Rapid bilateral visual loss as the initial clinical manifestation in idiopathic hypertrophic cranial pachymeningitis

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Abstract
A 59-year-old patient presented with 4-day acute painless bilateral visual loss, MRI results showed dura enhancement of the frontal, anterior cranial fossa. The patient was considered to have idiopathic hypertrophic cranial pachymeningitis based on laboratory tests and MRI data. After treatment with hormones, the visual acuity obviously improved.

KEYWORDS
dural thickening, idiopathic hypertrophic cranial pachymeningitis, optic nerve injury, visual loss

1 | INTRODUCTION

Idiopathic hypertrophic cranial pachymeningitis (IHCP) is a rare fibrose inflammatory disorder characterized by thickening of the dura matter at the base of the skull, tentorium, and falx. The clinical manifestations include chronic headache, cranial nerves affected, and epilepsy, a few patients present with bilateral or unilateral moderate visual loss. We describe the case of a patient who presented with bilateral visual loss without other positive manifestations; the patient was initially diagnosed with ischemic optic neuropathy (ION), but further investigation revealed IHCP. This study was approved by the ethics committee of the Third Medical Centre of Chinese People’s Liberation Army General Hospital. Written informed consent forms were obtained from the patient, who gave his consent that findings and images about himself were published in the journal and associated publications.

2 | CASE REPORT

A 59-year-old male patient presented with 4-day acute painless bilateral visual loss, with no headache, eye dis- tension, photophobia, lacrimation, or diplopia. The pa- tient denied a history of diabetes, hypertension, coronary heart disease, or auto-immune disease. Of note, ION di- agnosis was considered, and nutrient medicine such as
intramuscular injection of mouse nerve growth factor prior to hospitalization did not result in visual recovery.

Upon examination, the best-corrected visual acuity (BCVA) of the right eye was hand movement (HM), and the left eye demonstrated no perception of light (NPL). The movement of right eye in all directions was normal but the left eye was disorder, the pupils of both eyes were round, the pupil diameter of the right eye was 2.5 mm, and that of the left was approximately 5 mm. More, the eyes showed a relative afferent pupillary defect (RAPD), the right eye direct pupillary light reflex (PLR) was weak and indirect PLR was almost absent, the left eye direct and indirect PLR were almost absent, other neurological examinations were normal. Other examinations including Color fundus photography, optical coherence tomography (OCT) of the macula and optic disk photography of the right and left eye were normal (Figure 1).

Laboratory tests showed that the C-reactive protein (CRP) level was 13.1 mg/L (reference value: 0–8 mg/L), and the erythrocyte sedimentation rate (ESR) was 72 mm/h (reference value: 0–15 mm/h). The results of other tests including routine blood, urine routine, blood biochemistry, antinuclear antibody, tuberculosis antibody, mycobacterium tuberculosis γ interferon, syphilis serum antibody, tumor screening, TORCH antibody, IgG4 antibody, and thyroid function tests were normal. The results of cerebrospinal fluid (CSF) tests showed opening pressure was 190 mmH₂O (reference value: 80–180 mmH₂O), CSF protein was 2614 mg/L (reference value: 150–450 mg/L), white blood cell (WBC) was 104 × 10⁶ cells/L (reference value: 0–8×10⁶ cells/L), neutrophil rate was 6%, and lymphocyte rate was 94%. The other biochemistry, cytology, anti-ganglioside antibody, and aquaporin-4 tests were also normal. The results of other tests such as CSF tests to check for bacteria, fungi, cryptococcus neoformans, and acid-fast bacilli were also negative. Imaging tests, including magnetic resonance imaging (MRI) with gadolinium contrast, was presented with dura enhancement of the frontal, anterior cranial fossa, and the brain sickle, especially on the left side, the cavernous sinus and temporal lobe demonstrated possible aggressive signals (Figure 2). No significant high signal was found with diffusion-weighted imaging. In summary, the patient was considered to have IHCP based on laboratory tests and MRI data, the cranial nerves including bilateral optic nerves and left oculomotor nerve were injured, especially the left optic nerve. Treatment with 200 mg cyclophosphamide via intravenous drip infusion, respectively, on Day 1, Day 3, and Day 5, concurrently, methylprednisolone 1000 mg per day via
intravenous drip infusion and then halve reduced every three days, followed by oral prednisone acetate tablets at the end of the 60 mg intravenous drip course. Twenty days after treatment, the BCVA of the patient was significantly restored: that of the right eye reached 20/20, and that of the left eye was 20/200. Unfortunately, visual field examination showed that the visual sensitivity in the right eye had decreased, and examination of the left eye showed that one-quarter of the visual field was completely lost (Figure 3).
3 | DISCUSSION

Hypertrophic pachymeningitis (HP) is a chronic inflammatory disorder characterized by fibrous thickening of the cerebral and/or spinal dural mater. It is described in association with trauma, infections, tumors, autoimmune/inflammatory diseases, and cerebrospinal fluid hypotension syndrome, with some cases remaining idiopathic. Idiopathic hypertrophic cranial pachymeningitis (IHCP), the most common feature of which is chronic intermittent headache, can also be expressed as cephalalgia, local or whole brain blunt pain, or progressive palsy of cranial nerves. One important reason for the ease of cranial nerve damage is the narrowing of the skull gap, in which cranial nerves pass, due to endocranial hypertrophy. The thickened dura might invade the oculomotor nerve, abductor nerve, trochlear nerve, trigeminal nerve, and optic nerve. Severe injury of the optic nerve alone occurs infrequently but can lead to painless rapid bilateral visual loss. Mathew, R.G et al. reported one case of IHCP presenting as acute left-sided painless visual loss, and the patient complained of persistent left-sided headache and numbness on her forehead in the past few years. Our case presented as rapid bilateral visual loss as the initial clinical manifestation, and we have not found similar previous reports.

The diagnosis of IHCP can be made by combining clinical manifestations, imaging examination results, laboratory test results, and pathological features. The pathological characteristics are diffuse fibrous hypertrophy of the dura matter and lymphocytes as well as plasma cell infiltration; IHCP can also present with glass-like changes or caseous necrosis. It can also be observed on MRI and defined pathologically on biopsy; the performance of MRI was correlated with the clinical state in 80% of cases. Imaging demonstrated the diffuse thickening of the dura; the lesions appeared hypointense on T1-weighted sequences and isointense on T2-weighted sequences. The scope of the dura was larger on gadopentetate-enhanced T1-weighted sequences than on plain scans. According to others, laboratory findings might show increased ESR or CRP levels in patients, and some patients show positive results for autoantibodies including ANA, P-ANCA, ACA, and RF.

4 | CONCLUSION

Intracranial neuropathy might occur rapidly and progressively, and the damage is difficult to recover. A clear and correct diagnosis as soon as possible and timely treatment are critical. In this report, the visual acuity of the left eye in the patient was lost three days earlier than that of the right eye; as a result of treatment, the BCVA in the left eye was lower than that in the right eye. The results of this study demonstrate that we should pay attention to intracranial changes as soon as possible in patients with a sharp decline in visual acuity but with no obviously positive signs of the eye fundus.

AUTHOR CONTRIBUTIONS

WYS contributed to patient treatment, conception of the work, manuscript preparation, manuscript revision and final approval of the manuscript and agreed to be accountable for all aspects of the work. CYT, LXG, and BC contributed to the conception of the work, data search and patient treatment, and manuscript revision. QS contributed to patient treatment and manuscript revision.

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CONFLICT OF INTEREST

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

DATA AVAILABILITY STATEMENT

Data and original images in the current study are available from the corresponding author upon reasonable request. Authors can confirm that all relevant data are included in the article and/or its supplementary information files.

ETHICAL APPROVAL

All procedures performed were under the institutional and/or national research committee’s ethical standards and the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. This study was approved by the ethics committee of the Third Medical Centre of Chinese PLA General Hospital.

CONSENT

An informed written consent was obtained from the patient.

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