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CASE REPORT

Thunderclap Headache with Bilateral Blindness: A Rare Presentation of Idiopathic Intracranial Hypertension

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Abstract

Objective: Idiopathic intracranial hypertension (IIH) is known to present with thunderclap headache, transient visual obscurations and tinnitus. Here we report a case of idiopathic intracranial hypertension with a rare presentation of thunderclap headache associated with bilateral blindness.

Case presentation: A 28-year-old male with history of migraineous headaches since his late teens, presented with acute onset of thunderclap headache and episodes of bilateral complete blindness, lasting from 10 minutes to an hour. Routine blood investigations, CT brain, CT angiogram and CT venogram were all normal. Lumbar puncture with CSF manometry revealed elevated CSF opening pressure suggesting a diagnosis of idiopathic intracranial hypertension. His symptoms improved completely after CSF tap. He was started on acetazolamide for prophylaxis.

Conclusion: In migraine patients, a change in character of headache should immediately prompt the physician to search for intracranial organic pathologies. IIH should be considered in the differential diagnosis of thunderclap headache especially with visual phenomena, given the excellent response of it with early treatment.

Introduction

Idiopathic intracranial hypertension (IIH) is a disorder of elevated cerebrospinal fluid pressure of unknown cause [1]. It occurs more frequently in obese females [2]. The diagnosis of IIH requires that the patient meet all of the modified Dandy criteria: 1. Signs and symptoms of increased intracranial pressure. 2. No other neurological abnormalities or impaired level of consciousness. 3. Elevated CSF opening pressure with normal CSF composition. 4. A neuroimaging study that shows no etiology for increased ICP. 5. No other cause for intracranial hypertension is found [3, 4]. IIH patients usually presents with headache (94%), transient visual obscurations (68%), pulse synchronous tinnitus (58%), photopia (54%), and retro bulbar pain (44%), whereas diplopia (38%) and progressive visual loss (30%) are less common [1]. However to the best of our knowledge, IIH manifesting as sudden onset prolonged bilateral blindness has not been reported yet.

Case Presentation

A 28-year-old man with history of migraineous headaches since his late teens presented to the emergency with acute onset thunderclap headache with complete blindness lasting for 10-15 minutes. The headache was not related to any posture or activity. He was evaluated by ophthalmologist initially and was documented to have normal ophthalmological examination. He was then referred to our neurology service. He denied history of any medication use in the recent past. There was no history of significant medical or surgical illnesses in the past. His vital signs were stable. The neurological examination revealed normal higher mental functions; pupillary reflexes were normal and symmetrical without any afferent pupillary defect, extra-ocular movements were normal. Opt kinetic nystagmus was absent bilaterally. Optic fundi were normal. Rest of the cranial nerve examination, motor, sensory and cerebellar examination was normal. There were no meningeal signs. The working differential diagnosis at the time included bilateral occipital infractas, subarachnoid haemorrhage and cortical venous sinus thrombosis. Accordingly, the patient urgently underwent a CT brain, CT angiogram and CT venogram. All of which were reported to be normal. Occipital lobe epilepsy was considered unlikely due to the prolonged duration of the episode of vision loss.

He received analgesics and the headache gradually improved over a few hours. He was discharged home with the provisional diagnosis of Acute Migraine with visual aura and was started on Topiramate for migraine prophylaxis.

The next morning, he woke up with a severe headache and blurring of vision, which progressed to complete blindness lasting for an hour. There was associated nausea but no

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vomiting. He returned again to the emergency with the same complaints. Neurologic examination at the time showed no lateralizing neurological deficits. Optic fundi examination showed normal disc margins; however, spontaneous venous pulsations were absent on both sides. With the clinical picture suggestive of raised ICP, the possibility of Idiopathic intracranial hypertension was considered. Lumbar puncture with CSF manometry was done which revealed elevated CSF opening pressure confirming the diagnosis of IIH.

Investigations

Routine blood investigations including electrolytes, renal and liver functions, thyroid function tests and vitamin D levels were normal. CT brain, CT angiogram and venogram were normal. Lumbar puncture with CSF manometry revealed opening pressure was 270 mm of water, rest of the CSF parameters were normal. (Protein 27mg/dL, Sugar 60mg/dL). CSF microscopy was normal and CSF culture was sterile.

Treatment and Outcomes

After lumbar puncture, his headache subsided completely. He was started on acetazolamide for prophylaxis. His follow up at 2 weeks and later at 2 months showed that he was completely symptom free, with no further recurrence of headache or visual symptoms. Optic fundi examination was normal on follow up.

Discussion

IIH is uncommon to be seen in males and non-obese patients. In a population-based sample of 81 IIH patients, 76 were females while only 5 were males. The age at presentation was in the range of 8-55 years, with a mean age of 28-years-old [2]. IIH is known to cause visual abnormalities. Up to 68% of patients may present with transitent visual obscurations. Vision loss has also been reported with IIH in 30% of patients [1]. Papilledema is the commonest physical sign observed in visión loss associated with IIH [5]. Although it is usually bilateral, unilateral and highly asymmetric papilledema has also been documented in IIH [6]. Papilledema may be absent in about 6% of patients with proven IIH [5]. The pathogenesis of visual loss in IIH is postulated to be due to the disruption of axonal transport and intraneuronal optic nerve ischemia [7]. Visual obscurations in IIH may be monocular or binocular. They usually last less than 30 seconds and are followed by visual recovery back to baseline [2, 8]. Abducens nerve palsy, the only currently accepted false localizing sign in IIH can be unilateral or bilateral and is seen in about one-third of IIH patients [9]. Other focal deficits including various patterns of ophthalmoplegia such as transient or established pupillary-sparing third nerve palsy, fourth cranial nerve palsy, internuclear ophthalmoplegia, skew deviation, diffuse bilateral ophthalmoplegia, fifth and seventh nerve palsies have all been described in isolated case reports of patients with presumed IIH. Stanley TV, described a 14 year old girl with idiopathic intracranial hypertension that presented as hemiplegic migraine [10]. Sorensen and Corbett noted that pre-existing migraine may get worsened in IIH [11]. Ramadan reported episodic migraine getting transformed to chronic daily headache in a patient with IIH without papilledema [12]. Basilar migraine or migraine with brainstem aura is known to present with bilateral cortical blindness. Sturzenegger and Meienberg in their follow up study of 82 patients with basilar-type migraine found out that bilateral visual impairment occurred in 86% of patients while 18% had complete blindness [13]. Aura symptom duration lasted from 3 minutes to 60 hours with 75% of patients having auras between 5 and 60 minutes. The pathogenesis of blindness is supposed to be due to spreading depression in bilateral occipital cortices which has been proven in animals [14]. Shahar and Barak in 2003 reported that the duration of vision loss was between 1 and 10 minutes in their study including 14 patients with epileptic complete blindness [15]. However bilateral cortical blindness with thunderclap headache as initial presentation of IIH has not been reported. We hypothesize that IIH in our case would have precipitated a change in the character of headache and the mechanism of blindness could be similar to that of basilar migraine explaining the prolonged bilateral vision loss.

Conclusion

Our patient who was known to have history of migraineous headaches, presented to the hospital with acute onset of thunderclap headache and episodes of bilateral blindness. While thunderclap headache is a commonly presenting symptom of IIH, vision loss and specifically bilateral prolonged vision loss is less commonly seen [1]. We propose that the mechanism of blindness in IIH in this case is possibly similar to that of basilar migraine. In migraine patients, a change in character of headache should immediately prompt the physician to search for intracranial organic pathologies. IIH should be considered in the differential diagnosis of thunderclap headache especially with visual phenomena, given the excellent response of it with early treatment.

Statement of Ethics

The patient gave written consent to share his case.

Conflict of Interest Statement:

The authors declare that they have no conflicts of interest to disclose.

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