Schwannoma arising from the intermediate nerve and manifesting as hemifacial spasm

Case report

AKIRA KUDO, M.D., MICHIYASU SUZUKI, M.D., NAOHIKO KUBO, M.D.
KIYOSHI KURODA, M.D., AKIRA OGAWA, M.D., AND YUZO IWASAKI, M.D.

Department of Neurosurgery, Iwate Medical University, and Department of Neurological Sciences,
Tohoku University School of Medicine, Morioka, Japan

This 35-year-old man presented with left facial spasm that had persisted for 10 months. Microvascular decompression was performed to relieve the symptom. No responsible vessel could be identified during surgery, but a small mass seeming to arise from the intermediate nerve and compressing the seventh cranial nerve was removed. The histological diagnosis was Antoni-B type schwannoma. This unique case of schwannoma arising from the intermediate nerve was recognized by means of an operative microscope, and supports the idea that portions of the seventh nerve schwannoma originate from the components of the intermediate nerve.

Key Words • schwannoma • intermediate nerve • facial nerve • facial spasm • microvascular decompression • pathogenesis

CHWANNOMAS in the seventh cranial nerve are believed to originate from the intrinsic sensory fiber of the intermediate nerve or from aberrant ganglion cells. However, schwannoma arising from the intermediate nerve has not been described among over 300 cases since the first report in 1931. Recently, Saito and Baxter described two interesting cases of seventh cranial nerve schwannoma that could be distinguished microscopically from the seventh nerve motor root.

We describe a patient with a schwannoma that manifested as hemifacial spasm and arose from what we recognized as the intermediate nerve during surgery. We also discuss the pathogenesis of the seventh nerve schwannoma.

Case Report

This 35-year-old man was admitted to our department with left hemifacial spasm that had first occurred 10 months earlier. His family and medical histories were non-contributory.

Examination. The patient was alert with no neurological deficit except hemifacial spasm. Physical examination and laboratory data, as well as magnetic resonance imaging and x-ray computerized tomography, disclosed no abnormalities. However, left vertebral angiography demonstrated downward displacement of the left superior cerebellar artery. Microvascular decompression of the left seventh cranial nerve from the left superior cerebellar artery was scheduled to relieve hemifacial spasm.

Operation. A left suboccipital craniotomy was performed. Cerebrospinal fluid was aspirated, and the cerebellar hemisphere was retracted to disclose the seventh and eighth cranial nerve complex. No vessels responsible for the hemifacial spasm could be identified along the entire length of the seventh nerve. However, a small mass seeming to arise from the intermediate nerve was observed in the vicinity of the root entry zone of the seventh nerve, where it had partly adhered to and compressed the nerve (Fig. 1). The intermediate nerve was cut proximal and distal to the mass, because it was difficult to separate the mass and the intermediate nerve without injury. After removal, a prominent depression on the surface of the seventh nerve was observed. The mass measured approximately 2 × 3 × 5 mm and was relatively soft, grayish yellow, and slightly vascularized.

Postoperative Course. The postoperative course was uneventful. The facial spasm ceased immediately after the operation. One and one-half years after the operation, we scheduled a follow-up visit to check for facial spasm, and intermediate, seventh, and eighth nerve function. The
Facial spasm had not recurred. Objective examinations of the seventh and eighth nerves, including audiogram and stapedial muscle reflex, disclosed no laterality. Furthermore, there was no obvious decrease in tearing, and no taste disturbance found objectively, and the patient did not complain of sensory disturbance in the external auditory meatus.

**Histological Examination.** Light microscopy showed tumor cells that varied slightly in size that were aggregated against a loose-textured pattern of tissue (Fig. 2 left). Immunohistochemistry showed that the foamy tumor cells stained positively for S-100 protein, suggesting that the tumor origin was a Schwann cell (Fig. 2 right). The tumor was diagnosed as Antoni-B type predominant schwannoma.

**Discussion**

The origin of the schwannoma in the present case was considered to be the intermediate nerve itself, as seen under the operative microscope, and is important evidence supporting the idea that part of the seventh nerve schwannoma originates from the intermediate nerve.

There are three explanations for why no observation of intermediate nerve schwannoma has been made before. The first explanation is based on anatomical features. The intermediate nerve, which runs through the subarachnoid space from the brainstem to the internal auditory meatus, is divided into three segments: 1) the proximal segment, which exits the brainstem accompanying the eighth nerve and has a mean length of 6 mm; 2) the intermediate segment, which runs between and has a mean length of 10 mm; and 3) the distal segment, which passes into the internal auditory meatus with the seventh nerve motor root, and has a mean length of 5 mm. It is extremely difficult to find the intermediate nerve during surgery, even using the operative microscope, because it can only be clearly identified by the intermediate segment (10-mm length). Furthermore, the intermediate segment is absent in 22% of cases. A larger schwannoma arising from the intermediate segment easily involves and adheres to the neighboring seventh or eighth nerves. Such a tumor might be misdiagnosed as a seventh or eighth nerve schwannoma.

The second explanation is based on the location distribution of seventh nerve neurinoma. The vertical portion is the most common origin (58%), whereas the cerebellopontine angle, in which the intermediate nerve could possibly be identified, is the origin in less than 10% of cases. A. Kudo, et al.
Intermediate nerve and schwannoma

The third explanation refers to the symptomatology. According to reports, schwannoma located at the cerebellopontine angle only causes facial spasm in approximately 2.5% of cases.11,12 The most common symptom among the 15 cases of seventh nerve schwannoma in the cerebellopontine angle2,3,6–8,10,15,17 was auditory disturbance (10 of 15 cases) due to compression of the neighboring eighth nerve. Other symptoms included facial weakness (five of 15), tinnitus (four of 15), dizziness or vertigo (four of 15), and headache (two of 15 cases). Only three of the 15 cases showed facial spasm caused by simple compression of the seventh nerve motor root by a small mass. These considerations suggest that our case may be extremely rare.

The absence of neurological deficits on follow-up study of the patient’s intermediate, seventh, and eighth nerves raised doubts about what nerve-like structure it was that was cut during surgery. The structure was clearly demonstrated as the intermediate nerve in the intraoperative photograph (Fig. 1), and the surgical specimen consisted of neuronal elements. The report that the intermediate segment of this nerve is absent in 22% of cases16 suggests that our case may be extremely rare. The absence of neurological deficits on follow-up study of the patient’s intermediate, seventh, and eighth nerves may have been observed during surgery. This may be one reason why our patient did not show any neurological deficit after we cut the intermediate nerve. We need further studies of this nerve in the fields of anatomy and neurosurgery to obtain genuine answers.

References


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