Headache Medicine

Raeder's Syndrome - A Threatening Diagnosis Not To Miss

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Introduction
Raeder's paratrigeminal oculosympathetic syndrome, also known as Horner painful syndrome, is a rare headache disorder listed in the chapter "Painful lesions of cranial nerver and other faciaal pain" of ICHD-3. The diagnostic criteria involves pain localized to the distribution of the ophthalmic division (V1) of the trigeminal nerve (that can spread to maxillary division - V2) aggrevated by eye movement associated with ipsilateral Horner syndrome with imaging evidence of anatomical pathology of middle cranial fossa or ipsilateral internal carotid. Even though his rare condition often goes undiagnosed, its timely identification can lead to correct identification and treatment of the associated cause and reduce the risk of other neurologic complications.

Objective
This case report aids in raising awareness about this relatively rare condition, adding insights into the clinical manifestations, potential causes of this specific type of headache.
We detail the clinical approach, treatment and outcome of a patient with Raeder's syndrome caused by internal carotid dissection - an acute and pottencially morbid cause of this syndrome - highliting the need for urgent investigation in such cases.

Case Report
A 44 years old male patient entered the Emergency Department reporting a new onset of a left periorbital headache (V1 and V2 territory), worst-ever, non-pulsatile, which reached the highest intensity in 10 minutes. He denied head trauma, eye redness, tearing and nasal discharge, but described visual blur in the affected eye.
The patient had a history of treated hepatitis C with chronic liver disease, cryoglobulinemia (mononeuritis multiplex, cutaneous vasculitis, livedo reticularis, arthritis - last activity 5 years ago), smoking, overweight and hypertension. At the moment, he used only losartan as a continuous use medication (treatment of crioglobulenemia ceased in 2018 after 2 doses of Rituximab and hepatitis C treatment).
On physical examination, he was hypertense (210x120mmHg), had a discreet anisocoria with right pupil 2mm bigger than the left one in light and dark environments, and semiptosis on the left eye. He denied reduced tactile sensitiveness over periorbital V1 and V2 territory on the affected side, but reported a tingling sensation when touched. There wasn't redness or tenderness of the left eye. There were no other focal neurology deficits.
The hypothesis of Paratrigeminal oculosympathetic (Raeder's) syndrome was made. Brain CT did not show any pathologic changes, but CT angiogram of the neck evidenced left internal carotid dissection, later confirmed to be acute by MR neck angiogram. Cerebral angiogram (CT and MRI) was considered normal.
The patient was treated with Dipirone, acetylsalicylic acid and clopidogrel (for 21 days) with pain relief, and was discharged from hospital one day later.

Conclusion
Raeder's paratrigeminal sympathetic syndrome may be caused by acute and threatening neurologic and vascular disorders, and its early diagnosis can reduce long term morbidity by allowing treating the causative disorder. Even though trigeminal pain is caused by V1 and/or V2 involvement, petrous and cavernous carotid dissection may be hard to see even with advanced imaging, highlighting the importance of cervical arterial imaging as well.
The treatment of choice of acute cervical artery dissection ranges from double platelet antiagregation to anticoagulation and must be decided based on individual patients informations and clinicians's experience. Treatment of pain lacks evidence support and therefore should be indivualised and based on patients' response.

Keywords: Raeder's Syndrome; Carotid Dissection; Facial Pain.