Fulminant Idiopathic Intracranial Hypertension with Atypical Presentation

Dear Editor,

Idiopathic Intracranial Hypertension (IIH) is characterized by elevated intracranial pressure in the absence of a structural, parenchymal, or Cerebrospinal fluid (CSF) abnormality and absence of focal neurologic deficits with the exception of cranial neuropathy. Headache, transient visual obscurations, and pulsatile tinnitus are some of the typical features of IIH. Cranial nerve deficits most often in the form of sixth nerve palsy are seen in a minority of cases of IIH as false localizing signs. Many atypical presentations of IIH have been reported in the literature. We present a unique case of fulminant IIH complicated by complete ophthalmoplegia, blindness, and polyradiculopathy.

A 23-year-old woman with no known co-morbidities, presented with severe headache associated with nausea and vomiting of one month duration. She also complained of double vision, gradual decrease in vision, and neck pain, radiating to the medial three fingers of both upper limbs of 2 weeks duration. There was no history of fever, altered sensorium, or seizures. There was no history of intake of Oral contraceptives (OCPs) or other drugs. The patient was in the overweight range (BMI-27.50).

On examination, higher mental functions were normal. Cranial nerve examination revealed diminished visual acuity restricted to hand movements bilaterally along with loss of color vision. Visual fields could not be tested because of severely diminished visual acuity. Fundus examination showed bilateral established papilledema. Extra Ocular Movements were completely restricted in all directions. Other cranial nerves were normal.

Motor examination revealed normal bulk, tone, and power. Deep tendon reflexes were absent in all four limbs and the plantar reflex was flexor bilaterally. Sensory and cerebellar examination were normal. Kernig's sign was negative.

Basic hematologic workup and biochemical parameters were normal. MRI brain [Figure 1] showed tortuous optic nerves and prominent peri optic CSF space without any focal lesions. MR venogram was normal. MRI Spine done was normal. A contrast study was not done. A nerve conduction study revealed normal Compound muscle action potential (CMAPs) and SNAPs with absent F waves in the bilateral median, ulnar, tibial, and peroneal nerves.

She underwent an immediate lumbar puncture. CSF opening pressure was measured using the bedside method with an IV infusion set connected to an Lumbar puncture (LP) needle and was found to be grossly elevated (>60 cm of H_2O). CSF analysis revealed normal protein levels and normal sugar levels and was acellular. CSF CBNAAT was negative. The

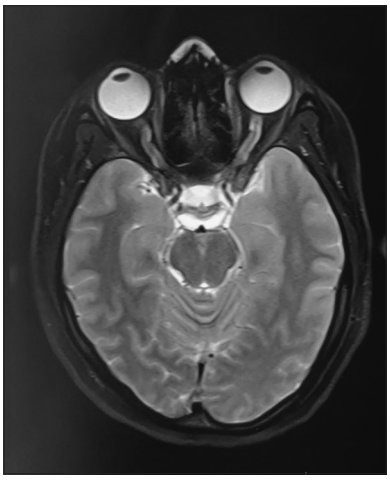

Figure 1: MRI brain (plain) axial images showing tortuous optic nerves with prominent perioptic CSF space

patient reported a marked reduction in the severity of headache following lumbar puncture. She was diagnosed with fulminant IIH and underwent Emergency Theco-Peritoneal (TP) shunting. Immediately after the procedure, her extraocular movements (EOM) improved significantly.

The vision improved gradually in her right eye to 20/70 but in the left eye, only hand movements could be perceived. Fundus examination on the 10th day following surgery revealed a normal optic disk on the right eye and the left eye revealed disk pallor s/o early optic atrophy [Figure 2]. OCT (RNFL) could not be done. All EOM in both eyes became normal. Deep tendon reflexes reappeared and repeat Nerve conduction study (NCS) showed a return of normal F-wave responses. She did not come for follow-up over the next 3 months and later presented with acute onset headache associated with worsening of vision and radiating pain in both upper limbs without any weakness. Examination revealed bilateral papilledema with optic atrophy. On evaluation, the TP shunt was found to have migrated away from the thecal space [Figure 3]. She underwent endoscopic assisted Ventricular-Peritoneal shunting following which headache and pain in the upper limbs subsided with improvement in vision. She was advised Tab. Acetazolamide 1 gm/day. She did not complain of headaches after the procedure.

Diagnostic criteria for IIH include symptoms and signs of intracranial hypertension with normal neurological examination excepting cranial nerve palsy (most frequently sixth cranial nerves) and papilledema along with the absence of structural lesions in neuroimaging and normal CSF constituents.[1,2] Diagnostic criteria for fulminant idiopathic intracranial hypertension include the criteria fulfilling IIH

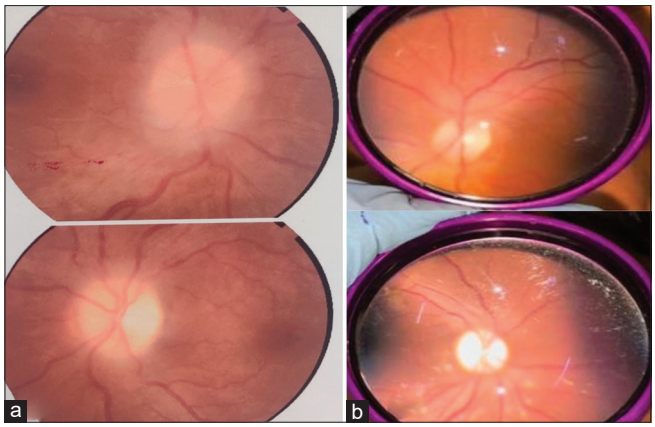

Figure 2: 2a-Fundus before surgery. OD: disk edema with hyperemia, dilated tortuous vessels, OS: disk edema with temporal pallor and dilated tortuous blood vessels. 2b-Fundus on 10th day post TP shunt. OD: normal disk, OS: disk pallor suggestive of early optic atrophy. OD=Once daily

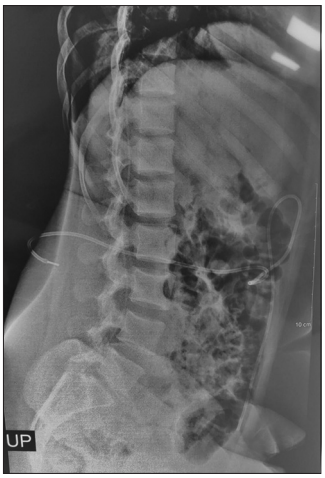

Figure 3: X-ray Lumbosacral (LS) spine lateral view revealing migration of thecal end of TP shunt

along with duration being <4 weeks between symptom onset and severe loss of visual acuity or field and rapid worsening of vision over days.[2]

Many atypical presentations of IIH have been reported in the literature with multiple cranial nerve palsies in varied frequencies.[3] The mechanism by which cranial neuropathies develop in IIH is unknown; however, it is thought to be due to intracranial pressure imposing traction forces on cranial nerves.[3-5] There have been a few cases of total ophthalmoparesis in IIH patients with high opening pressure that resolved following shunting similar to our case.[6]

Typically, visual acuity is rarely affected or affected late in the course. Patients with IIH usually have insidious and progressive vision loss following long-standing papilledema. Normally, autoregulation mechanisms maintain perfusion in the patients with IIH despite papilloedema. However, sudden elevations in Intracranial pressure (ICP) may occur to an extent that the ocular perfusion pressure at the optic disk is impaired before vascular regulation can cope and adjust resulting in acute vision loss.[7] According to Thambisetty *et al*.,[6] axoplasmic stasis and optic nerve ischemia following the sudden rise in ICP are most likely the cause for the severe, sudden, and rapidly progressing visual loss.

Polyradiculopathy was considered to be a false localizing symptom equivalent to sixth cranial nerve palsy in rare cases of severe intracranial hypertension (e.g., IIH and Cerebral Venous Sinus Thrombosis). The mechanical compression of nerve roots caused by high CSF pressure distending the subarachnoid space was hypothesized as the mechanism of radicular involvement in the case of raised ICP.[5,8] Groves *et al*. (1999)[9] proposed that the nerve roots, dura nerve junction, and dorsal root ganglia are surrounded by bony neural foramen and that these neural structures are potentially sensitive and susceptible to the effects of reduced venous outflow and edema caused by raised ICP. Parike and Watanabe (1985)[10] proposed that ischemia of the nerve roots produced by reduced venous outflow through the thin-walled radicular veins may play a role in the etiology of polyradiculopathy as these veins lack tunica media and are easily compressible by raised ICP.

In our patient, the restoration of normal F waves following TP shunting supports a cause-and-effect relationship between intracranial hypertension and polyradiculopathy. Further, the worsening of symptoms with TP shunt failure and recovery following Ventriculoperitoneal shunting strongly confirms the association.

Delay in diagnosis of fulminant IIH causes profound or irreversible vision loss.[9] Timely diagnosis of IIH patients presenting with these features is important as a timely intervention may prevent permanent visual disability. Studies reported fulminant vision loss in 2–3% of patients with IIH.

To conclude, we propose that, an under-recognized syndrome of polyradiculopathy with absent or prolonged F□waves may result along with complete ophthalmoplegia and blindness in particularly severe cases of intracranial hypertension. CSF shunting may provide a better prognosis than conservative management, particularly with regard to the preservation of vision. Rapid recognition of fulminant IIH is of critical importance as delay in definitive surgical intervention frequently results in permanent and profound visual disability.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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