ISSN 0194-5998

2573

Otolaryngology–Head and Neck Surgery

Official Journal of the AMERICAN ACADEMY OF OTOLARYNGOLOGY-HEAD AND NECK SURGERY FOUNDATION, INC and the AMERICAN ACADEMY OF OTOLARYNGIC ALLERGY

OCTOBER 2003 VOLUME 129 NUMBER 4

ORIGINAL ARTICLES

For additional Original Articles, Review Article, Case Reports, and Clinical Photograph, see Table of Contents, p. 5A

Contents continued

CASE REPORTS

CLINICAL PHOTOGRAPH

READER SERVICES

Dumbbell-type intracranial meningioma presenting as an aural polyp and a neck mass

ALOYSIO AUGUSTO TAHAN DE CAMPOS NETTO, MD, RAQUEL ANN REDTFELDT, MD, C. GARY JACKSON, MD, JAMES L. NETTERVILLE, MD, and CARL R. HAMPF, MD, São Paulo, Brazil, and Nashville, Tennessee

Meningiomas are usually benign neoplasms that constitute 12.5% to 15% of all intracranial tumors. Only 5% to 8% of them occur in the cerebellopontine angle (CPA), and they account for 10% to 15% of the lesions occurring in this site. These tumors are believed to arise from the arachnoid cap cells and rarely extend out of their intracranial boundaries. There are reports of meningiomas extending to the paranasal sinuses, the cavernous sinus, the orbit, and the table of the calvaria, and these cases represent very rare clinical situations. The extension of a meningioma into the parapharyngeal space is extremely rare; only 10 cases have been reported in the world's literature. Only 1.3% of intracranial meningiomas extend into the parapharyngeal or cervical area. When they do, they most often travel via the jugular foramen. These tumors have been referred to as "dumbbell-type" due to their shape and propensity to grow both intracranially and into the parapharyngeal space.[1] Meningiomas extending to the external ear canal (EAC) are even more uncommon than the situations mentioned above. Only 3 cases of intracranial meningioma presenting as an aural polyp have been reported.[2,3] To the best of the authors' knowledge, only 1 case of an intracranial meningioma presenting as an aural polyp and a parapharyngeal space mass has been reported. We report a case of a dumbbell-type meningioma that presented as an aural polyp and a neck mass.

From the Núcleo de Otorrinolaringologia de São Paulo,The Ear Foundation at Baptist Hospital (Drs Netto and Redtfeldt), The Otology Group (Dr Jackson), the Department of Otolaryngology–Head and Neck Surgery, Vanderbilt University Medical Center (Dr Netterville), and Neurological Surgeons, PC (Dr Hampf).

Reprint requests: C. Gary Jackson, MD, FACS, The Otology Group, 300 20th Ave N, Suite 502, Nashville, TN 37203-2138; e-mail, ozcycle2.aol.com.

Otolaryngol Head Neck Surg 2003;129:457-60.

0194-5998/2003/$30.00 + 0
doi:10.1016/S0194-5998(03)00632-6

CASE REPORT

A 43-year-old white woman with congenital bilateral sensorineural hearing loss presented with complaints of poor-fitting left ear mold, left otalgia, neck stiffness, and headaches. These symptoms were progressive over a 2- to 3-year period. She also had new-onset instability and mild dysphagia. The otolaryngologic examination revealed a left EAC soft tissue mass and a left 2 × 4 cm high-level II firm, mobile neck mass. Tongue deviation to the left was exhibited. There was fullness of her left posterior pharyngeal wall. Both cerebellar function and cranial nerves tests were normal. Temporal bone computed tomography scan showed bony erosion and an intracranial mass. The magnetic resonance imaging (MRI) with and without gadolinium demonstrated 5.4 × 3.5 cm left, posterior cranial fossa mass, that was consistent with meningioma (Fig 1 and 2). The mass involved the left cavernous sinus, the sphenoid sinus, and the floor of the EAC. There was a mass effect on the fourth ventricle and brainstem at the level of the foramen magnum. The sagittal view disclosed a connection between the posterior cranial fossa lesion and the parapharyngeal mass via the jugular foramen (Fig 1).

The parapharyngeal mass measured 5 × 2.3 cm. Angiography demonstrated a relatively hypovascular lesion that involved the internal carotid artery (ICA) at the skull base juncture. Only the occipital artery was embolized. One day after embolization, the patient underwent single-stage subtotal multidisciplinary surgical removal of the neoplasm. Operative findings included meningioma involving cranial nerves IX, X, XI, and XII as well as the internal auditory canal. The jugular foramen was confirmed as the portal of entry into the neck. An infratemporal fossa, translabyrinthine, and transcochlear approach to the skull base was performed to remove the temporal bone neoplasm. The facial nerve was mobilized and preserved. Tumor was removed from the jugular vein and the

Fig 1. MRI with gadolinium: The sagittal view demonstrates the connection between the posterior fossa lesion and the parapharyngeal space mass via the jugular foramen.

transverse sinus. It was dissected from the petrous portion of the internal carotid artery. Clear normal dural margins were obtained at the middle and the posterior cranial fossae.

The neoplasm was then cleared from the neck and finally intracranially. In addition, a left selective neck dissection and partial parotidectomy were performed. Surgical reconstruction consisted of an abdominal free fat graft, a superficial temporal parietal fascial flap, and overseeing the EAC. The resection was subtotal in regard to the disease that invaded the cavernous sinus. Histopathologic examination of the neoplasm revealed transitional and meningothelial patterns of meningioma, with no nuclear atypia. There was, however, a significant perivascular and perineural invasion. The lymph nodes were all negative for metastatic disease. The postoperative course was complicated only by an episode of upper airway edema that necessitated intubation. This resolved with steroids, and the patient was extubated within 48 hours. The patient went home with nasogastric feeds. She underwent a type I thyroplasty with a good voice result, however, she did require a gastrostomy tube, and it is presently working on swallow rehabilitation.

Fig 2. MRI with gadolinium (coronal view): Tumor extending into the ear and the external auditory canal (left side).

DISCUSSION

According to Nager et al,[1] this case can be classified as either an intracranial meningioma extending extracranially or as a meningioma arising in a neural foramen with extracranial extension. Of the 371 intracranial meningiomas they reported, 20% extended extracranially, The most common extension was into the orbit (8%) and the least common was into the parapharyngeal, parotid or cervical region (1.3%). They mention the possibility of middle ear involvement and then subsequent extension into the EAC if a meningioma gains access to the temporal bone but cite no case in their review. An intracranial meningioma that extends into the external auditory canal is by far the rarest type of extracranial extension. The great majority of aural polyps are associated with epidermoid cholesteatomas or inflammatory disease of the middle ear, such as granulation tissue, necrosis, fibrosis, cholesterol granuloma, and xanthogranulomatous inflammation. Aural polyps that arise de novo include benign lesions such as exostosis, osteomas, benign ceruminoma, sebaceous cyst, and keloid.[3] Neoplasms are scarcely encountered as polypoid lesions of the ear. Of a series of 162 aural polyp biopsy samples, only 5 were neoplasms and only 1 of them (0.6%) was a meningioma.[3] Neoplasms that erode the temporal bone and present as external auditory canal polyps include chondroblastoma, giant cell tumor, histiocytosis, adenocarcinomas, basal cells, and squamous cells carcinomas and, less commonly, malignancies.

The present case is unique because it extended extracranially both inferiorly and laterally. We believe it reached the parapharyngeal space via the jugular foramen and that it penetrated the middle ear and EAC via the hypotympanum. It is difficult to determine whether the patient in this case report had a very aggressive tumor or a very late presentation was due to her congenital hearing loss, allowing the tumor to grow unnoticed. It is likely that both are true.

Meningiomas are not usually considered in the differential diagnosis for parapharyngeal space tumors or aural polyps. To the differential diagnosis of both parapharyngeal and EAC mass, extracranial extension of a meningioma should be added. Certainly one would consider glomus jugulare much higher on this differential. They can manifest the same signs and symptoms as in this case report because they inhabit the same location and have similar modes of spread. Meningiomas in this location have been considered more treacherous than glomus tumors, requiring a large margin of resection.[4]

In the vast majority of the cases, MRI with gadolinium can differentiate meningiomas of the posterior cranial fossa from other types of tumors in this site, because of its unique characteristics on this imaging examination. They appear as large, oval, and sessile masses that possess a broad dural base on the posterior petrous bone. In this case, the MRI was helpful in illustrating the direct connection between the intracranial and the extracranial space tumors, ruling out the possibility of 2 separate lesions or potential metastatic disease. Complete unstaged surgical resection is the treatment of choice to extirpate these neoplasms; however, the added morbidity of cavernous sinus surgery in this patient was a significant deterrent. The risk of additional potential cranial nerve dysfunction in this woman and its effect on the quality of her postsurgical survival served to limit complete resection feasibility.

Two alternatives emerge in this patient's follow-up: plan a second-stage procedure in the future to remove residual disease and/or treatment by external focused radiation therapy, should her disease progress.

Postoperative external radiation therapy has its role in controlling residual microscopic disease and has been shown to lower recurrence rates in subtotal tumor excision and in lesions that have a high rate of surgical morbidity.[5]

In conclusion, we report our experience with an exceptionally rare presentation for an intracranial meningioma. This case underscores the spectrum of behavior of intracranial meningiomas, which commonly grow slowly and silently or, as in this case, may be so aggressive as to be life threatening.

REFERENCES

1. Nager GT, Heroy J, Hoeplinger M. Meningiomas invading the temporal bone with extension to the neck. Am J Otolaryngol 1983;4:297-324.
2. Brugler G. Tumors presenting as aural polyps: a report of four cases. Pathology 1992;24:315-9.
3. Huin E, Tan KP. Case report: meningioma presenting as an aural polyp. Clin Radiol 1998;53:619-22.
4. Molony BT, Brackmann DE, William WM. Meningiomas of the jugular foramen. Otolaryngol Head Neck Surg 1992;106:128-36.
5. Maniglia AJ, Page LK. Posterior cranial fossa and temporal bone meningioma in a child, appearing as a neck mass. Otolaryngol Head Neck Surg 1979;87:578-83.