

A case of idiopathic hypertrophic pachymeningitis presenting with chronic headache and multiple cranial nerve palsies

A case report

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Abstract

Rationale: Idiopathic hypertrophic pachymeningitis (IHP) is a rare condition, characterized by a chronic fibrosing inflammatory process usually involving either the intracranial or spinal dura mater, but rarely both. Here, we report a rare case of IHP affecting both the intracranial and spinal dura mater. We also discussed the diagnosis, management, and outcome of IHP.

Patient concerns: We reviewed the case of a 60-year-old woman presenting with chronic headache, multiple cranial nerve palsies and gait disturbance. Magnetic resonance imaging (MRI) of her head revealed thickened and contrast-enhanced dura in the craniocervical region as well as obstructive hydrocephalus and cerebellar tonsillar herniation. The patient had a suboccipital craniectomy and posterior decompression through C1 plus a total laminectomy. The dura was partially resected to the extent of the bony decompression, and a duroplasty was performed.

Diagnoses: Microscopic examination of the surgically resected sample showed chronic inflammatory changes, lymphoplasmacytic cell infiltration, fibrous tissue hyperplasia, and hyaline degeneration. Blood tests to evaluate the secondary causes of hypertrophic pachymeningitis (HP) were unremarkable.

Interventions: Steroid was used to treat suspected IHP.

Outcomes: Postoperatively, the patient showed gradual improvement in her headache, glossolalia, and bucking. Prior to discharge, a follow-up MRI showed improvement of the dura mater thickening.

Lessons: IHP is a chronic inflammatory disorder of the dura mater that usually causes neurological deficits. Clinical manifestations of IHP, MRI findings, and laboratory abnormalities are the essential components for making an accurate diagnosis. When the radiological or laboratory evaluation is uncertain, but neurological deficits are present, a prompt surgical approach should be considered. Postoperative steroid therapy and close observation for recurrence are necessary to ensure a good long-term outcome.

Abbreviations: CN = cranial nerve, HP = hypertrophic pachymeningitis, IHP = idiopathic hypertrophic pachymeningitis, MRI = magnetic resonance imaging.

Keywords: chronic headache, idiopathic hypertrophic pachymeningitis, multiple cranial nerve palsies

1. Introduction

Idiopathic hypertrophic pachymeningitis (IHP) is a rare disease caused by thickening of either the intracranial or spinal dura

mater, or rarely both simultaneously. Chronic headache, multiple cranial nerve (CN) palsies, and cervicodynia are the most common clinical manifestations.^[1,2] Here, we report a rare case of IHP involving both the cranial and spinal dura mater.

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2. Case report

In April 2014, a 60-year-old woman with persistent headache for 3 months was admitted to hospital for dizziness, lumbago, stomach upset, and insomnia. Her neurological examination revealed left 12th CN palsy. Brain magnetic resonance imaging (MRI) results were normal. She was misdiagnosed as lacunar infarction and paralysis of hypoglossal nerve after she had a normal brain MRI during the first visit. Subsequently, she was diagnosed with tension-type headache plus anxiety.

In February 2016, the patient was re-admitted again after a month long history of glossolalia, dysdipsia, cervicodynia, and gait disturbance. Her neurological examination demonstrated 9th, 10th, and left 12th CN palsies. MRI of her head revealed thickened and contrast-enhanced dura of the left tentorium cerebelli, temporal area, pons varolii, medulla oblongata, and cervical segments of the spinal cord (Fig. 1A and B), as well as obstructive hydrocephalus and cerebellar tonsillar herniation (Fig. 1C).

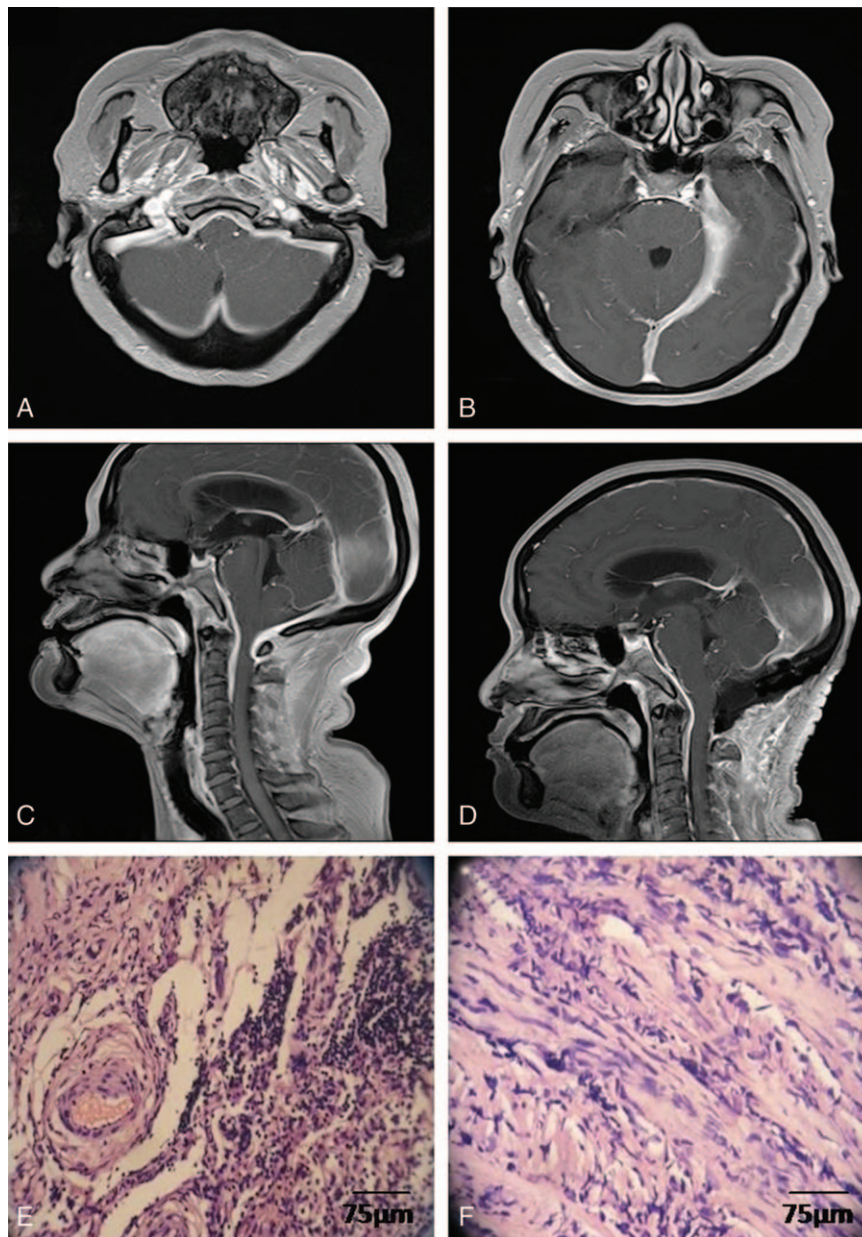


Figure 1. (A–C) Magnetic resonance imaging (MRI) of head revealed thickened and contrast-enhanced dura of the left tentorium cerebelli, temporal area, pons varolii, medulla oblongata, and cervical segments, as well as obstructive hydrocephalus and cerebellar tonsillar herniation. (D) After surgical resection and decompression, contrast-enhanced MRI showed improvement of the degree of enhancement. (E, F) Microscopic examination of the surgically resected material showed chronic inflammatory changes, lymphoplasmacytic cell infiltration, fibrous tissue hyperplasia, and hyaline degeneration.

The patient had normal laboratory tests for HIV, syphilis antibodies, tuberculosis, Torch, TB-PCR, tumor markers, complete blood count test, thyroid function, liver, and renal function. Her CRP, ESR, RF, ANA, dsDNA, antineutrophil cytoplasmic antibodies (p-ANCA and c-ANCA), Ro (SS-A), La (SS-B), U1RNP, Jo-1, and anti-CCP antibody were all negative. Her serum IgG4 level was within the normal range. Lung CT was normal. She had no fever, lethargy, weight loss, or vomiting. A history of contact with known cases of tuberculosis was absent. Because of the tonsillar herniation, lumbar puncture was not done. Considering the history and findings from these investigations, “idiopathic hypertrophic pachymeningitis” was suspected.

The patient had a suboccipital craniectomy and posterior decompression through C1 and a total laminectomy. Intraoperatively, the dura was found to be diffusely thickened and compressing on the spinal cord beneath. The dura was partially resected to the extent of the bony decompression, and a duroplasty was performed.

Microscopic examination of the surgically resected specimen showed chronic inflammatory changes, lymphoplasmacytic cell infiltration, fibrous tissue hyperplasia, and hyaline degeneration (Fig. 1 E and F). Blood tests to evaluate the secondary causes of hypertrophic pachymeningitis (HP) were unremarkable. Steroid treatment was started because of the microscopic findings.

Postoperatively and prior to discharge, a follow-up MRI showed an improvement in the dura mater thickening (Fig. 1D). Four weeks after discharge, the patient showed a gradual improvement in her headache, glossolalia, bucking, and gait disturbance. She had significant improvement at her 1-year-follow-up. Symptoms of glossolalia and dysdipsia disappeared, and her 9th, 10th, and left 12th CN palsies recovered. Chronic headache dissipated. Her gait abnormality resolved and she could walk independently at the 1-year-follow-up. The 1-year-follow-up MRI showed no new lesions comparing to the MRI of her brain 1 year ago. This is a clinical case report that does not involve clinical studies, drug trials, etc. and is not approved by the ethics committee. The patient provided written informed consent.

3. Discussion

HP is a rare chronic fibrosing inflammatory process that involves the cerebral and/or spinal dura mater. Causes include vascular abnormality, trauma, infection, neoplasm, and neuro-inflammatory conditions.^[2] IHP is diagnosed if no cause can be identified. IHP usually occurs in male adults presenting with chronic headache and CN palsies.^[1] IHP headache has no specific location or characteristic, but it can progressively become worse, and alleviated by NSAID. When the dura is not obviously thickened on plain MRI or CT scans, IHP can be misdiagnosed as primary headache of unknown etiology.

In our case, the patient was a 60-year-old woman misdiagnosed as having a tension-type headache. The diffuse dural inflammation and the associated mild increase in intracranial pressure were the causes of her headaches. The compression due to dural hypertrophy can lead to damage of the CNs close to the dura. In fact, damage of all 12 pairs of CNs has been reported.^[3] The involvement of CN IX and XII is relatively rare.^[3] However, our patient had CN IX, X, and XII palsies.

Contrast-enhanced MRI is vital when investigating IHP.^[2,4] Diffuse thickening of the dura was present in our patient (Fig. 1A–C). Our case was unique because it involved the junction of the craniocervical area, and her IHP caused obstructive hydrocephalus along with cerebellar tonsillar herniation. After surgical resection and decompression, contrast-enhanced MRI showed improvement of enhancement (Fig. 1D).

The gold standard for diagnosing IHP is a dural matter biopsy. Microscopic findings of IHP include the following: densely dispersed mixed inflammatory infiltrates, abundant lymphoplasmacytic cells, exuberant fibroplasia, and focal hyaline degenera-

tion.^[1,3] Dural matter biopsy should be considered if the symptoms are progressing. In our case, a biopsy was performed after decompression of obstructive hydrocephalus since malignancy was originally suspected.

The treatment of IHP remains controversial. Although there is no clear consensus on the dose and duration of steroid therapy, steroid therapy is effective in alleviating symptoms of IHP. Immunosuppressive therapy may be needed and, in refractory cases, surgical treatment is required. Our patient received surgical decompression first and then pulse therapy of a high-dose glucocorticoid. Managing the recurrence or relapse of IHP is challenging. The time interval to develop a relapse can range from 1 week to several years after the initial treatment.^[5] The symptoms of our patient were fortunately relieved after 10 weeks of treatment.

4. Conclusion

IHP is a chronic inflammatory disorder of the dura mater that causes neurologic deficits. Clinical indications of IHP, MRI findings, and laboratory evaluation are essential for making an accurate diagnosis. When radiological or laboratory evaluation is uncertain, or neurological deficits are present, a prompt surgical approach should be considered. Postoperative steroid therapy and close observation for recurrence are necessary to ensure a good long-term outcome.

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