A vascular malformation mimicking an intracanalicular acoustic neurilemoma

Case report

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✓ A patient with an enhancing, completely intracanalicular mass on magnetic resonance imaging was operated on for a presumed acoustic neurilemoma, but was found at surgery to have an intracanalicular vascular malformation. This rare lesion should be distinguished from angiomatosus change within an acoustic neurilemoma and in the past has been termed "vascular tumor," "hemangioma," or "fibro-angioma." The clinical distinctions between intracanalicular acoustic neurilemomas and intracanalicular vascular malformations and the ability of magnetic resonance imaging to distinguish between the two are discussed.

KEY WORDS ♦ acoustic neurilemoma ♦ arteriovenous malformation ♦ hemangioma ♦ cerebellopontine angle ♦ tumor, facial nerve ♦ magnetic resonance imaging

In most clinical series, 7% to 17% of intracranial vascular malformations arise in the posterior fossa.9,24,31 A prospective autopsy series of vascular malformations placed the figure at 13%25 while a retrospective autopsy series arrived at a figure of 32%.26 Vascular malformations of the posterior fossa can be divided into those in the cerebellum, in the brain stem, and in the cerebellopontine angle (CPA).10,22,35 Of the three types, those in the cerebellum are the most common.22,26

Vascular malformations constitute less than 1% of all pathological entities arising in the CPA.5,23,35 These lesions involve the pial surface of the brain stem or cerebellum but do not penetrate brain parenchyma.10,35 The internal auditory canal is a cerebrospinal fluid-containing space lined by dura which lies within the CPA. Four vascular lesions labeled "arteriovenous malformations" (AVM's) involving solely the internal auditory canal have been reported.3,19,21,34

We encountered an AVM 8 mm in diameter in a patient who underwent surgery for what appeared, by magnetic resonance (MR) imaging, to be an intracanalicular acoustic neurilemoma. The rarity of this entity and the fact that it was not distinguished preoperatively from an acoustic neurilemoma by MR imaging warrant publication. Authors have reported difficulty in distinguishing this lesion microscopically from other "vascular tumors" in the internal auditory canal including what have been termed "facial nerve hemangiomas," "facial nerve fibro-angiommas," "eighth nerve hemangiomas," and "ossifying hemangiomas of the temporal bone," as well as angiomatosus changes within acoustic neurilemomas;16 this reported difficulty also prompted our attempt to clarify some of these problems.

Case Report

This 39-year-old right-handed man was in good health until 5 years before presentation, when he had intermittent vertigo associated with periods of imbalance leading him to bump into objects. After 1 year, these symptoms spontaneously resolved. Three years before presentation, he noticed progressive hearing loss in the right ear without tinnitus. Six months before admission, he had only 50% speech discrimination in his right ear, and his hearing loss worsened after that. He also complained of rare, intermittent "twitches" or "spasms" of his right side of his face which had occurred unpredictably over the 4 months before presentation. He denied any pulsations in his right ear, headaches, or any other cerebellar or cranial nerve symptoms.

Examination. Neurological examination was normal except for decreased hearing in the right ear. There were no other cranial nerve or cerebellar signs. The patient had no nystagmus or intracranial bruits. Hemi-
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facial spasm could not be elicited with forced eyelid closure followed by rapid opening.

An audiogram revealed severe to profound hearing loss on the right with no speech discrimination and absent reflex thresholds (Fig. 1). Brain-stem auditory evoked responses were normal on the left and absent on the right. Magnetic resonance imaging enhanced with gadolinium-diethylenetriaminepenta-acetic acid (DTPA) demonstrated an 8-mm intracanalicular enhancing lesion on the right which was presumed to be an intracanalicular acoustic neuroma (Fig. 2).

Operation and Course. The patient underwent a right retromastoid craniectomy in the lateral decubitus position. The lesion was not visible until after the posterior lip of the porus acusticus had been drilled off with a high-speed drill. The porus was flared. When the dura was opened in the canal, the superior and inferior vestibular nerves appeared normal, although perhaps bulging a bit posteriorly. The vestibular nerves were split from one another and a lesion was palpated then exposed. Grossly it was gray-purple and appeared to be a meningioma. It was removed piecemeal and seemed to be adherent or attached to the dura caudally. This area of dura was coagulated. The nerves were not injured in any apparent way, but there was some evidence that the lesion was either adherent or attached to the facial nerve because the intraoperative facial nerve electromyelogram showed some evidence of hyperactivity on removal of the last fragment of tumor with sharp dissection. The patient recovered quickly from his operation and was discharged on the 7th postoperative day with mild (House and Brackmann Grade II) facial weakness which was gradually improving. He remained deaf in his right ear.

Pathological Examination. Histopathologically, the lesion consisted of dense collagenous connective tissue with multiple, irregular vascular channels lined by a single layer of endothelial cells. Most vessels were venous but a few had thick vascular walls resembling small arteries (Fig. 3). A few vessels had mural architecture with the features of both arteries and veins. There was no hemosiderin. No peripheral nerve tissue or Schwann cells were present within the specimen.

Discussion

Vascular Malformations

The histopathological classification now widely accepted for intracranial malformations divides the malformations into four types: AVM's, cavernous malformations, venous malformations, or telangiectases. Vascular malformations represent hamartomas or developmental anomalies, which enlarge by hemorrhage or engorgement of previously existing vascular channels. They may be referred to as “tumors” in the sense that they occupy space; however, they are not neoplastic in the usual sense and to label them as “angiomas” or “hemangiomas” is histologically and etymologically inappropriate.

![Image](https://via.placeholder.com/150)

**Fig. 1.** Preoperative pure-tone audiogram and speech discrimination scoring. PTA = pure tone acuity; SRT = speech reception threshold; DISCR = speech discrimination; RE = right ear; LE = left ear.

![Image](https://via.placeholder.com/150)

**Fig. 2.** Preoperative axial T1-weighted magnetic resonance images, unenhanced (left) and enhanced with gadolinium-DTPA (right), demonstrating a small enhancing intracanalicular lesion presumed to be an acoustic neuroma.
Cerebellopontine angle AVM's commonly cause cranial neuropathies.\(^9,33\) Dural AVM's have also been reported to cause cranial nerve symptoms\(^3,19\) including facial paraspasm.\(^3,17,23\) However, this patient did not have headaches, pulsatile tinnitus, or an audible bruit, all of which are described with dural AVM's in the CPA.\(^3,21\) Thus, our patient most likely had a CPA AVM which, in this location, most likely was supplied by the labyrinthine artery arising from the anterior inferior cerebellar artery.

**Hemangiomas**

The subject of intracanalicular vascular malformations includes a confusing array of entities in the literature including intracanalicular “vascular tumors” referred to as “facial nerve hemangiomas,”\(^4,6,11,18-21,30,32,34\) “facial nerve fibro-angiomas,”\(^9\) and “eighth nerve hemangiomas.”\(^29,30\) Reviewing the occasional published histology of these reported lesions, we concluded that they were not neoplasms, but many were cavernous or venous malformations.\(^3,11,21,30,32,34\) We expanded our search to include “vascular tumors,” and were able to find 21 reported cases of vascular malformations limited to the internal auditory canal; four AVM's\(^5,19,21,34\) and 17 “hemangiomas.”\(^1,6,11,19,20,29,30,34\)

“Hemangiomas” have been reported to involve the facial nerve\(^1,4,6,11,18-21,30,32,34\) as well as the eighth cranial nerve.\(^29,30\) Many “hemangiomas of the facial nerve” have been described at operation as merely compressing the facial nerve\(^21\) or even as being outside the dura of the facial canal.\(^19\) Given the published histology of “facial nerve hemangiomas,” it seems reasonable to consider these lesions as vascular malformations of the vascular plexus supplying these cranial nerves. While our specimen may have been adherent to the facial nerve at surgery, there was clearly no peripheral nerve tissue in the surgical specimen, making it unlikely that this malformation arose in either the seventh or eighth cranial nerve.

**Ossifying Hemangiomas**

In 1977, Fisch and Rüttner\(^1\) defined another category of “hemangioma,” termed “ossifying hemangioma” based on the presence of lamellar bone within the specimen. Since that time, others have begun to make the diagnosis of “ossifying hemangioma” based on the presence of bone spicules in lesions enlarging the facial canal on computerized tomography scans.\(^8,12\) Hemangiomas of bone do exist\(^12,15,36\) and could in theory secondarily invade the facial canal or the internal auditory canal from a primary temporal bone origin. It is possible that most extradural “ossifying hemangiomas” do represent primary temporal bone neoplasms. Along these lines, it is interesting to note that in only one of the six cases in the series of Curtin, et al.,\(^8\) and in none of the three cases reported by Glasscock, et al.,\(^13\) was facial nerve invasion apparent at surgery or by microscopy. “Ossifying hemangiomas” that involve the facial nerve and arise intradurally may not be temporal bone neoplasms but may represent vascular malformations of the cavernous type which have been noted to have significant incidence of mineralization or ossification.\(^25\)

**Angiomatous Change in Neurilemomas**

Small acoustic neurilemomas are less likely to be vascular and exhibit less angiomatous change than larger tumors.\(^16\) Still, it is necessary to distinguish our AVM from the angiomatous changes within acoustic neurilemomas which are characterized by blood vessels that are abnormal in size and that can be quite tortuous. The fact that this lesion was separate from the eighth cranial nerve at surgery and that the surgical specimen revealed no Schwann cells makes this distinction possible.

**Differential Diagnosis**

The clinical time course of symptoms in our patient was not unusual for an acoustic neurilemma; however, we were impressed by the extent of hearing loss and the presence of seventh nerve symptoms despite the small size of the lesion. The clinical observation of cranial nerve signs and symptoms out of proportion to lesion size, first made in 1976 by Sundaresan, et al.,\(^34\) has been noted by others reporting intracanalicular vascular malformations\(^6,19-21\) and should serve as a preoperative warning that one may not be dealing with an acoustic neurilemma.

The distinction between an intracanalicular acoustic neurilemma and an intracanalicular vascular malformation cannot be made on the basis of MR imaging alone. In patients with a completely intracanalicular enhancing mass revealed by MR imaging, who have signs and symptoms out of proportion to the small size...
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of their lesion, one should consider preoperative angiography to exclude the possibility of an AVM. Cavernous malformations are often not visualized on angiography due to the absence of a direct arterial input. Thus, the preoperative distinction between this type of vascular malformation and a intracanalicular acoustic neurilemoma may be impossible at the present time.

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