Complete ophthalmoplegia: A rare presentation of idiopathic intracranial hypertension

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Abstract

Idiopathic intracranial hypertension (IIH) is a disorder defined by clinical criteria that include signs and symptoms isolated to those produced by increased intracranial pressure (ICP; e. g., headache, papilledema, and vision loss), elevated ICP with normal cerebrospinal fluid (CSF) composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations. The most common signs in IIH are papilledema, visual field loss, and unilateral or bilateral sixth cranial nerve palsy. Here we report a case of IIH presenting as headache with vision loss, papilledema, complete ophthalmoplegia with proptosis in one eye, and sixth cranial nerve palsy in the other eye. Patient was managed with acetazolamide, topiramate, and diuretics. Symptoms remained static and she was planned for urgent CSF diversion procedure.

Key Words

Idiopathic intracranial hypertension, ophthalmoplegia, headache

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Introduction

Idiopathic intracranial hypertension (IIH) is a disorder defined by clinical criteria that include signs and symptoms isolated to those produced by increased intracranial pressure (ICP; e. g., headache, papilledema, and vision loss), elevated ICP with normal cerebrospinal fluid (CSF) composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations. While once called benign intracranial hypertension, to distinguish it from secondary intracranial hypertension produced by a neoplastic malignancy, it is not a benign disorder. Many patients suffer from intractable, disabling headaches; and there is a risk of severe, permanent vision loss. The most common signs in IIH are papilledema, visual field loss, and unilateral or bilateral sixth cranial nerve palsy. Here we report a case of IIH presenting as headache with vision loss, papilledema, complete ophthalmoplegia with proptosis in one eye, and sixth cranial nerve palsy in other eye.

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Case Report

A 22-year-old female presented with chief complaints of diminution of vision in both eyes from 8 days. Patient's symptoms started with diminution of vision, which was agradual onset involving the right eye. Her symptoms progressively worsened over time. There was no associated pain, watering, or redness of eye. Over the next 2 days, patient complained of vision loss in left eye; which was painless and progressive. Five days prior to this episode, patient had upper respiratory tract infection (fever, rhinorrhea, and sneezing) which resolved spontaneously.

There was no history of diurnal variation of symptoms, redness of eyes, floaters, tinnitus, nausea/vomiting, past history of any loss of vision, weakness of any side of body, dysphagia, ataxia, dyspnea, fever, loss of consciousness, intake of vitamin A, steroids, tetracyclines, oral contraceptive pills (OCP), arthralgias, or photo sensitivity. Patient had a history of headache from last 2 years which was unilateral, occipital in location associated with nausea, photophobia aggravated by fasting, and relieved by self-administered analgesics.

On examination; patient was conscious; cooperative; and oriented to time, place, and person. Pulse was 78/min regular, blood pressure (BP) was 110/70 mmHg, and body mass index (BMI) was 25 kg/m^2. CNS examination revealed normal higher

mental functions. Visual acuity in right eye was finger counting at 1 meter and left eye was finger counting at 3 meters. There was constriction of peripheral field of vision and color vision was impaired bilaterally. Fundus examination revealed bilateral papilledema with hemorrhage on left side at 2 O'clock position and mild disc pallor on right side. In right eye, there was restricted painful lateral gaze; while initially in left eye, extra ocular movements were normal. Rest of neurological examination was normal.

During hospital stay, her symptoms worsened. She developed pain on attempted eye movements and pressure over eyes with bilateral ptosis. Visual acuity in right eye decreased to perception of light and in left eye to finger counting at 1 meter. Extraocular movements in right eye were restricted in all directions (third, fourth, and sixth cranial nerve palsies) and in left eye there was sixth cranial nerve palsy [Figure 1]. Pupils were dilated and not reacting to light. The seventh cranial nerve was normal. Her hemogram, kidney function tests (KFT), liver function tests (LFT), and serum electrolytes were normal. Magnetic resonance imaging (MRI) brain revealed that bilateraloptic nerves were tortuous and showed increased perineural CSF spaces. There was mild flattening of posterior globe of sclera bilaterally [Figures 2-4]. MR venography (MRV) of brain was normal. CSF was grossly clear, with no cells, protein of 12 mg/dl, and sugar of 98 mg/dl. However, the opening pressure of CSF was 300 mm water. Her X-ray chest, echocardiogram (ECHO), antinuclear antibody (ANA), double stranded deoxyribonucleic acid (dsDNA), thyroxine (T4), thyroid stimulating hormone (TSH), cytoplasmic antineutrophil cytoplasmic antibody (cANCA), perinuclear ANCA (pANCA), neostigmine test, and hormonal profile were normal. On the basis of clinical feature and lab investigations, a diagnosis of IIH was made. Patient was managed with acetazolamide, topiramate, and diuretics. Lumbar puncture was done twice in 1 week and about 20 ml of CSF was drained each time. After CSF tap and starting medicines, patient's symptoms remained static. She was planned for urgent CSF diversion procedure.

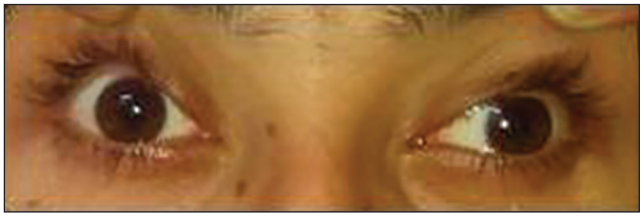

Figure 1: Complete ophthalmoplegia in right eye

Discussion

IIH is a disorder characterized by increased ICP of unknown cause is predominantly seen in women of childbearing age and associated with a history of recent weight gain. The incidence is approximately 1/100,000/year rising to 13/100,000/year in women between 20 and 44 years who are 10% above ideal body weight and 19/100,000/ year in those 20% above ideal body weight.[1-4] The first report of IIH was by the German physician Heinrich Quincke, who described it in 1893 under the name serous meningitis. The concept of raised ICP in the absence of a space-occupying lesion was first introduced by Max Nonneas 'pseudo tumour cerebri' (PTC) in 1904. Later the term 'benign intracranial hypertension' became popular and was often used interchangeably with PTC. The condition was considered 'benign' in comparison with cases of tumor, but it has been

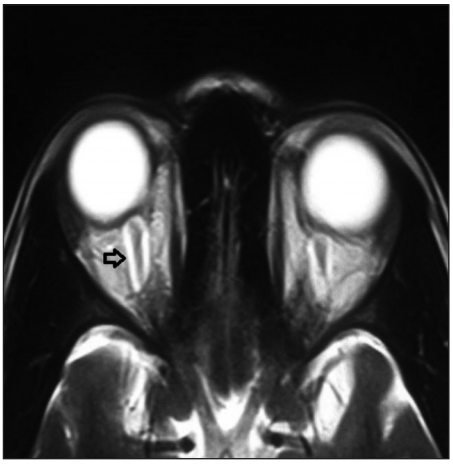

Figure 2: MRI showing optic nerves with increased perineural CSF spaces. MRI = Magnetic resonance imaging, CSF = Cerebrospinal fluid

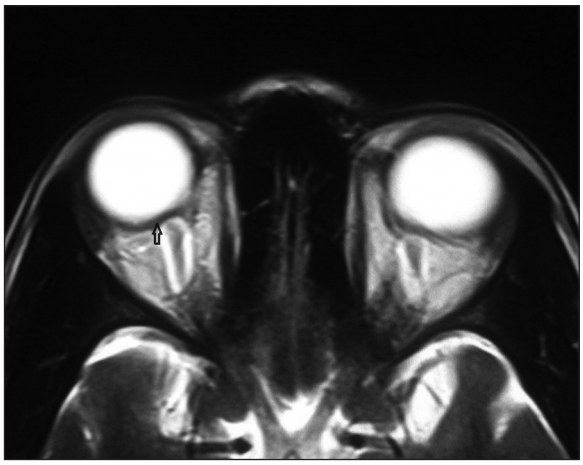

Figure 3: MRI showing flattening of posterior globe

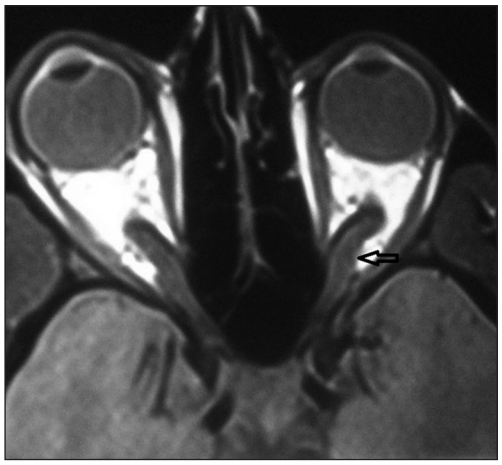

Figure 4: MRI showing tortuous optic nerves

argued that loss of visual function in up to 25% of cases and progression to blindness; if untreated means that it should not be considered 'benign'. Hence, the term IIH is now generally used. Its diagnosis is done using Modified Dandy criteria.[5]

Headaches are recorded in almost all IIH patients. They are typically nonspecific and vary in type, location, and frequency. The pain is generally described as being diffuse, worsening in the morning, and being exacerbated by the Valsalva maneuver. Patients who present with double vision, most frequently complain of horizontal displacement of the images. Horizontal diplopia is a symptom of false-localizing sixth cranial nerve palsy Vertical diplopia is rare, but it has been reported. Pulsatile tinnitus may be reported. This is a rhythmic sound, heard in one or both ears, with a pulsing synchronous rhythm that may be exacerbated by the supine or bending position. Radicular pain (usually in the arms) is an uncommon symptom.

IIH with total ophthalmoplegia is a rare presentation of IIH. Only few case reports have been published in the literature. Landan *et al*., have reported in 1987a case of IIH with complete bilateraloph thalmoplegia who recovered after optic nerve fenestration surgery.[6] Kidron *et al*., have reported a case of young female of IIH who presented with rapid visual loss and ophthalmoplegia and was managed with lumboperitoneal shunt.[7]

Here we report a case of IIH who presented with decreased vision and later developed complete ophthalmoplegia in one eye and sixth cranial nerve palsy in another. The reason behind complete ophthalmoplegia is not known, but is likely due to transmission of raised interferential current therapy (ICT) to the nerves. These patients need urgent management in order to preserve their vision. Many such patients need surgical management in the form of CSF diversion procedure like ventroperitoneal shunt or optic nerve fenestration, but till then they have to be managed with medical therapy consisting of carbonic anhydrase inhibitors and steroids. If vision is threatened, optic sheath fenestration is probably the best course; especially as a unilateral procedure often has effect on vision in both eyes. It consists of partial unroofing of the orbit and intraorbital incision of the dural–arachnoid sheaths surrounding the optic nerve. However, as this procedure carries a moderately high risk of vascular obstruction and unilateral visualloss, many now recommend a lumboperitoneal shunt as the first approach. In order to determine which procedure is better, clinical trials comparing the two procedures will be needed, which seem unlikely at present due to ethical reasons. Till we get more evidence about the better procedure among these two, it will be the clinical judgment of attending doctors that will help in deciding the treatment course of individual patients.

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