Recurrent Painful Ophthalmoplegic Neuropathy:

migraine variant or inflammatory cranial neuropathy?

Ludovica Tundo[1], Laura La Volpe[1], Valentina Marinato[1], Massimo Miccoli[1], Chiara Casellato[2], Tiziana Zaccone[2], Alberto Priori[1,2], Chiara Manfredi[2]

1. Department of Health Sciences, Aldo Ravelli Center for Neurotechnology and Experimental Brain Therapeutics, University of Milan, Milan, Italy.
2. Clinical Neurology Unit, ASST Santi Paolo e Carlo, San Paolo University Hospital Milan, Milan, Italy.

INTRODUCTION

Recurrent Painful Ophthalmoplegic Neuropathy (RPON) is a rare disorder characterized by recurrent episodes of cranial nerve palsy associated with ipsilateral headache. Long considered a migraine variant with cranial nerve involvement, RPON is now classified as a rare cranial neuropathy according to the ICHD-3. However, its classification remains controversial, reflecting challenges in its differential diagnosis and pathogenesis. We describe a case of RPON in a young adult with a history of migraine, which contributes to our understanding of this poorly recognized condition.

CASE PRESENTATION

A 26-yo woman with a history of episodic migraine without aura presented to the E.R. with severe hemicranial headache, followed 48 hours later by complete left eye ptosis and deep, gravative orbital pain. The headache resolved within 48 hours, while the third nerve palsy persisted. The previous year, she had experienced a similar episode fully resolved after corticosteroids. On admission, isolated ptosis was observed. Within three days, partial oculomotor nerve palsy developed, with impaired adduction, elevation, depression and diplopia in multiple gaze directions. Pupils remained isochoric and reactive, with preserved convergence.

RESULTS

Blood tests, autoimmune screening and infectious workup, including lumbar puncture, were unremarkable.
Brain CT and CT angiography were normal.
A PET scan excluded vasculitis.
MRI revealed thickening and FLAIR hyperintensity of the left oculomotor nerve in the cisternal segment, without gadolinium enhancement.
The patient received oral corticosteroids, resulting in gradual improvement of ptosis and diplopia.
Given the clinical presentation, the relapsing course, MRI findings and exclusion of other causes, she was diagnosed with RPON and discharged on prophylactic therapy with pregabalin.

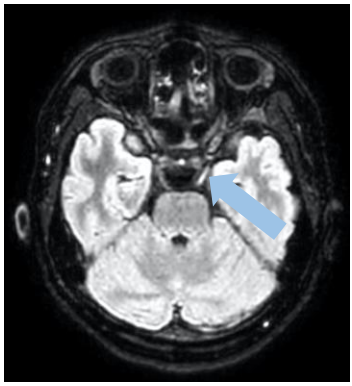

Fig.1 MRI T2 FLAIR hyperintensity of the left oculomotor nerve

DISCUSSION

RPON is a diagnosis of exclusion: neoplastic, vascular, infectious, inflammatory causes and Tolosa-Hunt syndrome must be ruled out. The clinical course and imaging findings in our case support the interpretation of RPON as a true cranial neuropathy rather than a migraine variant. The onset of oculomotor palsy after migraine resolution, its persistence beyond the pain phase, and the MRI evidence of nerve thickening and FLAIR hyperintensity, although without gadolinium enhancement, suggest a focal inflammatory process. Although some authors suggest a pathophysiologic connection between migraine attacks and recurrent cranial neuropathy, these should be considered as separate entities.

CONCLUSIONS

This case highlights the diagnostic complexity of RPON and its clinical overlap with migraine, reinforcing the hypothesis that RPON represents a distinct neuropathic entity rather than a migraine variant. Furthermore, it adds to the limited literature on RPON, aiding in its improved recognition among clinicians.

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