Headache Medicine

Recurrent Painful Ophthalmoplegic Neuropathy: case series from a tertiary hospital and a systematic review

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Introduction

Recurrent Painful Ophthalmoplegic Neuropathy (RPON) is a rare condition, commonly observed in the pediatric population, in which individuals experience recurrent episodes of headache and ophthalmoplegia. The pathophysiology is not yet well understood, and its diagnosis relies on the patient's clinical history and the exclusion of other causes. A comprehensive evaluation is necessary to rule out underlying conditions. The treatment of RPON aims to address the acute condition and prevent the recurrence of attacks.

Objectives

To explore the clinical manifestations of RPON, to emphasize the importance of diagnosing, distinguishing it from other conditions, and to discuss treatment and management options.

Case Reports

CASE 1: A 47-year-old hypertensive female patient presents with a history of headaches since the age of 20, associated with bilateral ocular movement paresis during or after episodes of pain, with complete improvement within days or weeks. During outpatient follow-up, an extensive investigation was conducted with complementary examinations to exclude underlying conditions justifying the symptoms. Serial magnetic resonance imaging (MRI) scans revealed no alterations, except for the last MRI in 2022, which identified hyperintensity in the T2 STIR sequence in the bilateral lateral rectus muscles. Other exams showed no relevant changes. She began experiencing permanent sequelae from the age of 35, with no complete recovery of abduction paresis in both eyes between headache episodes. For the treatment of RPON episodes, oral corticosteroid therapy with prednisone for 7 days, in combination with symptomatic medication, is administered. Various prophylactic medications were initiated in this context, with no adequate response. Currently, the patient is using venlafaxine with adequate pain control. CASE 2: A 19-year-old healthy female began experiencing episodes of headache associated with ophthalmoplegia at the age of 8, with no family history of neurological diseases. The headache exhibited characteristics of migraine, accompanied by ipsilateral ophthalmoplegia. The patient's past medical history and neurological examination were unremarkable. A diagnostic workup showed no abnormalities. Currently, the patient experiences rare RPON episodes (1 episode per year) and occasional migraine without aura, without ophthalmoplegia. Good control of RPON episodes is achieved with oral prednisone for 7 days, non-steroidal anti-inflammatory drugs (NSAIDs), and sumatriptan. Prophylaxis for migraine without aura is administered with amitriptyline, yielding a favorable response with no observed side effects. CASE 3: A 10-year-old male, with an uneventful personal medical history but with refractive error requiring corrective lenses, began experiencing headache episodes at the age of 2. However, it was only at 6 years old that he was hospitalized due to a presentation of headache, ptosis, mydriasis, and right esotropia. At that time, an extensive diagnostic workup, including laboratory review, MRI, and enhanced MRI, revealed no noteworthy findings. The patient had rare annual episodes of headache associated with ipsilateral ophthalmoplegia. Prophylactic treatment for headaches with flunarizine and abortive treatment with dipyrone were initiated. There is no history of corticosteroid use in the acute phase of RPON, and the events were self-limited without progression to sequelae.

Conclusion

RPON is a rare and highly debilitating condition. Oral steroids may offer potential benefits in treating acute exacerbations. Reports of cases that are resistant to corticosteroids propose other medication alternatives for the acute condition, although their efficacy remains unproven. Case reports exploring headache prevention in RPON patients include first-line prophylactic agents for episodic migraine. Imaging has proven to be an excellent tool in aiding the diagnosis of RPON. Ultimately, RPON remains a clinical challenge.

Palavras-chave: Recurrent Painful Ophthalmoplegic Neuropathy;Ophthalmoplegic Migraine; Headache; RPON; Cranial Neuropathy.