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Case report
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A rare case of fulminant idiopathic intracranial hypertension

Sylvester **Wong** Leong Kiat[1,2], **Tan** Vi Chee[1], **Tan** Li Mun[1,3], **Lam** Chenshen[1,4]

[1]Department of Ophthalmology, Hospital Sibu, Sarawak, Malaysia; [2]UM Eye Research Centre (UMERC), Department of Ophthalmology, Faculty of Medicine, Universiti Malaya, Kuala Lumpur, Malaysia; [3]Department of Ophthalmology, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia; [4]Department of Ophthalmology, Faculty of Medicine, Universiti Kebangsaan Malaysia, Kuala Lumpur, Malaysia

Abstract

Background: Idiopathic intracranial hypertension (IIH) is characterised by raised intracranial pressure and normal cerebrospinal fluid composition with no detectable intracranial aetiology. Fulminant IIH is rare and is defined by the acute onset of symptoms and signs (less than 4 weeks between onset of symptoms and severe vision loss) with progressive vision loss over days.
Case presentation: A 22-year-old woman with body mass index of 27.6 was diagnosed with fulminant IIH. Her presenting vision was hand movement and it progressed rapidly to no light perception by day 4. She was managed with oral acetazolamide and intermittent lumbar punctures. Nevertheless, she did not regain her vision.
Conclusion: Fulminant IIH is a vision-threatening condition that must be recognised and treated early to prevent irreversible vision loss.

Keywords: blindness, fulminant idiopathic intracranial hypertension, pseudotumor cerebri

Correspondence: Dr. Lam Chen Shen, Department of Ophthalmology, Faculty of Medicine, Hospital Canselor Tuanku Muhriz Universiti Kebangsaan Malaysia, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia.
E-mail: chenshen_lam@yahoo.com

Kes hipertensi intracranial idiopatik fulminan yang jarang ditemui

Abstrak

Pengenalan: Hipertensi intrakranial idiopatik (IIH) adalah penyakit yang bercirikan dengan peningkatan tekanan intrakranial tetapi kandungan cecair serebrospinal yang normal, tanpa sebarang etiologi intrakranial. Fulminan IIH jarang berlaku dan didefinisikan dengan simptom dan tanda yang akut (kurang daripada 4 minggu antara permulaan simptom dan kehilangan penglihatan yang teruk) dengan kehilangan penglihatan yang progresif dalam tempoh beberapa hari.
Pembentangan kes: Seorang wanita berumur 22 tahun dengan indeks jisim badan 27.6 telah dikesan menghidapi fulminan IIH. Pesakit pada mulanya datang dengan penglihatan yang terhad hanya dapat melihat pergerakan tangan sahaja. Namun 4 hari kemudian, penglihatan pesakit menjadi bertambah teruk sehingga tiada persepsi cahaya. Pesakit diberi rawatan dengan acetazolamide secara oral dan tebukan lumbar secara berkala. Walau bagaimanapun, penglihatan pesakit ini tidak dapat dipulihkan.
Kesimpulan: Fulminan IIH adalah keadaan yang mengancam penglihatan dan perlu dikenal pasti awal. Rawatan awal diperlukan untuk mengelakkan kehilangan penglihatan yang kekal.

Kata kunci: buta, hipertensi intracranial idiopatik fulminan, pseudotumor cerebri

Introduction

Idiopathic intracranial hypertension (IIH) is a disorder characterised by raised intracranial pressure (ICP) that predominantly affects young, obese women with no established pathogenesis.[1] Common symptoms of IIH include headaches, transient visual disturbance, pulsatile tinnitus, and back or neck pain.[2] Diagnosis is made based on modified Dandy criteria,[3] which requires the presence of:

1. signs and symptoms of increased intracranial pressure;
2. no other neurological abnormalities or impaired level of consciousness (with the exception of cranial nerve VI palsy);
3. elevated ICP with normal cerebrospinal fluid (CSF) composition;
4. a computed tomography (CT) scan which shows no aetiology for increased ICP;
5. no other apparent cause.

The overall age-adjusted and gender-adjusted annual incidence is reported to be 2.4 per 100,000 (2002–2014).[4]

Fulminant IIH is defined by its acute onset (less than 4 weeks between symptom onset and severe vision loss) and rapid worsening of vision over days.[5] Fulminant IIH occurs in 2–3% of patients with IIH.[5] These patients are at high risk of permanent legal blindness if diagnosis and definitive surgical treatment are delayed.[5]

Case report

A 22-year-old overweight Melanau woman with underlying systemic lupus erythematosus (SLE) presented to the emergency department with sudden onset of reduced bilateral vision for 1 day. She also complained of headache, shortness of breath, neck and back pain. She had no eye pain, redness, or discharge. She denied any recent trauma, fever, or infective symptoms. Her body mass index was 27.6. Her blood pressure (BP) was 185/142 mmHg on arrival.

The patient had been diagnosed with SLE in 2015. However, she frequently defaulted her treatment. Due to poor control of the SLE, she often presented to the emergency department with fluid overload, acute pulmonary oedema, or hypertensive emergency. Her last clinic visit was 8 months prior to the admission reported herein. She was on oral mycophenolate mofetil 500 mg BD and prednisolone 15 mg BD.

Bilateral visual acuity (VA) was hand movement and relative afferent pupillary pathway defect was negative. Anterior segment examination was unremarkable with normal intraocular pressure. Fundus examination revealed bilateral swollen optic discs with dilated and tortuous vessels, flame-shaped haemorrhages, and multiple blot haemorrhages in all quadrants. Macular star, vascular sheathing, and ghost vessels were not seen on presentation. Optic cups were obscured by swelling. There was no retinitis, vasculitis, or vitritis. Further examination revealed right abducens nerve palsy.

Among the differentials were hypertensive emergency and hypertensive retinopathy as her BP was high. Another differential was papilledema. Increased ICP can result from hypertensive emergency, space-occupying lesion, or venous stasis/thrombosis. The third differential was optic neuritis. Multiple sclerosis (MS) and neuromyelitis optica (NMO) spectrum disorder needed to be considered as she was female, age 22, and had bilateral optic disc swelling with poor VA. The fourth differential was central nervous system (CNS) infection. She had symptoms of headache, neck stiffness, and positive Kernig's sign. Even though there was no fever, CNS infection could not be ruled out as she was immunocompromised. The patient was diagnosed with grade 4 hypertensive retinopathy and hypertensive emergency, and treated for cerebral lupus.

On admission, the non-enhanced computerized tomography (CT) of the brain showed no intracranial abnormality. She was restarted on her medication and the BP was brought under control on day 2 of treatment. However, her vision further

deteriorated to light perception despite well-controlled BP. Other ophthalmological assessment findings remained similar.

Contrast-enhanced CT of the brain reported bilateral symmetrically thickened optic nerves with no space-occupying lesion noted. Venous sinuses were patent. Lumbar puncture was performed and the opening pressure was greater than 50 cm H_20. Normal CSF opening pressure is 10 to 25 mm H_20. The CSF sample was otherwise clear with normal composition. Infective screening of the CSF was all negative as well.

The patient was subsequently diagnosed with fulminant IIH and immediately started on tablet acetazolamide 250 mg QID. She was referred to the neurosurgery team for a CSF shunting procedure, which the patient refused.

On day 4 of admission, bilateral VA further dropped to NLP and progressed to bilateral abducens nerve palsies. Therapeutic lumbar puncture was performed. The opening pressure was 38 cm H_2O. Shortly after, the abducens nerve palsies resolved, but vision remained at NLP. Fundus examination showed reduced swelling of the optic discs with less vessel tortuosity, but both optic discs were already palish (Fig. 1).

Magnetic resonance imaging (MRI) of the brain and orbit on day 6 of admission demonstrated enhancement of both optic nerves (intraorbital) and flattening of posterior pole (Fig. 2). She was referred to the neurology team. After reviewing the MRI findings, the neurology team concurred with the diagnosis of fulminant IIH. CSF oligoclonal band test as well as serum and CSF aquaporin-4 test came back negative, which ruled out MS and neuromyelitis optica. The MRI findings likely represented the cuff of the CSF signal surrounding the optic nerve. As the patient had increased ICP, more CSF fluid accumulated within the dural sheath of the optic nerve. The patient's bilateral vision did not recover upon discharge and follow-up.

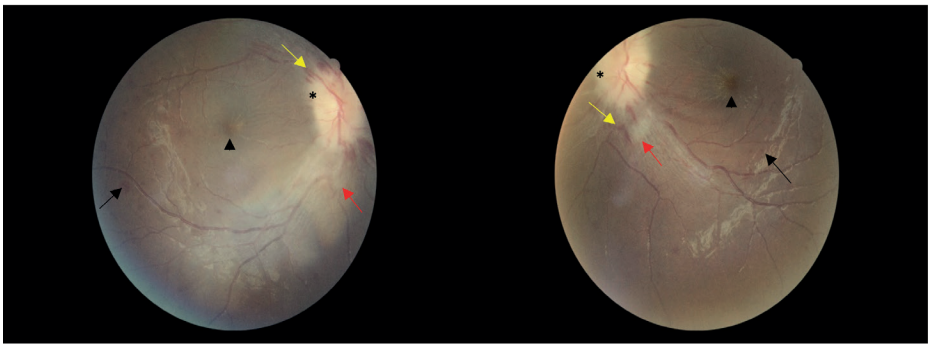

Fig. 1. Fundus images of the right eye (*right*) and left eye (*left*) taken on day 11 of admission. The images show optic nerve head oedema (asterisk), macular star (arrowhead), dot-blot haemorrhage (black arrow), flame-shaped haemorrhage (yellow arrow), and cotton-wool spots (red arrow).

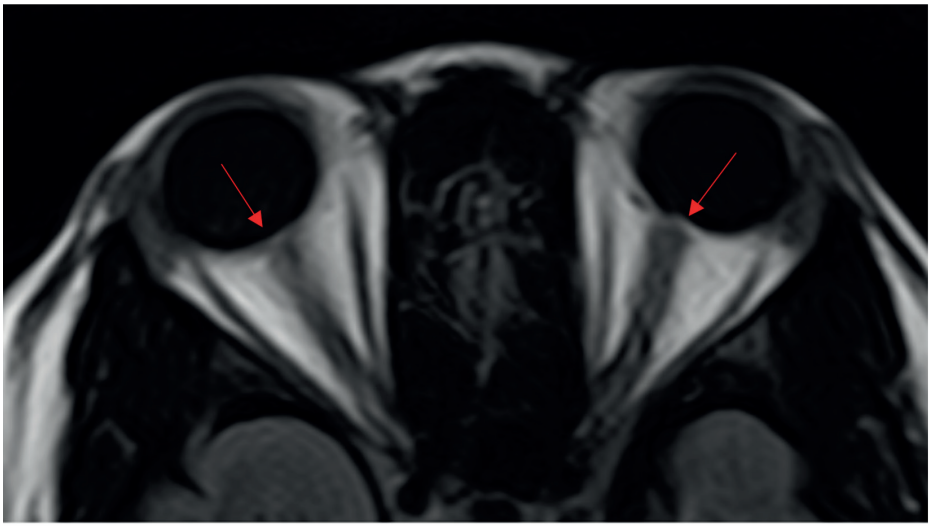

Fig. 2. Axial cut of a fluid attenuated inversion recovery magnetic resonance image showing posterior globe flattening (red arrows).

Discussion

This patient presented with bilateral optic disc swelling and rapid deterioration of vision. Differential diagnoses included papilledema, space-occupying lesion, malignant hypertension, venous stasis, venous thrombosis, optic neuritis from MS or NMO spectrum disorder, bilateral sequential non-arteritic ischaemic optic neuropathy, giant cell arteritis, sarcoidosis, infiltrating optic neuropathy, and compressive optic neuropathy.[6]

Such a patient must undergo neuroimaging to rule out a space-occupying lesion. If neuroimaging is normal, lumbar puncture should follow to measure opening pressure and check CSF composition. If the patient is suspected to have optic neuritis, MRI of the brain and orbit with fat suppression and gadolinium contrast should be ordered.

Fulminant IIH can lead to severe and rapid vision loss. Visual loss is often irreversible unless prompt treatment is administered. Retrospectively, this patient had risk factors for IIH as she was an obese female of child-bearing age with SLE and a history of steroid therapy.

The management of fulminant IIH differs fundamentally from that of typical IIH. Patients with fulminant IIH are at high risk of permanent, profound visual loss if surgical intervention is delayed.[5] Effective treatment of fulminant IIH requires a combination of medical and surgical intervention.[5] All patients should be started on a high dose of carbonic anhydrase inhibitor.

The IIH Treatment Trial has shown that acetazolamide treatment improves perimetric mean deviation, reduces ICP, and increases weight reduction.[7] The dosage was started at 500 mg BD and subsequently increased to 1 tablet every week to a maximum dose of 4 g per day.[7] Wall *et al.* also recommend surgical management for patients with perimetric mean deviation worse than -7 dB.[7]

Surgical interventions include CSF shunting, venous sinus stenting, and optic nerve sheath fenestration. CSF shunting is a procedure performed to reduce ICP. Options include ventriculoperitoneal shunt and lumbar peritoneal shunt. Venous sinus stenting places a tubular metallic mesh in the area of narrowed cerebral venous sinuses to expand the vein and resolve the narrowing. Optic nerve sheath fenestration creates holes in the optic nerve sheath to release CSF from subarachnoid space around the optic nerve.

If surgical intervention is expected to be delayed for more than 24 hours or if vision loss was rapid, temporizing CSF drainage should be considered via lumbar drain, extraventricular drain, or serial large-volume lumbar puncture.[8] In the largest series of patients with fulminant IIH, Thambisetty *et al.* found that a faster surgery with a median delay of 2 days showed better visual recovery compared to a median delay of 6 days. The 8 patients whose surgery was delayed had an outcome of legal blindness.

Our patient was managed with oral acetazolamide. She refused the CSF shunting procedure. In the end, the VA did not recover and remained as NLP in both eyes. Prompt diagnosis and surgical intervention may have helped improve her VA. During the patient's in-patient treatment, optical coherence tomography was not done as there was no facility in-house. The patient was unable to proceed with visual perimetry test as she was unwell and unable to sit properly due to headache and neck pain.

Diagnosis was also delayed because of the presence of other comorbidities, which led to the presumption of hypertensive retinopathy with cerebral lupus, MS, or NMO. The headache and visual loss did not improve with initial management. Lumbar puncture and further imaging were done, leading to diagnosis of fulminant IIH. Delay in diagnosis and management of fulminant IIH most likely contributed to the poor visual outcome in this patient. Weight loss is the only disease-modifying therapy in IIH.[2]

Conclusion

Fulminant IIH is a vision-threatening condition that needs to be recognised and treated early to prevent irreversible vision loss. This involves a multidisciplinary team approach, imaging, and investigations. Close communication between all teams is necessary to hasten the examinations and investigations necessary to establish a prompt IIH diagnosis. Patients should be counselled on the importance of reducing weight and referred to a dietician.

Declarations

Consent for publication
The patient provided informed consent for the use of the clinical images and data contained in this case report.

Competing interests
None to declare.

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