Intracranial hypoglossal schwannoma as an unusual cause of facial nerve palsy

Case report

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A case is presented in which facial palsy resulted from a hypoglossal schwannoma encircling the nerve in its course through the temporal bone.

KEY WORDS • schwannoma • facial nerve • hypoglossal nerve • posterior fossa tumor • neurilemmoma • neurinoma • jugular foramen

The facial nerve can be compromised anywhere in its course, from the nucleus in the brain stem until it arborizes within the parotid gland. We present a patient with facial palsy due to a hypoglossal schwannoma encircling the nerve in its course through the temporal bone.

Case Report

This 24-year-old woman initially presented in October, 1977, for evaluation of numbness and marked hemiatrophy of the left side of the tongue. Investigations at that time included basal skull films, jugular foramen views, and a computerized tomography (CT) scan, all of which were interpreted as normal. Vertebral angiography showed the left anterior inferior cerebellar artery to be stretched in comparison to the right, but this was thought to be a normal variant. No hypoglossal canal tomograms were made at this time, and no diagnosis was made.

She next presented in July, 1979, during the first trimester of pregnancy because of the development of tinnitus and decreased hearing in the left ear. At that time, a mass was noted behind the left tympanic membrane but investigation was deferred. On March 22, 1980, 1 day before delivery, she awoke with a left facial palsy.

Examination. She had a complete paralysis of all branches of the left facial nerve, a complete left sensorineural hearing loss, decreased sensation on the left side of the pharynx, left vocal cord weakness, and weakness of the left sternomastoid muscle. Also, there was marked atrophy of the left side of the tongue. The rest of the neurological examination was normal. There were no stigmata or family history of von Recklinghausen's disease.

The left hypotympanic mass that had been seen previously was still present. Tomograms of the hypoglossal canals revealed severe erosion of the left petrous temporal bone from the foramen magnum to the clinoid processes, and erosion of the jugular bulb laterally (Fig. 1). Angiography revealed a left cerebellopontine angle and jugular foramen mass, with complete obstruction of the left transverse and sigmoid sinuses. A CT scan revealed a left cerebellopontine angle mass which enhanced after the infusion of contrast material, and destruction of a wide area of the skull base in the region of the jugular foramen (Fig. 2).

Operation. In July, 1980, the patient underwent a left partial mastoidectomy and labyrinthectomy, and decompression of the whole facial nerve. The tumor, which proved to be a schwannoma, was wrapped around the facial nerve. With the aid of the operative microscope, the continuity of the facial nerve was preserved. At operation, it was thought that all the intraosseous tumor had been removed; however,
Intracranial hypoglossal schwannoma

FIG. 1. Tomography of the base of the skull, showing extensive erosion of the left occipital bone lateral to the foramen magnum.

FIG. 2. Left: Enhanced computerized tomography scan showing the intracranial extent of the tumor (arrows). Right: Reverse image of the same scan showing both the intracranial and intraosseous portions of the tumor.

follow-up CT revealed residual intracranial tumor (Fig. 3). Ten days later, the patient underwent a left suboccipital craniectomy with microsurgical removal of the intracranial part of the tumor. The seventh through 11th cranial nerves were seen to be stretched over a large extradural mass. The 12th cranial nerve could not be visualized intradurally. After a cruciate incision of the dura, the mass was removed, piecemeal. At the end of the operation, the seventh through 11th cranial nerves were clearly seen throughout their intracranial course. The 12th cranial nerve still could not be visualized either intradurally or in the evacuated extradural tumor cavity.

Postoperative Course. There were no changes in the patient’s pharyngeal sensation or gag reflexes, and a tracheostomy was not necessary. A postoperative leak
of cerebrospinal fluid through the ear was successfully treated through the mastoidectomy incision.

Although the left 12th nerve was not visualized intraoperatively, the clinical presentation, with early unilateral atrophy of the tongue, and the radiological evidence of marked enlargement of the hypoglossal canal, supports the belief that this tumor originated from the left 12th nerve.

**Discussion**

Extracranial and intracranial schwannomas of the hypoglossal nerve are unusual but important lesions because they are benign and potentially curable. The hallmark of the intracranial hypoglossal schwannoma is the early, marked unilateral lingual atrophy. However, because of the minimal disability produced by the deficit, patients do not usually present until quite late, by which time they are showing signs of cerebellar or brain-stem compression or hydrocephalus. There is often involvement of the lower four cranial nerves, and less often of the facial, vestibulocochlear, and trigeminal nerves. Twelfth nerve involvement has been noted for 5 months to 10 years prior to definitive diagnosis.

Our patient did not have a definitive diagnosis made at her first admission, and it was 2 more years before progression of the tumor again brought her to medical attention. She is also typical of the sex (73% female) and side (73% left) noted in previous reports. She represents one of the youngest cases to be reported; only Bailey, et al., and Morelli have recorded younger cases. However, in the case of Bailey, et al., the 12-year-old girl had von Recklinghausen's disease.

Of the previous hypoglossal schwannomas, only two have been of the "dumbbell" type characteristic of spinal schwannomas. The present case is the third example of this, and is the second case requiring both an intra- and an extracranial approach to the tumor. Based on the available operative reports, the tumor in the present case is the only one located both intracranially and extradurally. As such, it in theory could have been entirely removed via the mastoid approach extradurally, and initially this was what was thought. Follow-up CT, however, demonstrated the residual tumor clearly, and the necessity for the posterior fossa approach.

The facial nerve is not usually involved until late in patients with hypoglossal schwannomas, and even then there is usually only a mild weakness. This was evident in six of the previous 15 cases. None had a complete facial palsy, however.

Thus, this case of a "dumbbell" type of hypoglossal schwannoma is the first example of a complete facial nerve palsy produced by a hypoglossal schwannoma. It also illustrates the usefulness of a staged extracranial and intracranial approach to these tumors when they are large, and shows the usefulness of CT scanning in determining the adequacy of tumor removal.

**References**


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**Fig. 3.** Enhanced computerized tomography scan following the removal of the petrous intraosseous portion of the tumor, showing residual intracranial tumor and air.
Intracranial hypoglossal schwannoma


Manuscript received April 22, 1981.
Accepted in final form October 19, 1981.
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