Case Report

Idiopathic intracranial hypertension presenting as acute bilateral loss of vision

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ABSTRACT

Acute bilateral vision loss is a very rare presentation of idiopathic intracranial hypertension (IIH). We report the case of a 17-year-old female student who presented with a history of moderate degree headache from 7 days and severe loss of vision of 6 days duration. The vision loss initially affected the right eye and over the next 2 days, her left eye was also involved. Her visual acuity in both eyes was 6/60 and she had a relative afferent pupillary defect on the right side. Fundus examination showed bilateral papilledema and perimetry showed severe bilateral constriction of visual fields with the involvement of central areas as well. Cerebrospinal fluid opening pressure was raised and magnetic resonance imaging showed signs suggestive of IIH. She was managed with acetazolamide, topiramate, and lumbar drainage. She showed a gradual improvement in her vision, and finally, her visual acuity returned to normal.

Keywords: Idiopathic intracranial hypertension, Papilledema, Vision loss

Idiopathic intracranial hypertension (IIH) is a headache syndrome characterized by (a) signs and symptoms of generalized intracranial hypertension; (b) elevated cerebrospinal fluid (CSF) pressure; (c) a normal CSF composition; (d) no evidence of hydrocephalus or space-occupying lesion on neuroimaging, and (e) no other cause of intracranial hypertension [1]. The terms such as serous meningitis, pseudotumor cerebri, and benign intracranial hypertension were previously used for this condition. IIH is a rare condition, occurring in the general population with an incidence of 0.9 cases per 100,000 populations in the United States. It is most common in women of reproductive age group and among obese individuals with a female-to-male ratio of 8:1 [2,3]. The course of the disease is highly variable from being absolutely benign with minimal symptoms to being progressive with permanent vision loss. IIH presenting as acute severe vision loss is exceedingly rare with only a few cases reported in the literature. The present case highlights this atypical presentation of IIH with a favorable prognosis with early treatment.

CASE REPORT

A 17-year-old female presented was referred to the neurology outpatient department with a history of the headache of 7 days and diminution of vision of 6 days duration. Headache was moderate in intensity, global in location, and aggravated by straining and coughing. One day after the headache, the patient developed a diminution of vision in the right eye. The vision progressively worsened over the next 2 days and she developed vision loss in her left eye also. She also had episodic worsening of vision lasting for seconds during which she felt completely blind. She assured that she had been able to read until 7 days before the presentation. There was no history of pain or redness of eyes, floaters, diplopia, fever, vomiting, and photophobia, drug intake, and rash, alteration in sensorium, abnormal body movements, or weakness of any part of the body.

On examination, she was conscious and oriented. The pulse rate was 72 bpm and blood pressure 110/80. Her weight was 60 kg and the body mass index was 24. Visual acuity in both eyes was 6/60 and intraocular pressure was normal. Ocular movements were normal but she had a relative afferent pupillary defect (RAPD) on the right side. Fundus examination showed bilateral papilledema with peripapillary vessel attenuation (Fig. 1). Perimetry showed severe bilateral constriction of visual fields with the involvement of central areas as well, more so in the right eye (Fig. 2). The rest of the examination was normal.

Her investigations revealed a normal hemogram, kidney function tests, liver function tests, blood glucose, and chest X-ray. Magnetic resonance imaging (MRI) showed bilateral tortuous optic nerves, posterior flattening of the globe, and partially empty sella (Fig. 3). There was no contrast enhancement and

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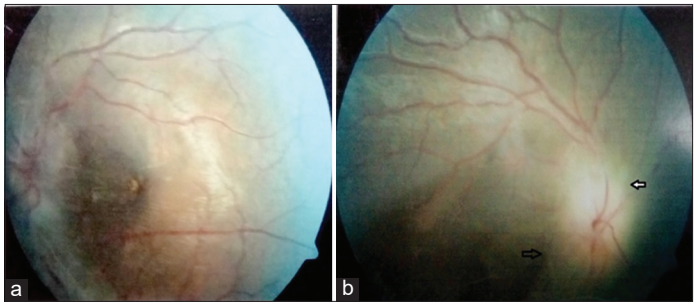

Figure 1: (a) The left fundus showing papilledema. (b) The right fundus showing papilledema (black arrow) with attenuation of vessels (white arrow)

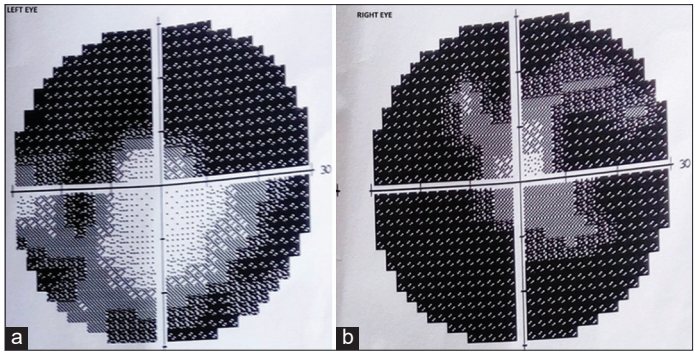

Figure 2: Perimetry showing bilateral severe constriction of visual fields with involvement of central visual areas as well more on the right side

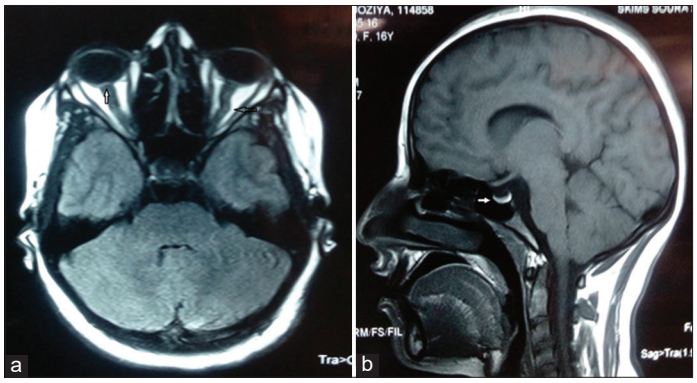

Figure 3: (a) Axial T2 magnetic resonance imaging (MRI) showing flattening of the globe (white arrow) and tortuous optic nerve (black arrow); (b) sagittal view contrast MRI brain showing partially empty sella (white arrow)

cranial venography was also normal. CSF examination revealed an opening pressure of 320 mm of water. There were no cells, and protein and glucose levels were normal. Her autoimmune antibody profile was negative. Furthermore, aquaporin-4 antibody was negative.

A diagnosis of IIH was made and the patient was started on acetazolamide 250 mg tid and topiramate 25 mg bid with a gradual escalation of dosage to 500 mg tid and 50 mg bid, respectively. Lumbar drainage with the removal of about 40 ml of CSF was done twice during the first 4 days of admission. She showed a gradual improvement in her vision, and after 2 weeks of treatment, her visual acuity was 6/12 in both eyes. After 2 months, her visual acuity returned to normal.

DISCUSSION

IIH is a disorder characterized by raised intracranial pressure of unknown cause, predominantly seen in women of childbearing age and associated with a history of recent weight gain. The association of this disorder with weight gain suggests that its incidence and prevalence will increase in accordance with the global epidemic of obesity [4].

Headache is the most common clinical manifestation of IIH in the majority of patients and is also the most frequent presenting symptom. Headache is characteristically pressuring like, throbbing, usually unremitting, and may be accompanied by nausea. The headache may resemble a migraine or tension-type headache or have features of raised intracranial pressure with worsening by coughing, straining, or the Valsalva maneuver [5]. The headache usually precedes the final diagnosis by months. Our patient, however, had a short history of headache which is unusual for IIH.

Visual abnormalities are the next most common symptoms. Transient visual obscurations, often described as dark patches or black spots, are the most common visual manifestation. IIH causes blindness in up to 10% of cases which can be progressive [6]. Although vision loss may initially be reversible, it tends to be permanent once retinal nerve fiber layer loss and retinal ganglion cell atrophy develop. However, central vision is usually spared until late in the course of the illness resulting mainly in visual field constriction and tunneling of vision. In the majority of patients, visual impairment typically evolves slowly with deficits arising over several weeks to months [7,8]. Our patient's course and tempo of visual loss were atypical for IIH with acute severe bilateral loss of both central and peripheral vision.

The important differential diagnosis of a patient presenting exclusively with acute onset headache and progressive bilateral vision loss includes cerebral venous thrombosis, bilateral optic neuritis as seen in neuromyelitis optica, posterior reversible encephalopathy syndrome, bilateral posterior cerebral artery stroke, pituitary apoplexy, and acute demyelinating disorders. All of these disorders were excluded with appropriate investigations in our patient.

The arrival of MRI marked a paradigm shift in the utility of imaging in IIH. The various radiological features of IIH detected through MRI include empty or partially empty sella, flattening of the posterior sclera, enlargement of perioptic subarachnoid space, increased tortuosity of the optic nerve, intraocular protrusion of optic nerve head, enhancement of optic nerve, and slit-like ventricles [9]. Our patient had four of these seven features and fulfilled the criteria for the diagnosis of IIH [1].

Restoration of visual acuity and resolution of papilledema are the main goals of management in IIH. Most patients can be managed with conservative treatment. Acetazolamide has been the most commonly used drug to treat IIH. Topiramate also reduces CSF pressure besides having beneficial action against headaches and helping in weight reduction. Shaia and Elzie described a case of fulminant IIH with right RAPD which

was managed successfully with acetazolamide [10]. However, in patients with threatened vision, early surgical intervention may be necessary. Our patient presented with rapid onset vision loss with bilateral involvement. Many such patients need CSF diversion procedures such as ventriculoperitoneal shunting or optic nerve fenestration. Mensah *et al*. reported a case of malignant IIH which showed no response to steroids and acetazolamide and stabilized only after ventriculoperitoneal shunting [11]. Our patient was managed with drugs along with CSF drainage and showed a satisfactory response to these modalities.

CONCLUSION

We can conclude that any patient of IIH with a short history of visual loss may initially be managed medically before shifting to more invasive surgical treatment. The present case highlights this atypical presentation of IIH with a favorable prognosis with early treatment.

AUTHORS' CONTRIBUTIONS

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