Introduction

Raeder’s syndrome (RS) consists of the combination of a postganglionic partial Horner’s syndrome (HS) (oculouvpathetic paralysis: ptosis and miosis) with facial pain localized in the distribution of the ophthalmic division of the fifth cranial nerve. RS differs from HS in that ipsilateral facial sweating is preserved, even though anhidrosis of a small ipsilateral forehead area can be present. Perisellar nerve involvement (cranial nerves III, IV, V, VI) represents a dangerous lesion in the anterior portion of the middle cranial fossa requiring appropriate investigations [1].

We present a patient with RS secondary to dolichoectasia of the left intracavernous internal carotid artery (ICA).

Case history

This 38-year-old man developed sudden, severe, nonthrottling but pressing pain. Headache was localized in the left retro- and peri-orbital areas. By the next day, the pain was less severe but persisted with fluctuating intensity. At the same time, he became aware of drooping of the left upper lid. Numbness was present in the distribution of the first division of the ipsilateral trigeminal nerve. The patient denied any nausea or vomiting. His past medical history was unremarkable except for smoking two packs of cigarettes for 17 years. Five days later, neurological examination revealed left miosis and ptosis. No tearing or redness of the eye was noted. The pupils reacted to light and accommodation. Fundoscopic examination was normal. Extraocular movements were full. Decreased function was noted in the ophthalmic division of the left fifth cranial nerve. Left corneal reflex was reduced. There was preservation of facial sweating. The remainder of his physical and neurological examinations was normal. Routine laboratory tests were normal. A brain computed tomography scan disclosed no abnormalities. Two weeks after the clinical picture started, brain magnetic resonance imaging (MRI) was performed and showed enlargement and dilation of the cavernous portion of the left carotid artery. The signal within the vessel was normal (Fig. 1). The gasserian ganglia and cavernous sinuses were of normal signal intensity and size. The brain

Abstract

A 38-year-old man presented with ptosis, miosis, facial pain and hypoesthesia in the ipsilateral ophthalmic division of the trigeminal nerve. Brain magnetic resonance imaging and magnetic resonance angiography showed fusiform dilation of the cavernous portion of the left carotid artery. A diagnosis of Raeder’s syndrome (RS) was made. Carbamazepine selectively relieved the facial pain but the partial Horner syndrome persisted. Our case adds dolichoectasia of the intracavernous internal carotid artery to the list of causes of RS.

Key words Raeder’s syndrome • Dolichoectasia • Cavernous sinuses • Internal carotid artery
was normal. Magnetic resonance angiography (MRA) confirmed a dilatation and a tortuous course of the intrapetrosus, intracavernous and supraclinoid portions of the left ICA (Fig. 2). Nevertheless, right ICA and extracranial carotid arteries were normal. A chest radiograph with apical views and a cervical spinal cord MRI study showed no structural lesions.

A diagnosis of RS associated with dolichoectasia of the left intracavernous ICA was made. The patient was treated symptomatically with carbamazepine with resolution of the headache. However, the partial HS persisted.

Discussion

In 1924, George Raeder published five cases of paratrigeminal oculosympathetic paralysis [2]. This clinical entity indicates pathology near the trigeminal ganglion. Many of the cases reported since then did not conform to the clinical picture reported by Raeder. Moreover, some authors included cluster headache patients in this group, increasing the confusion concerning this clinical entity [3]. Therefore, RS is an unclear diagnosis and there are many authors who do not believe in its existence as an independent syndrome. In fact, this syndrome has not been included in the latest Headache Classification of the International Headache Society (IHS) [4]. According to IHS criteria, a diagnosis of RS only represents a location of a non-specific pathological process in the middle fossa.

Boniuk and Schlezinger [5], in an attempt to clarify prognosis, pathogenesis and treatment, divided RS into two distinct types: Group 1, characterized by hemicrania, ipsilateral oculosympathetic paresis with parasellar cranial nerve involvement, or true RS; and Group 2, characterized by hemicrania and ipsilateral oculosympathetic paresis without parasellar cranial nerve involvement, also called “pericarotid syndrome”.

Our case fulfills Mokri’s criteria for the diagnosis of RS: incomplete HS and trigeminal dysfunction [6]. Therefore, it belongs to Group 1 since the patient has a single perisellar nerve disturbance. As our case shows, if ipsilateral parasellar nerve involvement is documented, a thorough neuroradiological investigation is indicated to rule out parasellar lesions. Furthermore, the combination of MRI and MRA allows confirmation of the diagnosis in the cavernous and paracavernous regions with high accuracy. By contrast, in pericarotid syndrome the cervical ICA needs to be studied.

The ocular sympathetic paralysis is due to a pericarotid lesion affecting the perivascular sympathetic fibers. These fibers, originated from third-order sympathetic neurons situated in the superior cervical ganglion, proceed to divide into two branches: (a) an extracranial sympathetic branch travels with the external carotid artery, responsible for facial sweating; and (b) the intracranial sympathetic branch travels with the ICA to form a plexus around the vessel. These

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**Fig. 1** Coronal T2-weighted MR image discloses an increase in the diameter of the left intracavernous internal carotid artery (arrow). The right intracavernous internal carotid artery shows normal size.

**Fig. 2** MRA images in anteroposterior projections. The left intracranial carotid artery has a tortuous course and a wide luminal diameter compared to the normal appearance of the right intracranial carotid artery.
fibers provide sympathetic innervation to the dilator of the pupil, the palpebral muscle of the upper lid, ciliary muscle, blood vessels of the globe and sweat glands of the forehead. Therefore, a lesion proximal to the bifurcation of the ICA results in HS, but a lesion distal to the bifurcation involving the ICA produces the characteristic miosis and ptosis without facial anhidrosis of RS.

Two reports have estimated the frequency of intracranial dolichoectasia to be 0.06% and 5.8% of all angiographic studies obtained for any reason [7, 8]. It is admittedly possible that the presence of the ectasia of the ICA in this patient was coincidental. We believe that the enlarged carotid artery caused the symptoms in the patient because we did not demonstrate a lesion in the sympathetic pathway in any other anatomical location [9]. Finally, the oculosympathetic paralysis was associated with a parasellar nerve injury.

We agree with Ford and Walsh in that sympathetic damage resulted from dilation of the ICA caused by recurrent edema and thickening of this vessel [10]. In our patient the trigeminal injury may also have been due to involvement of the artery of the inferior cavernous sinus which supplies the intracavernous cranial nerves [11]. Involvement of the intracavernous vessels may be attributed to compression, distortion of the carotid branches or thrombus formation in the dolichoectatic artery, causing occlusion of the origin of the small vessels that arise from it. Another feasible mechanism is the reduction of the blood flow by the dolichoectatic artery [12]. On the other hand, the direct stimulation by irritation, traction or pressure of trigeminal nerve fibers innervating the meninges near the cavernous sinus may be responsible for the referred pain to the ipsilateral orbital area [13].

The causes of RS are numerous and include abnormalities of the ICA, neoplasm, trauma or inflammatory process (sinusitis, basal arachnoiditis or neurosyphilis). The majorities of the vascular lesions are aneurysm or pathology of the cervical portion of the ICA. To our knowledge, only a few cases of isolated RS associated with intracranial ICA dilatation have been reported in the literature. Their etiopathogeneses include various conditions: extension of inflammatory disease such as sinusitis [14] or post-stenotic dilatation [15]. Although the etiology of cerebral ectasia is not well understood, we assume that the abnormality of the ICA was principally caused by congenital defects in the internal elastic lamina in the vessel wall [12].

In conclusion, our case adds dolichoectasia of the intracavernous ICA to the list of causes of RS.

Acknowledgements The authors are grateful to Julio Pascual for discussions and comments on the manuscript.

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