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CASE REPORT

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Combined Salvage Treatments in a Case of Neurosyphilis

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ABSTRACT

Purpose: Visual loss and optic nerve head (ONH) edema are hallmarks in diagnosing optic neuritis, perineuritis, neuroretinitis and papillitis, but clinicians often disregard the role that intracranial hypertension could play in neurosyphilis.

Methods: A case of syphilitic optic neuropathy with visual loss and bilateral ONH edema was presented, where the detailed diagnosis procedure of chronic intracranial hypertension and its treatment were described.

Results: Once the syphilitic intracranial hypertension was diagnosed, intravenous mannitol, penicillin G and emergent bilateral optic nerve sheath decompression were given. Vision improved from bilateral no light perception to light perception in the right eye and count fingers in the left. Headaches and ONH edema were also relieved.

Conclusions: Visual loss and bilateral ONH edema can be caused by not only syphilitic optic neuritis but also syphilitic intracranial hypertension or both. The importance of assessing and treatment for intracranial hypertension in neurosyphilis should be kept in mind.

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Abbreviations

ONH: Optic Nerve Head

OU: both eyes

OD: right eye

OS: left eye

CT: Computed Tomography

MRI: Magnetic Resonance Imaging

ONS: Optic Nerve Sheaths

ICP: Intracranial Pressure

CSF: Cerebral Spinal Fluid

WBC: White Blood Cell

HIV: Human Immunodeficiency Virus

MRV: Magnetic Resonance Venogram

TPPA: Treponema Pallidum Particle Assay

TRUST: Tolulized Red Unheated Serum Test

RPR: Rapid Plasma Reagintest

CSF-VDRL: Cerebral Spinal Fluid-Venereal Disease Research Laboratory.

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Background

Visual loss and optic nerve head edema is not an unusual manifestation in neurosyphilis. It usually causes clinicians to make the diagnosis of optic neuritis, perineuritis, neuroretinitis and papillitis. In addition to optic neuritis, perineuritis, neuroretinitis and papillitis, intracranial hypertension should be considered in differential diagnosis [1].

We report a case of syphilitic optic neuropathy with intracranial hypertension that presented with progressive visual loss and bilateral optic nerve edema. The patient has previously been misdiagnosed, mismanaged and misinformed on their prognosis. The defining moment was the relief of intracranial hypertension associated with neurosyphilis that provided protection and improvement of the optic nerve head and visual function while in our care.

Case Presentation

A 37-year-old male presented with progressive visual loss OU and headache for eight months was admitted to ophthalmology department.

Eight months ago he noted bilateral vision blurring and mild headache.

Six months ago, his local ophthalmologist noted bilateral optic nerve head (ONH) edema and obtained a brain CT which was reportedly unremarkable. The patient refused further work-up or treatment at the time. But he was admitted to his local hospital due to worsening vision and headaches two months ago. His visual acuity was 20/200 OD, 20/40 OS with significant bilateral ONH edema. An enhanced cranial/spinal MRI revealed sphenoid sinusitis. Endoscopic sphenoid sinus drainage surgery was performed which was unremarkable for fungal sinusitis. Intravenous cephalosporin and methylprednisolone was given for 7 days postoperatively and was then discharged.

Three days prior to admission to our hospital, his vision had progressed to no light perception bilaterally.

On admission, examination revealed significant bilateral ONH edema (Figure 1), dilated and unreactive pupils, and mild bilateral abduction limitation. He was noted to have a normal body mass index, with no other neurological deficits detected. No significant family history was revealed. He denied illicit drug usage or toxic exposure. He did admit to occasional usage of self-bought antibiotics such as amoxicillin for colds. Previous MRI and CT were carefully reviewed, which were remarkable for dilated bilateral retra-bulbar optic nerve sheaths (ONS). This finding was confirmed with a repeat MRI of the brain and orbits (Figure 2). Intracranial hypertension was suspected and a lumbar puncture was performed with intracranial pressure (ICP) over 500 mmH2O. Cerebral spinal fluid (CSF) analysis demonstrated WBC of 2 cells /μL with normal protein and glucose. No bacterium or fungi was found. MRV was performed without abnormal findings. Intravenous mannitol was administered.

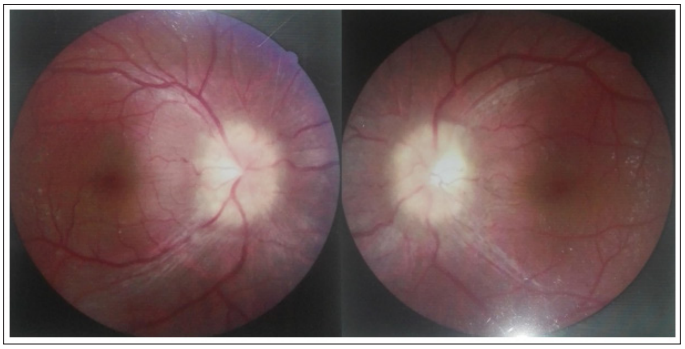

Figure 1: Significant bilateral ONH edema

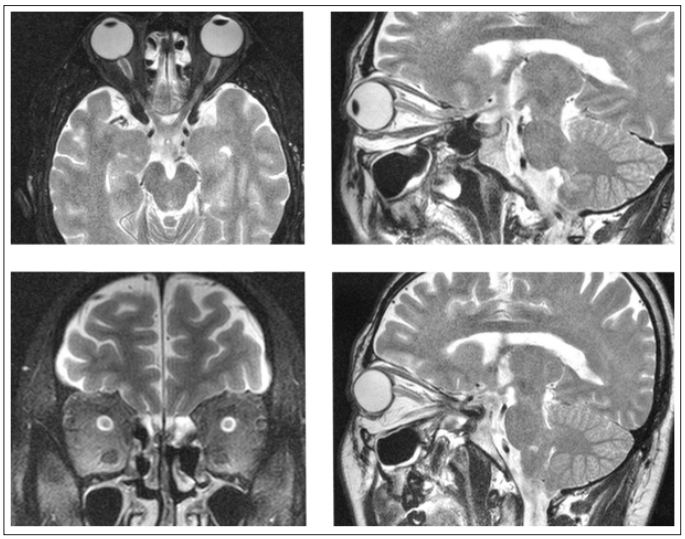

Figure 2: MRI of the brain and orbits shows dilated bilateral retra-bulbar optic nerve sheaths (ONS)

Lab tests including HIV, autoimmune antigens/antibodies returned negative. Serum TPPA and TRUST were weakly positive with a titer of 1:1, repeated serums TPPA, TRUST, RPR from other two hospitals were similar. Upon further questioning, he admitted a recurring history of unprotected sexual intercourse over the past eight years, but specifically denied genital ulcer, rash, or standard anti-syphilis treatment. CSF was re-collected and CSF-VDRL was positive (1:1).

A syphilitic optic neuropathy was suspected, in combination with chronic intracranial hypertension. Intravenous penicillin G (4 million units every 4 hs) was administered over seven days but with no improvement in vision or relief of symptoms. Bilateral optic nerve sheath decompression, a surgery to stabilize or improve visual function in idiopathic intracranial hypertension, was performed emergently. Within two days, the vision had improved to light perception in the right eye and count fingers in the left, and the headache and ONH edema were relieved with residual optic atrophy [2]. Penicillin was continued for a total of 20 days with no improvement in visual acuity and was discharged. He was lost one month after the discharge.

Discussion and Conclusions

Syphilitic intracranial hypertension is not an uncommon complication in neurosyphilis. In pre-antibiotic era, as high as 66% patients presented with intracranial hypertension in syphilitic meningitis, of which 15% had ICP over 400 mmH2O, and 51% had ICP between 210-400 mmH2O. There was also 15% with ICP over 210 mmH2O in cerebrovascular neurosyphilis, and intracranial hypertension could also occur in general paresis [3]. In the current post-antibiotic era intracranial hypertension has decreased, but should not be disregarded as there still are a few confirmed reports [4, 5].

It is well known that chronic intracranial hypertension may lead to visual loss and other nervous system damage. The visual impairment in our patient was likely multifactorial such as persistent intracranial hypertension, direct damage to the optic nerve and/or vasculature from Treponema pallidum. Although the penicillin was initially given, the patient's history and the immediate visual improvement after optic nerve sheath decompression indicated the chronic intracranial hypertension was probably an important cause to the visual loss.

From this case the following notes might help clinicians in identifying and treating neurosyphilis accompanied with chronic intracranial hypertension:

1. The visual loss in intracranial hypertension is a slow, progressive one with headache, while that in neuritis is usually a rapid vision loss associated with eye pain.
2. Bilateral dilated ONS found in the MRI of the orbits, an identified characteristic sign of idiopathic intracranial hypertension, contributed to confirming the diagnosis of syphilitic intracranial hypertension [6]. We continue to recommend orbital MRI as a helpful tool in determining the presence of intracranial hypertension. It should be considered in neurosyphilis with presumed intracranial hypertension.
3. In addition to shunt surgery, optic nerve sheath decompression has been successfully used for the maintenance or restoration of visual function and recommended as the primary choice for the patient with visual loss in intracranial hypertension [7]. For secondary intracranial hypertension due to intracranial sinus thrombosis or Cryptococcus meningitis, optic nerve sheath decompression has also shown potential for the vision protection [8]. Optic nerve sheath decompression should be considered early in treatment to restore and preserve visual function, such as in the case of our patient.
4. Although penicillin is the primary drug of choice for the treatment of syphilis, there are reports of resistance to penicillin [9-11]. With the aforementioned resistance, chronic intracranial hypertension, an ICP over 500 mmH2O, and binocular blindness for ten days, we decided to perform the bilateral optic nerve sheath decompression emergently, 7 days after standard penicillin treatment was given.

In general, clinicians should consider that visual loss and bilateral ONH edema could be caused by not only syphilitic optic neuritis but also syphilitic intracranial hypertension or both. The importance of assessing and treatment for intracranial hypertension in neurosyphilis should be kept in mind. Delayed diagnosis and treatment can lead to permanent visual loss in neurosyphilis.

Declarations

Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Competing Interests: The authors declare that they have no competing interests.

Authors' Contributions: Hui Chen, Huan Wang treated the patient and in doing so acquired the case data, they were also involved with drafting of the manuscript. Hui Chen, Huan Wang revised the manuscript. All authors read and approved the final manuscript.

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