Alternating Involvement of Bilateral Oculomotor Nerves in Recurrent Ophthalmoplegic Cranial Neuropathy

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INTRODUCTION

Recurrent ophthalmoplegic cranial neuropathy (ROCN), also known as ophthalmoplegic migraine, is a rare disorder of episodic unilateral headache with reversible paresis of ipsilateral cranial nerves III, IV, and/or VI. Almost exclusively, laterality is consistent between recurrences\(^1\). We report a case of alternating laterality of headache and oculomotor paresis between ROCN episodes.

CASE PRESENTATION

A 59-year-old male presented with acute diplopia and right-sided ptosis following 13 days of severe throbbing right-sided headache with nausea and photophobia. Systemically, the patient was well and there was no antecedent trauma or illness. There were no other symptoms of GCA. Past medical history included migraine, but no diabetes, hypertension, smoking, or thyroid disease.

Neuro ophthalmic examination revealed partial third nerve palsy on the right (Fig. 1a). Pupils were normal. Examination of other cranial nerves was normal. The patient was admitted due to pain. Extensive laboratory investigations (Table 1) and neuroimaging (Fig. 2) were normal, aside from minimally elevated CSF protein. The headache resolved 3 weeks after onset.

The diplopia gradually improved until 6 weeks later when the patient re-presented with acute worsening of diplopia following 1 week of left-sided headache. There was a new left-sided pupil-sparing third nerve paresis and improved function of the right third nerve (Fig. 1b). Laboratory and imaging studies were stable. Based on International Classification of Headache Disorders criteria, ROCN was diagnosed (Table 2). The headache responded well to corticosteroid.

Four months later, ocular motility was significantly improved (Fig. 1c) and there has been no recurrent headache. Neuroimaging obtained 8 months after initial presentation remains normal (Fig. 3).

Table 1. Systemic investigations

<table>
<thead>
<tr>
<th>Bloodwork</th>
<th>CSF</th>
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<tbody>
<tr>
<td>ANC 5.5</td>
<td>LP</td>
</tr>
<tr>
<td>ESR 2</td>
<td>Cell count</td>
</tr>
<tr>
<td>Hb 13.0</td>
<td>Glucose</td>
</tr>
<tr>
<td>Hct 41</td>
<td></td>
</tr>
<tr>
<td>Platelets 250.0</td>
<td>Protein</td>
</tr>
<tr>
<td>Fibrinogen 2.2</td>
<td></td>
</tr>
<tr>
<td>INR 0.9</td>
<td>ESR</td>
</tr>
<tr>
<td>BMI 25.0</td>
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Table 2. Diagnostic Criteria: Recurrent Ophthalmoplegic Cranial Neuropathy

A. At least two attacks fulfilling criterion B
B. Both of the following
1. Unilateral headache
2. Ipsilateral paresis of one or both ocular motor nerves
C. Orbital, parasecondary or posterior fossa lesion has been excluded by appropriate investigation
D. Not better accounted for by another ICD-10 diagnosis.

DISCUSSION

ROCN is a diagnosis of exclusion. In this case, a broad differential for sequential painful CNII palsies was considered, including:

- Microvascular ischemia
- Inflammatory: Vasculitis, Tolosa-Hunt Syndrome
- Infectious: VZV, Lyme, syphilis
- Antibody mediated: myasthenia, thyroid disease, paraneoplastic
- Sarcoid

This case illustrates typical features of ROCN:

- Headache preceding cranial nerve palsy by several days
- Resolution of headache with corticosteroids
- Resolution of cranial nerve palsy weeks after headache
- Normal neuroimaging (findings more common in children\(^1\))

However, to our knowledge, this is only the second report of alternating laterality between episodes of ROCN, and the only reported case with normal imaging. Choi et al reported a case with alternating laterality of internal ophthalmoplegia with headache and gadolinium enhancement of bilateral oculomotor nerves\(^1\).

There remains debate whether ROCN is a primary headache disorder or a neuropathy. The quality of headache and normal imaging in the present case may support a migraine etiology.

REFERENCES

2. The International Classification of Headache Disorders, 3rd edition.