Jacobson's Nerve Schwannoma Presenting as Middle Ear Mass

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Summary: Schwannoma is one of the common benign middle ear space tumors. Middle ear space schwannomas may originate from the nerves of the tympanic cavity or by extensions from outside the middle ear space. In the English-language literature, the facial nerve and chorda tympani nerve, but not yet the tympanic branch of glossopharyngeal nerve (Jacobson’s nerve), have been reported as the origins of intrinsic middle ear space schwannomas. We present the clinical and radiologic features of a middle-space schwannoma originating from Jacobson’s nerve, and suggest that such a tumor be included in the differential diagnosis of middle ear tumors.

Tumor and tumorlike conditions of the middle ear include hemangioma, meningioma, cholesterol granuloma, primary cholesteatoma, adenomatous tumor, and choristoma. Most neurogenic tumors arising in the middle ear are facial schwannomas. This report documents the imaging findings of a schwannoma arising from the tympanic branch of the glossopharyngeal nerve (Jacobson’s nerve).

Case Report

A 55-year-old woman presented with left-sided hearing loss and intermittent otalgia. She denied history of dizziness, tinnitus, or otorrhea. Physical examination revealed a mass within the middle ear space with an intact but bulging tympanic membrane. Audiography showed normal hearing on the right with a large conductive hearing loss on the left. Facial nerve testing was normal as was the remainder of the neurootologic examination. The patient did not have any stigmata of neurofibromatosis.

CT of the temporal bones revealed a well-circumscribed mass in the mesotympanum extending to the epitympanum (Fig 1 and 2). The long process of the incus and the head and crura of the stapes were eroded and the malleus and incus were displaced laterally by the mass. The promontory was also eroded, but there was no sign of labyrinthine invasion. The cochlea, vestibule, internal auditory canal, and the mastoid and labyrinthine segments of the facial nerve canal were normal. The tympanic segment of the facial nerve could not be distinguished separately from the mass, but the facial nerve canal was not enlarged.

On MR images, the mass was isointense to brain on T1- and T2-weighted sequences and showed marked homogeneous contrast enhancement (Fig 3). The mass occupied the mesotympanum and hypotympanum. There was no abnormal contrast enhancement in the internal auditory canal, facial nerve canal, jugular foramen, inner ear, or petrous apex.

The preoperative diagnosis was a benign tumor of the middle ear. The patient underwent middle ear exploration with intact canal wall mastoidectomy via facial recess approach. Findings at the time of surgery included: 1) facial nerve dehiscence along the entire tympanic segment, 2) carotid artery dehiscence and exposure, 3) promontory erosion with exposed cochlea, 4) erosion of the long process of incus and the head and crura of the stapes. The tumor was separated from the dehiscent facial nerve. The tumor was not of facial nerve origin. The mass was clearly encapsulated and removed in total. The pathologic diagnosis was schwannoma. Postoperatively, the patient did well without vestibular symptoms; however, subsequent audiography showed that she had lost hearing in the ear.

Discussion

Of the tumors of the middle ear and mastoid, paraganglioma is the most common. Other tumors include facial nerve schwannoma, hemangioma, meningioma, cholesterol granuloma, primary cholesteatoma, adenomatous tumor, and choristoma (1–3). An aberrant carotid artery and protruding jugular bulb are nonneoplastic masses important in the differential diagnosis. Schwannomas presenting as middle ear space masses may originate from the tympanic cavity or by extensions from outside the middle ear space. Schwannoma arising in the vestibulocochlear nerve may extend into the vestibule or cochlea along the internal auditory canal. They then grow through the lumen of the inner ear and prolapse into the middle ear space, usually through the round window (1, 4, 5). These tumors tend to cause a mixed type of hearing loss and vertigo. Schwannomas arising from cranial nerves IX, X, and XII may also present as middle ear masses, eroding through the lateral margins of the jugular foramen (6). These tumors may be confused with glomus jugulare. Radiologic clues to the identity of schwannomas include discrete borders and less vascularity than glomus tumors (7).

Schwannomas arising intrinsically in the middle ear may potentially originate from the facial nerve, the chorda tympani nerve, the tympanic branch of the glossopharyngeal nerve (Jacobson’s nerve), or the auricular branch of the vagus nerve (Arnold’s
nerve). The facial nerve is the most frequent origin of middle ear schwannomas. Schwannomas arising in the tympanic segment of the facial nerve may expand into the tympanic cavity and cause conductive hearing loss rather than facial nerve symptoms (8, 9) as did the Jacobson’s nerve schwannoma in our report. Schwannomas originating from the chorda tympani branch of the facial nerve have also been reported (10), but none yet from Arnold’s nerve or Jacobson’s nerve.

Jacobson’s nerve arises from the inferior ganglion of the glossopharyngeal nerve, enters the tympanic cavity through the inferior tympanic canalculus, and ascends in a groove or canal on the medial wall of the middle ear, usually on the cochlear promontory (Fig 4). It provides the main sensory fibers to the mucosa of the mesotympanum and the eustachian tube, and joins the caroticotympanic nerve to form the lesser superficial petrosal nerve (7). On rare occasions, the inferior tympanic canal can be identified on high-resolution CT (11) (Figure 5). Arnold’s nerve (auricular branch of the vagus nerve) arises from the superior ganglion of the vagus nerve. It reaches the descending facial nerve canal via the mastoid canaliculus, which is also visible on high-resolution CT scans (12).

In retrospect, erosion of the cochlear promontory shown by CT is the only clue to suggest the correct origin of the tumor as Jacobson’s nerve in our case. Another anatomic sign would be enlargement of the inferior canaliculus, which was not detected in our case.

Clinically, the outcome of surgical resection of a Jacobson’s nerve schwannoma is likely to be more favorable than that of a facial nerve schwannoma in that the potential of facial nerve palsy can be spared.

In conclusion, Jacobson’s nerve is a previously unreported site of middle ear schwannomas. Imaging clues to the correct origin of such a tumor are absence of facial nerve canal involvement, er-
osion of the cochlear promontory, and possible enlargement of the inferior tympanic canaliculus.

References