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Raeder Syndrome: MR Appearance

Mauricio Castillo¹ and Larry Kramer

Summary: A 40-year-old woman presented with symptoms compatible with Raeder syndrome. MR demonstrated narrowing of the left cavernous carotid artery. The high cervical portion of the left internal carotid artery was not identified. Angiography confirmed the narrowing of the cavernous carotid artery and showed marked and irregular stenosis of the distal cervical internal carotid artery. Involvement of the left sympathetic plexus and of ipsilateral cavernous sinus arteries is believed to have been the cause of the Raeder syndrome in this patient.

Index terms: Arteries, carotid; Nerves, trigeminal (V); Raeder syndrome; Migraine

Raeder syndrome (RS) is an unusual condition consisting of unilateral headache and facial pain in the distribution of the ipsilateral ophthalmic and maxillary divisions of the trigeminal nerve (1). An incomplete Horner syndrome is also usually present and is characterized by pupillary constriction, ptosis, and preservation of ipsilateral facial sweating. RS is believed to result from lesions involving the distal and cavernous internal carotid artery (ICA) (2). We present the magnetic resonance (MR) imaging and angiographic findings in one patient with RS.

Case Report

A 40-year-old woman presented with a 10-day history of left-sided headache, left ptosis, and left hemifacial pain. Past medical history was noncontributory. Physical examination revealed drooping of the left eye lid, conjunctival redness, and miosis on that same side. Pain and numbness was present in the distribution of the ophthalmic and maxillary divisions of the left trigeminal nerve. The remainder of the physical and laboratory evaluations were unremarkable.

MR was performed and showed severe narrowing of the left intracavernous internal carotid artery (ICA) (Figs. 1A). The left cavernous sinus was of normal signal intensity and size. The high cervical ICA on the left side was not identified (Fig. 1B). The right ICA and cavernous sinus were normal.

The gasserian ganglia were normal. The brain was normal. Conventional angiography was performed and showed marked and irregular narrowing of the left distal cervical ICA. Although the left cavernous carotid artery did not appear significantly narrowed on the angiogram, when compared to the right side, moderate to severe narrowing was present (Fig. 2). The right ICA showed multiple areas of narrowing, suggesting fibromuscular dysplasia.

The patient was treated symptomatically with analgesics and is doing well.

Discussion

RS was initially described in 1924 by a Norwegian neurologist (3). This condition is also known as the "paratrigeminal" syndrome. Clinically, it is characterized by unilateral migraine-like headache, pain in the distribution of the first (V1) and second (V2) divisions of the ipsilateral trigeminal nerve, and an incomplete Horner syndrome (miosis, ptosis, and preserved facial sweating) (1). The miosis can be of the retained type or be associated with absent pupilloconstriction reactions (2). RS may persist for years or may spontaneously disappear within several months (4).

RS can be clinically divided into two varieties, the migrainous or reflex type, and the symptomatic type with multiple parasellar cranial nerve involvement (4). Lesions involving the ipsilateral distal ICA are believed to be responsible for the majority of cases (1). Carotid artery dissections, aneurysms, pseudoaneurysms (related to biopsy, radiation therapy, and/or adjacent inflammatory processes such as sinusitis), and arteritis are the most common etiologies of RS (1–5). Rarely, glioblastomas of the temporal lobe and meningiomas of the Meckel cave may also cause RS (1, 6).

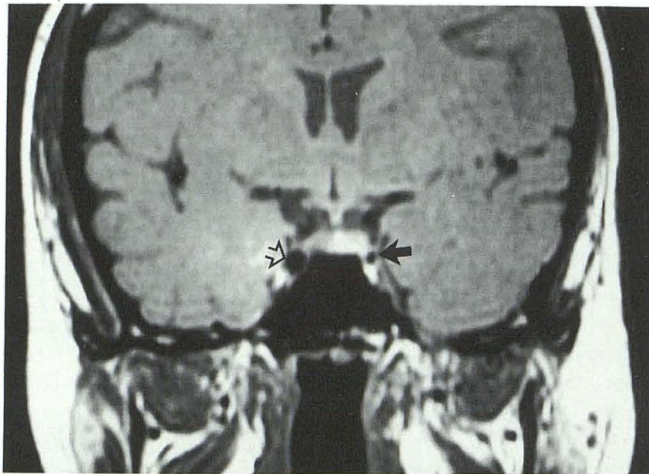
In RS, only the postganglionic segments of the fifth cranial nerve are involved (4). The artery to

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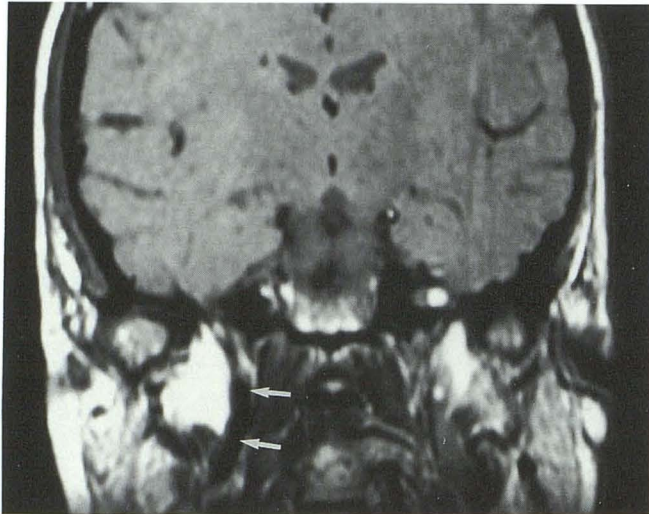
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A



B

Fig. 1. A, Noncontrast coronal MR T1-weighted image (800/25/4) (TR/TE/NEX) demonstrates normal right cavernous sinus and a normal size intracavernous right ICA (*open arrow*). Significant narrowing of the left intracavernous ICA (*arrow*) is present. The left cavernous sinus contains a small focus of high signal intensity that is believed to be related to an artifact secondary to slow flow or to normal fat. The right side of the pituitary gland is of slightly decreased intensity and probably related to partial volume averaging with the dorsum sellae since it was normal in all other sequences.

B, Noncontrast coronal MR T1-weighted image (800/25/4) slightly posterior to A shows flow void in the high portion of the right cervical ICA (*arrows*). The left ICA is not identified.

the inferior cavernous sinus (lateral main stem artery) is one of the most important dural branches of the cavernous ICA. This vessel supplies the intracavernous cranial nerves. The most important branch is the artery of the foramen rotundum that specifically supplies the maxillary division of the trigeminal nerve (7). Therefore, involvement of the cavernous carotid artery and

its branches may lead to ischemia of V1 and V2, giving rise to facial pain. We are not certain why the intracavernous portions of the third, fourth, and sixth cranial nerves are spared in patients with RS. The blood supply to the third (V3) division of the trigeminal nerve derives from smaller branches arising from the accessory middle meningeal artery that in itself is a branch of the internal maxillary artery. Also, the sympathetic fibers controlling facial sweating emerge between C6 and T4 and then accompany branches of the external carotid artery to the skin. Sparing of the external carotid artery explains normal sweating in the ipsilateral face, as well as the absence of pain in the distribution of V3. The portion of the sympathetic chain that innervates the pupillary dilator and Mueller muscles (paralysis of these muscles leads to miosis and ptosis, respectively) accompanies the high cervical ICA. Thus, involvement of this artery accounts for the incomplete Horner syndrome present in patients with RS (2).

Since the majority of the lesions responsible for RS are vascular in nature, angiography has been suggested as the imaging modality of choice. Because in many patients RS is a self-limited entity, several authors have recommended that angiography not be performed unless the symptoms last over 3 months (1, 8). In one small series, three patients with RS were

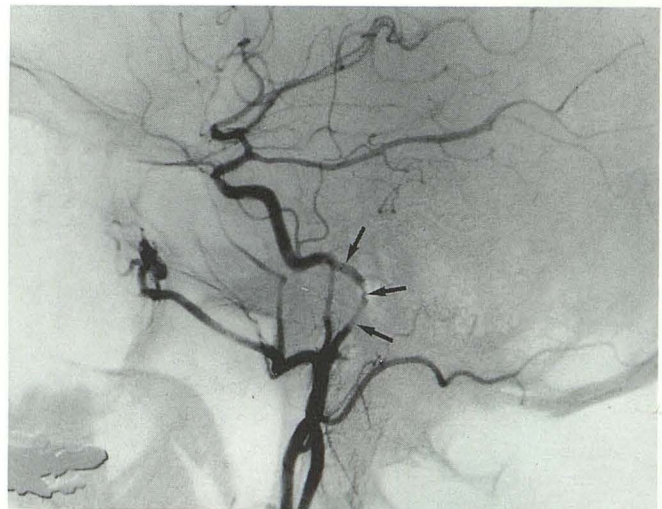


Fig. 2. Lateral view of the left common carotid artery injection shows marked narrowing (*arrows*) of the high ICA. Although not obvious in this view, there is mild narrowing of the left cavernous segment when compared to the right side (data not shown). Although the findings are nonspecific, the presence of fibromuscular dysplasias in the contralateral ICA (data not shown) makes the possibility of a dissection likely.

evaluated using angiography (1). In those cases, angiograms demonstrated fusiform dilatation of the cavernous ICA in one patient and aneurysms of the cavernous ICA in two patients. Most cases of RS usually require only symptomatic treatment; however, patients with severe pain may have to undergo surgical sympathectomy.

In our case, MR clearly showed marked narrowing of the left cavernous ICA (Fig. 1A). By MR, the left distal cervical ICA was not identified (Fig. 1B). Angiography showed findings suggestive of a previous dissection of the distal left ICA (Fig. 2). Although the etiology of this dissection is uncertain, it may have been related to the presence of underlying fibromuscular dysplasia. Findings characteristic of fibromuscular dysplasia were present in the right ICA. This disease is known to be bilateral in approximately 65% of cases (7). Moreover, spontaneous dissections may occur in up to 50% of patients with fibromuscular dysplasia (9). We also propose that diminished blood flow (leading to ischemia) through the left cavernous branches that supplied V1 and V2 gave rise to the clinical findings in our patient. Involvement of the distal ICA and preservation of the external carotid artery explains the presence of an incomplete Horner syndrome. No aneurysms were seen. We believe CT is rela-

tively insensitive in the evaluation of abnormalities involving the cavernous sinuses, and although our experience with this rare syndrome is limited to the case presented here, we suggest MR as the initial imaging modality in patients with RS. If the MR findings are indeterminate or suggest the presence of a cavernous ICA aneurysm, angiography should be obtained.

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