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Primary Paraganglioma of the Facial Nerve Canal

L. V. Petrus and W. M. M. Lo

Summary: We report two cases of primary paraganglioma of the facial nerve canal. This entity should be considered in patients presenting with facial paresis or pulsatile tinnitus. Paraganglioma should be considered when a lesion appears to arise from the facial nerve canal.

Index terms: Paraganglioma; Nerves, facial (VII)

Three cases of primary paraganglioma of the facial nerve canal have been reported in the literature (1, 2). We present two cases to highlight the imaging findings and suggest that facial paragangliomas should be considered in patients presenting with facial palsy or pulsatile tinnitus.

Case Reports

Case 1

A 74-year-old woman presented with a 5-year history of slowly progressive drooping of the left face. Clinical examination confirmed a complete left lower motor neuron seventh-nerve palsy. A small red mass was seen in the inferior aspect of the fundus of the external auditory canal. Computed tomography (CT) of the temporal bones demonstrated circumferential widening of the distal left descending facial nerve canal. In addition, there was breeching of the posterior wall of the external auditory canal and irregular bone destruction of the inferior medial mastoid air cell. A 1-cm soft-tissue mass extended down and out the stylomastoid foramen (Fig 1A and B). Selective left external carotid angiogram revealed a 2 × 1.5-cm hypervascular mass in the left mastoid region, fed by enlarged branches of the left occipital and left posterior auricular arteries, with rapid shunting into the left internal jugular vein. The ascending pharyngeal artery and the jugular bulb were normal (Fig 1C and D).

Mastoidectomy revealed a reddish nodular tumor invading the stylomastoid foramen and surrounding the facial nerve sheath in the mastoid. Biopsy of the lesion revealed nests of epithelioid tumor cells, with round to oval nuclei containing moderate amounts of foamy cytoplasm. The cell nests were arranged in an alveolar pattern, separated from one another by a richly vascularized trabecular connecting tissue, consistent with a paraganglioma (Fig 1E). Complete excision of the tumor was not possible and postoperative radiotherapy was recommended.

Case 2

A 74-year-old woman reported right pulsatile ringing. Clinical examination revealed a vascular impression on the right tympanic membrane. There was no evidence of facial paralysis. CT of the temporal bones demonstrated a mass expanding the middescending facial nerve canal, with erosion of the adjacent posterior wall of the external auditory canal and extension into the middle ear cavity (Fig 2A and B). Exploration and biopsy of the mass showed endothelium-lined vascular structures surrounded by circumscribed nests of spindle and ovoid cells with hyperchromatic nuclei. The diagnosis of paraganglioma was confirmed by pathologists of the Armed Forces Institute of Pathology (Washington, DC). Surgical resection of the tumor was not advised because of the patient's age and the mild symptoms.

Discussion

Paraganglioma is the most common neoplasm in the temporal bone, aside from acoustic schwannoma (3). Most paragangliomas arise from the jugular bulb, the skull base, and the tympanic cavity, or involve a combination of the above sites (4). Large jugular paragangliomas often involve the descending facial nerve by extension (5). Paragangliomas arising from the facial nerve canal itself, however, are an extremely rare entity.

In Guild's histologic study of 88 temporal bones, he found along the course of the inferior tympanic branch of the glossopharyngeal nerve (Jacobson's nerve) and the auricular branch of the vagus nerve (Arnold's nerve), 248 paragan-

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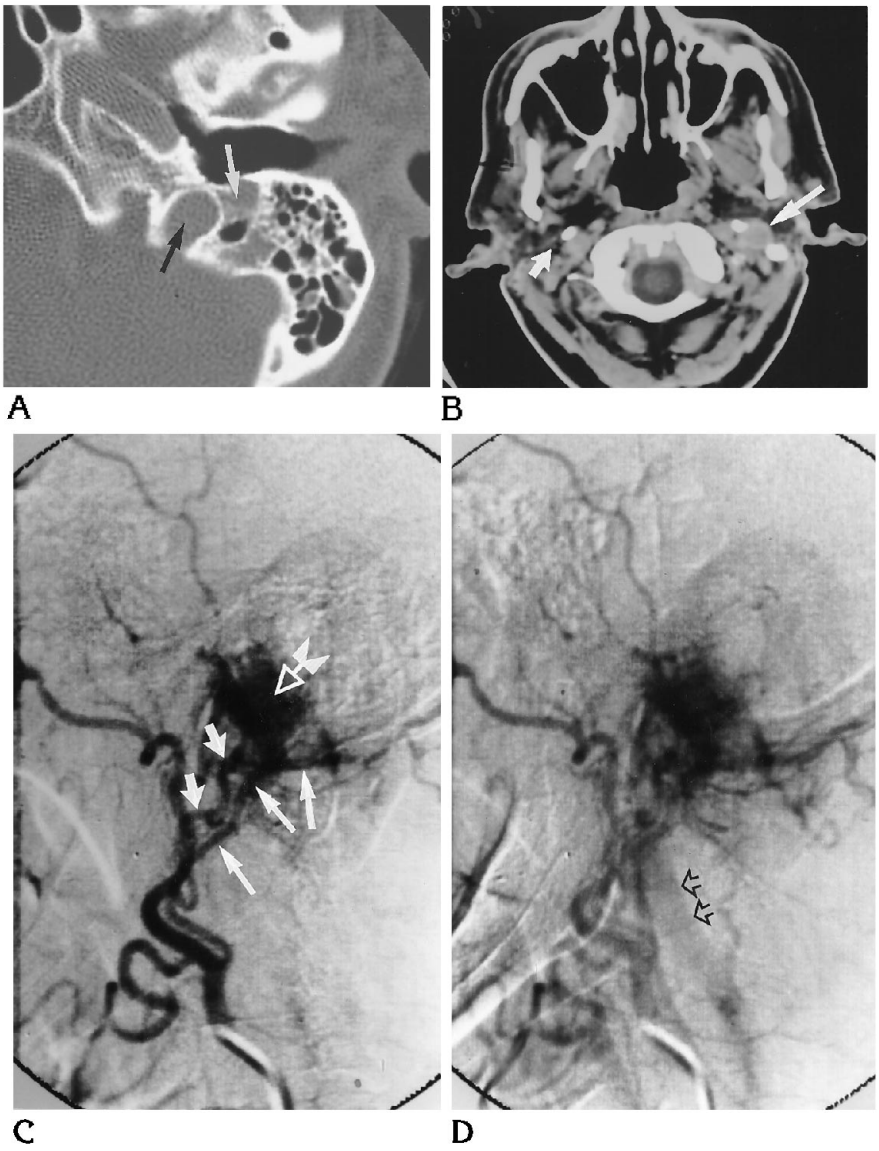

Fig 1. Primary paraganglioma of the left facial nerve canal.

A and *B*, Axial CT scans through the left temporal bone and at the level of the stylomastoid canal. On *A*, there is circumferential widening of the descending facial nerve canal (*white arrow*). The jugular foramen (*black arrow*) is intact. On *B*, a soft-tissue mass is present in the left stylomastoid canal (*larger arrow*). On the *right*, the normal facial nerve surrounded by fat is noted (*smaller arrow*).

C and *D*, Selective left external carotid arteriograms, lateral projection. On *C*, a densely staining tumor is present (*open arrowhead*), fed by branches of the left occipital (*three larger arrows*) and left posterior auricular arteries (*two smaller arrows*). On *D*, there is early filling of the left internal jugular vein (*arrows*).

Figure continues.

glia (glomus bodies or glomus jugulare). Approximately one half of them were located in the adventitia of the jugular bulb, one fifth in the inferior tympanic canaliculus, and one tenth over the cochlear promontory. These prevalent locations of the paraganglia account for the sites of origin of the vast majority of paragangliomas in the temporal bone. The remaining small number of paraganglia found were along Jacobson's nerves, superior or distal to the cochlear promontory, and along branches of Arnold's nerve in or below the mastoid canaliculus between the jugular bulb and the descending facial nerve canal, and within the descending facial nerve canal itself. These locations are thus additional sites in the temporal bone in which paragangliomas may arise on rare occasions. The two tumors in this report, as well as the three reported earlier (1, 2), presumably originated from paraganglia located within the descending facial nerve canal.

The three previously reported patients and the two present ones included one man and four women who ranged from 20 to 70 years in age. Their symptoms were facial weakness in three, pulsatile tinnitus in two, and aural fullness in one. Notably one of our patients did not have facial paralysis. A mass in the external auditory canal and/or tympanic cavity was seen in four. In all three patients who underwent tumor re-

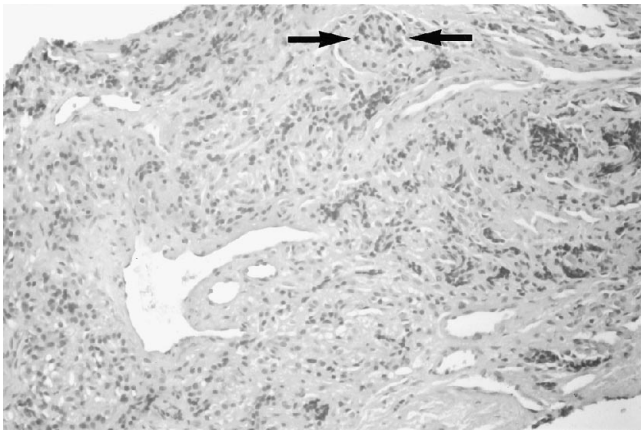

Fig 1, continued. *E*, Photomicrograph of the lesion shows a vascular tumor with scattered epithelioid cells arranged in compact Zellballen cell nests (*arrows*).

section, segmental facial nerve resection and interposition of great auricular nerve graft were required. All had satisfactory results.

The differential diagnosis of a facial nerve canal paraganglioma in these patients included principally, facial nerve schwannoma (6), perineural spread from a parotid gland carcinoma (7), intratemporal benign vascular tumors (hemangioma/vascular malformation) (8), and jugular paraganglioma (9).

On CT, the tumor in both of our patients showed circumferential expansion of the descending facial nerve canal, with relatively smooth and well-preserved bone margins, similar to those caused by a schwannoma (6). The portion of the tumor beyond the facial nerve canal in the mastoid process, however, showed moth-eaten bone destruction typical of jugular paragangliomas (9). The moth-eaten bone changes were nonspecific, but spoke against a schwannoma (7). Both tumors extended anteriorly, one into the fundus of the external auditory canal and one into the facial recess of the tympanic cavity. This anterior direction of extension might have followed the course of the chorda tympani. One tumor grew inferiorly out of the stylomastoid foramen to indent the parotid gland, and raised the concern of a parotid gland carcinoma.

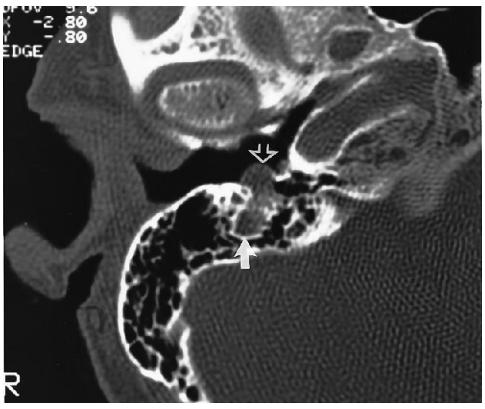

Fig 2. Primary paraganglioma of the right facial nerve canal. There is a mass widening the descending facial nerve canal (*closed arrow*). The mass has destroyed the posterior wall of the external auditory canal and protrudes into the hypotympanum (*open arrow*).

Neither of our patients underwent magnetic resonance imaging. However, both tumors might have been too small to show the salt and pepper pattern described by Olsen et al in paragangliomas larger than 2 cm (10). This salt and pepper appearance is thought to be caused by serpiginous areas of signal void, representing high vascular flow interspersed among areas of high signal intensity caused by slowly flowing blood and tumor cells.

In patient 1, the angiographic findings of marked hypervascularity, enlarged feeding arteries, and early draining vein were characteristic of a paraganglioma (11, 12). The exclusive supply of this tumor by branches of the posterior auricular and occipital arteries was in contrast to that of jugulotympanic paragangliomas that, until they extend into the mastoid, derive their supply from the ascending pharyngeal artery (11, 12). The rapid arteriovenous shunting helped to differentiate this tumor from a hemangioma (8).

In conclusion, paraganglioma of the facial nerve canal should be considered in the differential diagnosis in patients presenting with facial nerve palsy or pulsatile tinnitus. Paraganglioma should be considered in the differential diagnosis of lesions apparently arising in the facial nerve canal.

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