealed no
acid-fast bacilli or fungal lesions. His adenosine deaminase
level was normal. His rheumatoid arthritis factor was within
normal limit. A test for sarcoidosis was also negative. Serum
angiotensin converting enzyme, C-Reactive Protein (CRP), and
Venereal Disease Research Laboratory (VDRL) tests were also
negative. A biopsy revealed inflammatory areas and the patient
was put on steroid therapy with mark recovery of symptoms.
MRI findings show improvement as a decrease in enhancement
in postcontrast study [Figure 4].

Discussion
Idiopathic hypertrophic cranial pachymenigitis is a chronic
fibrogenic inflammatory disease of dura leading to focal and
diffuse thickening of it.¹⁵,¹⁶ The patient usually present with complains which are usually due to the local factor related
to its thickening. Pressure effects are seen on exiting nerves with
flow disturbance of CSF being noted. The diagnosis is done
first by exclusion of all the common conditions leading to
thickening of dura and it is confirmed by biopsy of the dura.¹⁷,¹⁸
Mark typical enhancement of dura matter is seen in MRI.¹⁴
The enhancement is eveny seen with no gyral pattern, which
is seen in pachymenigitis. As the inflammatory condition
resolves the enhancement also decreases.

In South Asians the common condition of tuberculosis has
to be excluded. The patient should be closely monitored
and long-term evaluation of the patient is needed in this
disease.
Figure 1: T1 weighted postcontrast sagittal images are showing thickened dura matter at the base of skull pointed by the arrowhead.

Figure 2: T1 weighted postcontrast axial images showing thickened thecal sac.

Figure 3: Not much changes are seen in comparison to Figure 1 after full course of antitubercular treatment.

Figure 4: Mark improvement is seen with the lesion not enhancing much and resolving after the patient was put on steroids.

References


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