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Case Report

Bilateral ophthalmoplegia in a child with migraine

Rachana Dubey, Biswaroop Chakrabarty, Lokesh Saini, Priyanka Madaan, Sheffali Gulati *

Division of Child Neurology, Department of Pediatrics, All India Institute of Medical Sciences, New Delhi 110029, India

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Abstract

Background: In children, migraine with or without aura is a common entity, however variants like recurrent painful optic neuropathy (RPON) is rarely encountered.

Case result: A 9 year old boy presented with headache for 1 week and restricted movements and drooping in both eyes for last 3 days. On examination he had bilateral ophthalmoplegia and ptosis. History of migrainous headache was present in the patient as well as his mother. His MRI brain with venogram, serum autoimmune markers, serum and urine toxicology screen and repetitive nerve stimulation test were normal. He received intravenous pulse followed by oral steroids for 6 weeks and was started on antimigraine prophylaxis. Eighteen months since the attack, he has improved completely with mild asymmetric mydriasis persisting.

Discussion and conclusion: This may represent first attack of RPON in a child with migraine. Rarely this may herald the onset of migraine as well, index of suspicion should be high as it is a diagnosis of exclusion and a treatable entity.

Keywords: Migraine; Ophthalmoplegia; RPON; Steroids; Treatable

1. Introduction

Migraine, with and/or without aura are common in children, but variants like recurrent painful optic neuropathy (RPON) are exceedingly rare [1]. RPON has been defined as repeated attacks of paresis of one or more ocular cranial nerves (commonly the third), with ipsilateral headache [2]. A case of bilateral ophthalmoplegia in a child is presented here which represents the first attack of RPON.

2. Case summary

A premorbidly normal, 9-year-old boy, presented with throbbing, nonlateralising diffuse headache of 1 week duration along with vomiting, double vision and restricted eye movements for last 3 days. The headache increased in severity within 2–3 days, was bifrontal in location and responded partially to analgesics. The diplopia was noted equally in all directions. The restriction of eye movement started unilaterally toward leftward abduction and upgaze, which progressed within 2 days to complete restriction of movement in all directions. Drooping of both eyelids was also noted for last two days.

There was no associated altered sensorium, speech abnormality, involvement of other cranial nerves, motor or sensory deficit, movement disorder or any feature of incoordination. There was no history of preceding febrile illness, animal bite or vaccination. For last two years, he is having recurrent episodes of hemicranial throbbing headache of either side with associated nausea, vomiting, photophobia and phonophobia getting

* Corresponding author. Tel.: +91 9868397532.
E-mail address: sheffaligulati@gmail.com (S. Gulati).

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relieved by rest and analgesics within 6–8 h without any historical evidence of pupillary abnormality. There was history of migraine without aura in his mother since her adolescence.

Salient features on clinical examination were stable vitals, normal fundus, bilateral ophthalmoplegia (bilateral pupils were 5 mm, regular, with no response to light or accommodation, absent vestibulo-ocular reflex, Fig. 1A) and mild ptosis. However, he had transient sinus bradycardia at admission signifying underlying autonomic dysfunction in a child with migraine. His higher mental functions, other cranial nerves, motor, sensory and cerebellar system were normal. There were no meningeal signs.

With this clinical background, differentials considered were RPON, bilateral cavernous sinus thrombosis, acquired demyelination, Tolosa-Hunt syndrome, neurosarcoidosis, Lymes disease and brainstem encephalitis. MRI brain with contrast with optic nerve cuts (Fig. 2) and magnetic resonance venography (MRV) were normal. Hematological and biochemical parameters were normal. Cerebropinal fluid (CSF) showed elevated proteins (116 mg %) without pleocytosis. CSF IgG index was normal and oligoclonal bands were negative. Serum levels of angiotensin-converting enzyme (ACE), anti-TPO (thyroid peroxidase) antibodies, antinuclear antibody (ANA) and anti-double stranded DNA were normal. Serology for Lyme disease, urine for atropine and organophosphates were negative. Electroencephalography (EEG), repetitive nerve stimulation test (RNST) and Autonomic function tests were also normal.

After excluding all other causes, a diagnosis of first attack of RPON was considered and intravenous methylprednisolone (30 mg/kg/day) was given for 5 days. Headache responded within 48 h of treatment. He was discharged on oral steroids (2 mg/kg/day for 2 weeks followed by tapering over 4 weeks) and flunarizine (5 mg/day). Ophthalmoparesis improved gradually; ptosis improved within two weeks and bilateral extraocular movements recovered completely in 6 weeks. Eighteen months since the illness, mydriasis is still persisting, although resting pupillary size and response to light and accommodation has improved (Fig. 1B and C). During this period he had one episode of hemicranial headache without ophthalmoparesis. Currently he is on topiramate (4 mg/kg/day) as migraine prophylaxis; flunarizine was withdrawn after 6 months due to excessive weight gain.

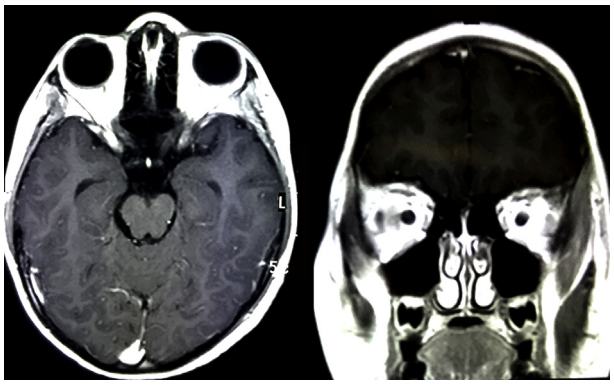

Fig. 2. MRI brain T1 weighted axial and coronal contrast images showing no enhancement of the bilateral optic nerves and other cranial nerve nuclei.

3. Discussion

The two notable underlying mechanisms for pathogenesis of RPON are ischemia and inflammation and demyelination. The ischemic pathway is mediated by swelling of the walls of the carotid or basilar arteries leading to occlusion of the smaller arteries supplying the cranial nerves. Internal ophthalmoplegia can be caused by ischemia in the Perlia and Edinger–Westphal nuclei [3]. According to the theory of inflammation and demyelination, it has been postulated that at the onset of a migrainous attack, the ophthalmic branch of the trigeminal nerve may cause release of neuropeptides into the circle of Willis. This results in sterile inflammation leading to demyelination which is also responsible for contrast enhancement of cranial nerves on imaging, elevated CSF protein and high IgG index [4]. In the current case, although there was no contrast enhancement of the cranial nerves on MRI, there was evidence of sterile inflammation in the form of raised CSF protein and globulins.

In children with painful ophthalmoplegia, the diagnosis of RPON should only be considered after ruling

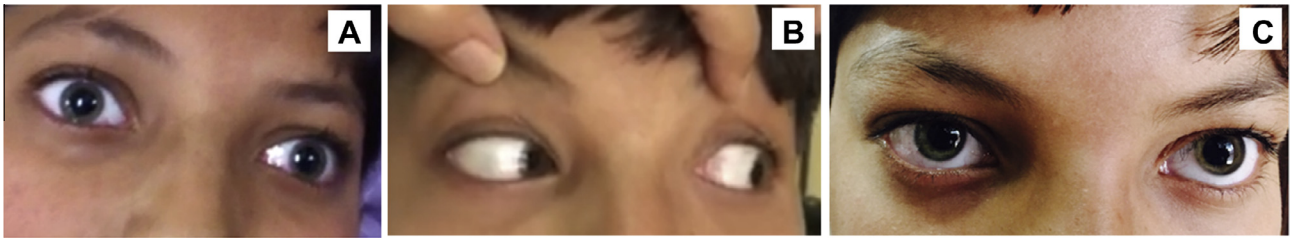

Fig. 1. (A) Bilateral complete ophthalmoplegia at presentation. (B) Complete improvement at 18 months in terms of range of ocular movements. (C) Improved but residual mydriasis persisting at 18 months.