Hemilingual spasm: a new neurosurgical entity?

Case report

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Hemilingual spasm is a little-known movement disorder, presenting as intermittent paroxysmal involuntary contractions of half of the tongue muscles. The authors report a case of hemilingual spasm caused by an arachnoid cyst. After marsupialization of the cyst, the patient’s symptoms immediately resolved. There has been no recurrence of hemilingual spasm during the follow-up period of more than 40 months.

KEY WORDS • arachnoid cyst • movement disorder

In comparison with hemifacial spasm, hemilingual spasm (our term) is a little-known clinical entity described in only two patients by Lees, et al., as paroxysmal hemiglossal twitching. It consists of sudden episodes of speech difficulty due to involuntary spasms of half of the tongue. The spasms start as a feeling of stiffness or numbness lasting only seconds, and triggers include talking, eating, and stress. We report on a patient who had a similar clinical picture to the ones described by Lees, et al.; our patient responded well to a marsupialization of a premedullary arachnoid cyst.

History. This 50-year-old man presented to our clinic with paroxysmal spasms of the right side of his tongue, which were causing speech difficulties. These episodes had already lasted for 8 months and had recently become bilateral; the spasms never occurred simultaneously on both sides but alternated. The symptom-free intervals shortened and the spasms lasted progressively longer. The paroxysms were triggered by eating, talking, and stress. Apart from the disturbing feeling, the spasms also created speech and swallowing difficulties. While having these spasms he experienced his tongue becoming thicker and stiff, and it became purple on the spastic side, associated with ipsilateral numbness. The spasms lasted seconds or minutes and disappeared as quickly as they came.

The patient had a history of moderate chronic obstructive pulmonary disease, hypertension, and hyperlipemia. When he first noted the hemilingual spasms, he went to a local hospital where he was treated with intravenously administered heparin for a suspected transient ischemic attack. Results of admission computerized tomography, and MR images were normal except for a temporal arachnoid cyst on the right side. Four-vessel angiography demonstrated only a 40% stenosis of the left internal carotid artery. When the symptoms recurred, the patient was given warfarin and later ticlopidine.

Examination. Results of the patient’s clinical and neurological examinations were normal, including his hypoglossal function.

Neuroimaging and Functional Studies. A thorough look at his admission T2-weighted MR study (Fig. 1 upper) revealed a previously unnoticed premedullary arachnoid cyst predominantly on the right side that was compressing his lower brainstem. Results of an electromyographic study of his tongue were normal.

Treatment. Because we considered this arachnoid cyst to be the cause of our patient’s unique clinical picture, the ticlopidine was discontinued and 200 mg carbamazepine was administered three times per day. His symptoms resolved for 3 months, after which they recurred. The patient was then offered surgery consisting of a marsupialization of the premedullary cyst.

Operation. The patient was placed in a three-quarters prone position with his head rotated to the right. An extreme-lateral inferior transcondylar approach allowed a low retrosigmoid suboccipital craniectomy after skeletonization of the sigmoid sinus and the jugular bulb. The dura mater was incised and the premedullary arachnoid cyst was readily seen; it was stretching the hypoglossal nerve fibers. The cyst was marsupialized and most of the cyst wall was resected.

Abbreviation used in this paper: MR = magnetic resonance.
Postoperative Course. The involuntary hemilingual spasms ceased immediately after surgery and have not recurred during a 40-month follow-up period. The immediate and late postoperative course was uneventful. On postoperative MR imaging, again performed with T2 weighting (Fig. 1 lower), flow voids were demonstrated in the region where the arachnoid cyst had been located, indicating that cerebrospinal fluid motion had occurred after cyst wall resection.

Discussion

Arachnoid cysts of the posterior fossa are lesions that produce a variety of symptoms, depending on the location of the lesion and the degree of brainstem, cerebellar, or cranial nerve compression. Because of the often vague and varying symptomatology, the lesions are often misdiagnosed or not diagnosed until after a protracted clinical course. Larger cysts can give rise to increased intracranial pressure, but smaller cysts such as the one seen in this patient only create focal neurological dysfunctions by nerve compression. Most often only hypofunctioning of cranial nerves is described, such as hearing loss, facial paralysis, and trochlear nerve palsy. Rarely, patients with arachnoid cysts present with hyperactive cranial nerve syndromes such as hemifacial spasm, contralateral trigeminal neuralgia, or vestibular nerve excitation, which are readily seen in microvascular compression syndromes.

The pathophysiological features of the condition are unknown, but Arbusow, et al., argue that, based on oculomotor analysis and electronystagmography, ectopic discharges in the eighth cranial nerve might be the cause of vestibular nerve excitations in a patient in whom an arachnoid cyst was compressing the vestibulocochlear nerve. The same mechanism is also proposed for microvascular compression syndromes; thus, we can propose the following pathophysiological hypothesis explaining the 12th nerve hyperactivity, which presents clinically as hemilingual spasm. The arachnoid cyst causes a stretching of the hypoglossal fibers. This nerve stretching can create a focal demyelination, as has been demonstrated in guinea pigs. A focal demyelination leads to ectopic excitation, resulting in the reorganization of the hypoglossal nucleus. This has been called the kindling theory. The reorganization of the brainstem motor nucleus is believed to be responsible for the hyperfunctioning of the affected nerve, in this case the 12th cranial nerve. This can be supported indirectly by the fact that carbamazepine resolved the hemilingual spasms for 3 months, in the same way that it can temporarily alleviate trigeminal neuralgia, glossopharyngeal neuralgia, and, sometimes, hemifacial spasm.

The preferred treatment for symptomatic arachnoid cysts has generally been surgical drainage, either by resection of the cyst wall as completely as possible or by marsupializing the cyst into the subarachnoid space.

The importance of this case report lies in the fact that it demonstrates that a small arachnoid cyst of the posterior fossa can result in an isolated cranial nerve dysfunction, even a hyperactive dysfunction, as is clinically demonstrated in hemilingual spasm.

Acknowledgment

We thank M. Pieters for her enthusiastic technical support in preparing this manuscript.

References

Hemilingual spasm


Manuscript received July 31, 2001. Accepted in final form March 29, 2002.
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