A tale of two discs- normal to blurry, and the diagnostic dilemma! How to proceed?

A 25-year-old female presented with sudden-onset diminution of vision in OS for 3 days' duration. On examination, best corrected visual acuity (BCVA) in OD was 20/20 and in OS was finger counting (FC), OS showed grade II relative afferent pupillary defect (RAPD) and red-green color deficit, and OU had normal optic discs [Fig. 1a and b]. Magnetic resonance imaging (MRI) brain was normal. MRI of OS optic nerve showed hyperintensity in T2 phase [Fig. 2a]. Cerebrospinal fluid (CSF) analysis showed normal contents and opening pressure and negative result for neuromyelitis optica (NMO) antibodies. Visual evoked potential (VEP) showed prolonged OS P100 latency. She was diagnosed with OS retrobulbar neuritis and treated with intravenous methylprednisolone followed by oral prednisolone (Optic neuritis treatment trial (ONTT) regimen).[1] BCVA improved to 20/20 OS at 3 months of follow-up. There was no further recurrence of the symptoms until 1 year later when she presented again with bilateral blurring of vision associated with headache for 1 week duration. BCVA was 20/25 OU; pupillary reactions were sluggish in OU. Fundus examination OU revealed disc edema with blurred margins (OD >> OS), peripapillary edema, and engorged and tortuous retinal veins. Disc in OD was hyperemic, while disc in OS showed mild temporal pallor [Fig. 1c and d]. Differential diagnoses included bilateral recurrent papillitis, papilledema, and idiopathic intracranial hypertension (IIH).

What is the next best course of action at this stage?

A. Treat as recurrent bilateral optic neuritis
B. VEP and CSF analysis
C. MRI brain and orbit, MR venogram, and CSF analysis
D. Blood pressure and CSF analysis.

Findings

MRI brain and orbit revealed the following:

- mild atrophy with minimal enhancement of the intraorbital portion of the left optic nerve suggestive of post-optic neuritis sequelae [Fig. 2b];
- partial empty sella with distension of the subarachnoid space around optic nerve (>2 mm) on both sides suggestive of IIH [Fig 2c and d]; and
- MR venogram ruled out cavernous sinus thrombosis and deep vein thrombosis.

CSF analysis showed raised opening pressure (40 mmHg), normal protein (55 mg/dl), normal glucose (66 mg/dl) and chloride (128 mEq/dl), and was microscopically acellular.

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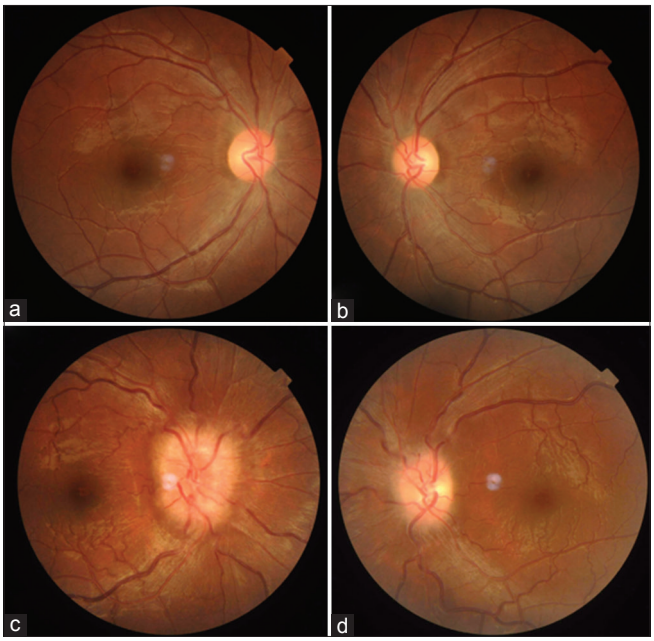

Figure 1: During the first episode: (a and b) OU normal optic discs with no evidence of disc edema, hyperemia, or pallor. During the second episode: (c) OD disc hyperemia, blurred margins, cup obliterated, peripapillary edema, engorged and tortuous retinal veins; (d) OS disc edema with blurred margins, obliterated cup, mild peripapillary edema, temporal disc pallor, engorged and tortuous retinal veins

Blood pressure (BP) was 122/76 mmHg.

VEP showed prolonged p100 latency on the left side.

Based on the above evidences, a diagnosis of OU IIH and OS post-optic neuritis sequelae was established.

She was prescribed oral acetazolamide 250 mg three times daily. On the last follow-up visit 3 months later, BCVA improved to 20/20 OU. Fundus examination revealed resolved disc edema in OU, normal disc color in OD, and temporal disc pallor in OS.

Diagnosis: OU IIH with OS post-optic neuritis sequelae.

Correct answer: C.

Discussion

Optic neuritis (ON) can be associated with multiple sclerosis (MS) or NMO or can occur independently as idiopathic optic neuritis (ION).[2] In the current case, neurological

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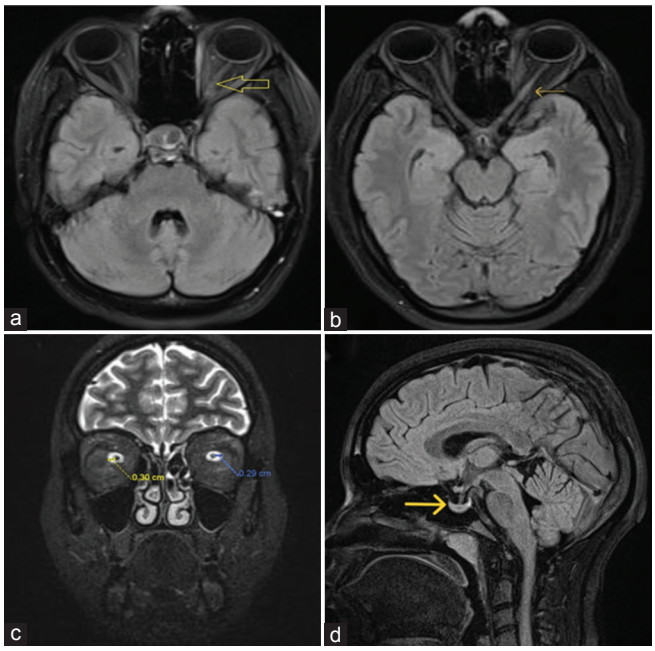

Figure 2: MRI. During the first episode: (a) OS hyperintensity in T2 phase (arrow) in the retrobulbar intraorbital segment of the optic nerve. Repeat MRI during the second episode: (b) OS mild atrophy with minimal enhancement of the intraorbital portion of the optic nerve (arrow) suggestive of post-optic neuritis sequelae. (c) Bilateral enlarged optic nerve subarachnoid space (>2 mm) suggestive of IIH and (d) partial empty sella (arrow) with concave shape of the pituitary gland suggestive of IIH. IIH = idiopathic intracranial hypertension, MRI = magnetic resonance imaging

evaluation and MRI ruled out MS and NMO during the first episode. While prognosis of ON is generally good, the probability of recurrence after the first episode is relatively high – the 5- and 10-year risks of recurrence are 28% and 35%, respectively.[1] Unilateral optic nerve involvement at the initial stage of the disease also increases the risk of ION recurrence.[2] In another study, the estimated cumulative incidence of ON recurrence in either eye was 26% at 1 year, 33% at 3 years, 37% at 5 years, and 50% at 10 years after the first episode of ON. The recurrence rate was highest within 1 year after the episode.[3]

In the current case, the patient presented with bilateral disc edema 1 year after an initial episode of unilateral retrobulbar neuritis. Therefore, considering the past history of OS retrobulbar neuritis and the associated high risk of recurrence, bilateral recurrent ON was the initial differential diagnosis during the second episode. However, presence of headache, fairly good vision in both eyes, and disc edema with engorged retinal veins hinted toward other mimickers like papilledema and IIH, thereby leading to a diagnostic dilemma. This necessitated further evaluation including brain imaging and lumbar puncture. Repeat MRI, MR venogram, and CSF analysis established a diagnosis of IIH, while ruling out bilateral ON and papilledema. High index of suspicion is, thus, needed in such a case to diagnose IIH. IIH is typically seen in young obese women of the reproductive age group. Rarely, IIH can also be seen in young nonobese women as seen in this case.[4]

Statement of ethics

Written informed consent for publication (including the images) has been obtained from the parent of the patient. All procedures carried out were in accordance with the tenets of the Declaration of Helsinki. Institute Ethics Committee approval is not required for a case report according to the Indian Council of Medical Research guidelines.

Ethics approval

The study was approved by the Institute Ethics Committee and was conducted in accordance with the ethical standards of 1964 Helsinki declaration and its later amendments.

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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