Hemicrania, Oculosympathetic Paresis, and Subcranial Carotid Aneurysm: Raeder's Paratrigeminal Syndrome (Group 2)

Case Report

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Hemicrania with ipsilateral oculosympathetic paresis in the absence of other neurological signs is usually regarded as a benign, self-limiting process which does not require angiographic investigation.1,8,10 We are reporting a patient with this symptom complex who also had a subcranial internal carotid aneurysm.

Case Report

A 36-year-old white woman was asymptomatic until July 8, 1967, when she noted the gradual onset of left facial pain which resolved on the following day. She then developed burning, tearing, and redness of the left eye and drooping of the left upper eyelid. The ptosis persisted but the other ocular complaints abated during the next few days. On July 10 she had a constant intense "sore throat" on the left side which was unaffected by swallowing. She was able to relieve the throat discomfort by applying her index finger over the left palato-pharyngeal arch and pushing laterally and posteriorly. She also had pain over the left cranial vertex. For an entire day she had episodes of momentary, sharp, shooting pains which radiated inward through the left ear canal. Left supraorbital pain then occurred. She denied any asymmetry of facial sweating.

The patient's past history was unremarkable except for high blood pressure since 1962. One week after the onset of her pains she was admitted to another hospital for angiographic studies. Daily doses of reserpine 0.1 mg, Hygroton 100 mg, and Vistaril 100 mg were prescribed. A left carotid arteriogram demonstrated an extracranial aneurysm arising from the left internal carotid artery near the base of the skull. A right carotid arteriogram, left vertebral arteriogram, and renal arteriogram showed no abnormalities.

The patient was transferred to the University of California Medical Center on August 21, 1967. On admission, her blood pressure was 160/100 and her pulse was 88. Visual acuity was 20/20 in both eyes. The left eye had ptosis and miosis. The right palpebral fissure measured 10 mm and the left, 8 mm. The right pupil measured 5 mm and the left 3 mm in a dimly lighted room. Extraocular movements were full. Corneal sensations were intact. The fundi showed slight arteriolar narrowing. Intraocular pressure was 6 with a 5.5 gm weight (Schiotz) in both eyes. The results of Schirmer's tear test were 5 mm in the right eye and 1 mm in the left eye. One drop of 10% cocaine in each eye failed to dilate either pupil when the patient was taking reserpine, but the right pupil became markedly dilated when the cocaine test was repeated 4 days after reserpine was discontinued. One drop of 1/1000 epinephrine failed to dilate either pupil. The remainder of the physical, neurological, and laboratory examinations were normal.

A second left carotid arteriogram confirmed the presence of a 5 mm subcranial internal carotid aneurysm extending medially and superiority at the base of the skull (Fig. 1). Surgical correction was not deemed advisable. The patient was discharged on September 10, 1967, essentially pain-free but with persisting left ptosis and miosis.

Discussion

Raeder* in 1924 reported five patients with somewhat different neurological symptoms and diagnoses; all five had Horner's syndrome without facial anhydrosis. Pain or numbness in the distribution of the ophthalmic branch of the trigeminal nerve accompanied the oculosympathetic paresis, which Raeder therefore described as "paratri-
Raeder's Paratrigeminal Syndrome

Fig. 1. Selective internal carotid arteriograms. Left: Lateral projection. The aneurysm (arrow) partly overlaps the distal extracranial portion of the internal carotid artery. Right: Oblique projection. The aneurysm (arrow) is easily seen.

geminal.” Some of his patients also had multiple cranial nerve palsies.

The designation “Raeder's paratrigeminal syndrome,” which has been the title of a number of subsequent publications, required clarification in the light of the varied clinical presentation of Raeder's original cases. Boniuk and Schlezinger consequently divided the syndrome into two types:

Group 1: Characterized by hemicrania, ipsilateral oculosympathetic paresis, and parasellar cranial nerve (III, IV, V, VI) involvement. These signs suggest a mass in the middle cranial fossa and warrant intensive diagnostic study.

Group 2 (in which our patient belongs): Characterized by hemicrania and ipsilateral oculosympathetic paresis but no other neurological signs. This variety of the syndrome is most frequently caused by vascular headache of the migraine type, and arteriography is felt to be unwarranted.

Boniuk and Schlezinger and Nano could not find a single verified Group 2 case in which an aneurysm was present. Our review confirms this except for possibly Sir Astley Cooper's case in 1836.

The patient we have presented with a Raeder's syndrome (Group 2) is unusual in that a subcranial internal carotid aneurysm was demonstrated angiographically.

Addendum

After submission of this paper, Law and Nelson (Neurology, Minneap. 1968, 18:43–46) reported a patient with a Group 2 Raeder's syndrome who had an ipsilateral aneurysm of the supracavernous portion of the internal carotid artery. Ambler, et al. (Neurochirurgia, 1967, 10:169–175), in a thorough review of extracranial internal carotid aneurysms, stated that reports have appeared in the Soviet literature of supraorbital pain and oculosympathetic paresis accompanying this variety of aneurysm.

References
6. Kunkle, E. C., and Anderson, W. B. Significance of minor eye signs in headaches of