Recurrent Painful Ophthalmoplegic Neuropathy: Report of the Two New Pediatric Cases

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INTRODUCTION

Recurrent painful ophthalmoplegic neuropathy (RPON), previously known as ophthalmoplegic migraine, is a rare syndrome characterized by repeating attacks of one or more ocular cranial nerve palsies with an ipsilateral headache, in which secondary causes have been excluded.(1) There are few publications in the literature up to now (2,3), and here we aimed to present two pediatric patients with RPON according to the ICHD-3 criteria and emphasize the diagnostic significance of the loss of enhancement or lesion during the symptom free period and discuss valproate as a useful prophylactic alternative.

Case 1

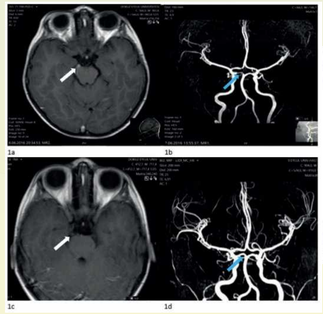
A four-year-old girl presented with a one week right-sided frontal throbbing headache, vomiting and photophobia which occurred every 1-2 months for the last year. Previous episodes of headache were unrelated to infections and had lasted for 2-3 days with vomiting 10-15 times a day. There was no family history of neurological disease except for a history of migraines in the patient's aunt. Neurological examination was normal. After 3 days, the patient presented with ptosis, ophthalmoparesis with impaired adduction and slowed pupillary light reflex in the right eye. Brain MRI revealed 5.5 mm contrast enhanced nodular lesion on the right at the level of the ambient cistern belonging to the cisternal part of the 3rd cranial nerve (Fig.1a). Magnetic resonance angiography (MRA) revealed reduced flow at P1 segment of the right posterior cerebral artery (PCA), moderate narrowing of the P1 segment posteriorly, compared to the left P1 segment, secondary to the lesion originating from the 3rd cranial nerve (Fig.1b). Two days later, the findings had spontaneous regression (normal light reflex, partial regression in the others). Propranolol prophylaxis was started and the patient was followed up with the prediagnoses of RPON and trigeminal nerve schwannoma. The patient had many further attacks and prophylaxis of propranolol, topiramate and flunarizine were used, respectively, at the maximum effective doses. After a 5 month attack-free period under flunarizine, the patient presented with a similar attack. Valproate was added to flunarizine. After 6 months without attacks under dual therapy, the valproate dose was gradually decreased. In the 41st month, when the last attack was 6 months ago, the patient was asymptomatic, the size of the lesion was 4 mm and there was no contrast enhancement on brain MRI (Fig.1c). Repeated MRA was normal (Fig.1d). Headache and ophthalmoplegia episodes regressed with methylprednisolone therapy used for 5-10 days without any sequelae. The patient has been symptom-free for 24 months.

Case 2

A previously healthy seven-year-old girl presented with a right-sided frontal headache persisting for the last 10 days, with the addition of limited inward ocular movement and double vision for the last day. The patient described previous similar headache episodes that occurred almost every day for 3-4 months, lasting for 1-4 hours. Neurological examination was normal except for limited inward gaze and ptosis in the ipsilateral eye. Brain MRI revealed a 4.5 mm, enhancing, nodular lesion in the 3rd cranial nerve lodge in the right perimesencephalic area (Fig. 2a). Brain CT angiography was performed to rule out vascular causes and was normal. Dexamethasone was initiated with pre-diagnoses of trigeminal nerve schwannoma and RPON. Headache regressed in 8 days and ophthalmoplegia in 6 days. Propranolol prophylaxis (1 mg/kg/day) was started. The patient had a second attack in the 9th month of the follow-up. Headache and ophthalmoplegia in this attack had the same characteristics as in the previous one. The patient received only analgesic treatment as the patient presented after the symptoms regressed. The propranolol dose was increased to 1.5 mg/kg/day. Brain MRI revealed similar lesion size and contrast enhancement. No lesions and enhancement in the cranial nerve lodge were observed in the brain MRI in the symptom-free period (Fig.2b). At the 13th month of follow-up, the patient had an attack and valproate was added to the propranolol prophylaxis. The patient is currently in the 18th month of the follow-up (5-month symptom free).

Conclusion

The changes in the naming and classification of RPON for nearly two decades indicate the ongoing debate around this rare disorder. A brain MRA during initial work-up and a brain MRI with contrast after a symptom-free period of at least 3 months may be helpful in both differential diagnosis and diagnostic challenges of RPON. A combined treatment approach with acute attack and prophylactic treatments can manage acute symptoms as well as minimizing recurrence for which valproate may be an effective prophylaxis option.



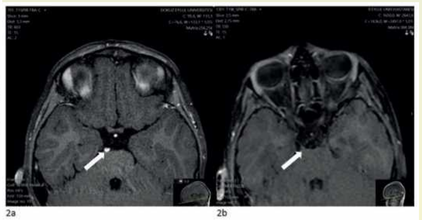


Fig. 1. (a) Axial postcontrast T1 images show thickening and increased enhancement 5.5 mm nodular lesion on the right at the cisternal portion of the 3rd cranial nerve. (b) Reduced flow at right P1 segment of posterior cerebral artery secondary to lesion. (c) After 6 months, the lesion size decreased to 4 mm and there was no enhancement on postcontrast T1 weighted image. (d) Repeated MR angiography study was normal.

Fig. 2. (a) Nodular lesion in the right 3rd cranial nerve at the perimesencephalic area show prominent contrast enhancement. (b) 12th month follow up study shows no lesion and no abnormal enhancement of the right oculomotor nerve at postcontrast T1 image.

References

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