Commentary

The authors reported a very rare case of hemifacial spasm (HFS) caused by an epidermoid tumor in the cerebellopontine angle (CPA) and emphasized the significance of the MRI study since HFS could be the sole symptom of CPA epidermoids.[1] Because surgical removal of the tumor is the curable approach, early diagnosis is essential. This young patient was operated on and HFS completely disappeared after surgical removal of the epidermoid. As the authors described, hyperactive dysfunction of the cranial nerves such as trigeminal neuralgia (TN) and HFS may be the initial and only symptom that patients with CPA epidermoids experience. The occurrence of these symptoms at a younger age is characteristic in patients with epidermoids in contrast to those due to a vascular cause. An early radiological study is recommended to avoid non-curable treatment without correct diagnosis.

Epidermoid or "pearly" tumors were described by Cruveilhier[2] and designated the "most beautiful tumors of the body" by Dandy.[3] They are slow-growing congenital lesions of ectodermal origin, representing approximately 1% of all primary intracranial tumors. The CPA is one of the most common sites affected, and patients with this tumor may present symptoms of cranial nerve, cerebellar, and brainstem dysfunction, as well as hydrocephalus and

meningeal irritation.[4] Of interest, hyperactive dysfunction of cranial nerves, such as TN and HFS, has been reported in association with this rare tumor. Meta-analysis of 263 cerebellopontine angle epidermoids disclosed that hearing difficulty was the most common symptom, accounting for 37.6%, followed by TN, 29.7%; dizziness or vertigo, 19.4%; facial palsy, 19.4%; headache, 17.9%; and diplopia, 16.7%. HFS was found in only 4.9% of patients in these reports.[4] According to the recent reports, 55 of 6910 HFS patients (0.8%) who underwent microsurgical procedure harbored CPA tumors, 41 of them (74.5%) were epidermoids.[5] Among 2050 HFS patients, CPA tumors were detected in 9 (0.4%) and 2 of them were epidermoids.[6]

As epidermoids flow into any available subarachnoid space and slowly increase their volume, they conform to the shape of the cavities they enter and do not displace normal neurovascular elements until all available subarachnoid space is occupied. As a result, the cranial nerves and arteries are embedded or displaced by the tumor. The tumor-neurovascular relationship is categorized in four types. The cranial nerves may be wrapped (A) or compressed (B) by the tumor as the tumor increases its volume. The nerve may be displaced by the tumor to contact the artery at the opposite side of the tumor in one side, resulting in its being pinched by both the tumor and the artery (C). When the artery is situated between the tumor and the nerve, the artery may be trapped and may begin to compress the nerve as the tumor grows (D).[4] Recently, arterial compression at the root exit zone was also reported in the majority of HFS cases with CPA epidermoids.[5,6]

The neurosurgeon must keep in mind that the symptom is elicited by compression of the nerve by the tumor per se, by an artery that is displaced to the nerve, or by both. Careful resection of the tumor, whose capsule occasionally is strongly adherent to the neurovascular structures, is necessary, and microvascular decompression should be performed in some cases to achieve a complete, permanent cure of symptoms with a low rate of recurrence. Total removal is ideal and must be the goal of the operation; however, the occasional tendency of strong adhesion of the tumor to vital neurovascular structures and tumor extension far beyond the midline may prevent its total removal. A recent series of reports on CPA epidermoids showed total removal rates ranging from 18 to 97%.[4-8]

In the literature, the regrowth rate of epidermoid in long-term follow-up has been reported to vary from 0% to 30%, with a trend toward a higher rate in cases with a longer follow-up period.[4] Therefore, a long-term follow-up is warranted for this young patient.

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