Meningioma Extending into the Middle Ear Mimicking Chronic Otitis Media

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Meningiomas arising in the middle ear or presenting as a middle ear lesion are extremely uncommon and only a few cases are reported in the literature. The case presented here illustrates the challenge in diagnosing extracranial meningiomas extending to the middle ear, since it mimics chronic otitis media.

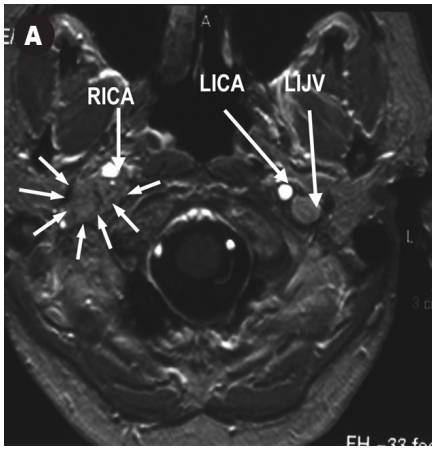
Patient Description

A 49 year old woman with a history of multiple primary breast and uterine carcinoma diagnosed and treated more than 6 years earlier was referred to the otolaryngology department because of right chronic otitis media and prolonged hearing loss on the right side. On referral, otoscopy on that side revealed an opaque tympanic membrane with solid fullness behind it (otoscopy was defined at that time as not resembling primary cholesteatoma). Audiometry showed mixed conductive and sensorineural hearing loss. High resolution computed tomography demonstrated clouding of the middle ear, the antrum, and the mastoid air-cell system, without significant destruction of the ossicles. In view of her history and the somewhat atypical otoscopic findings, the patient was scheduled for immediate middle ear exploration and mastoidectomy. At surgery only fluid was found in the mastoid. The attic and the middle ear were occupied by non-bleeding soft tissue suspected for neoplasm.

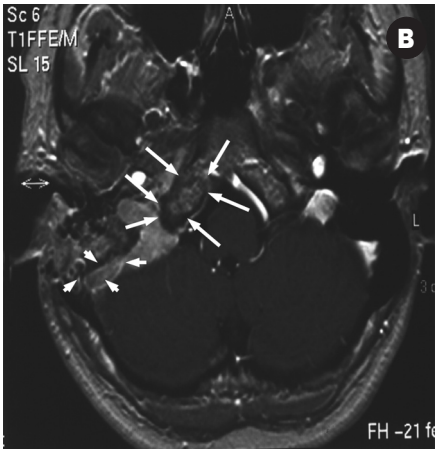
On frozen section, the possibility of benign non-epithelial tumor such as glomus tumor was raised; however, it was clinically negated due to the non-bleeding nature of the tissue found. Histological study of permanent sections (hematoxylin & eosin staining) demonstrated cellular aggregates with epithelioid appearance and whorling pattern composed of cells with indistinct cell borders and round-to-oval nuclei. Neither nuclear pleomorphism nor mitotic activity was found.

Immunohistochemistry showed positive staining with epithelial membrane antigen and vimentin but negative staining for S100 protein and cytokeratin. Morphological features and immunohistochemical profile of the tumor were typical for meningioma.

Magnetic resonance imaging performed only at this point demonstrated invasion of the posterior fossa by a lesion compressing the upper portion of the right internal jugular vein inferiorly and extending to the right internal carotid artery canal without compressing the artery [Figure A]. The lesion invaded the right clivus medially and extended laterally to the sigmoid sinus without invading it [Figure B]. The lesion was restricted to the temporal bone and unaccompanied by an intracranial



[A] MRI study (T1-weighted-images with gadolinium) demonstrating a gadolinium-enhanced lesion (arrows delineate the lesion), which inferiorly compressed the right internal jugular vein (RIJV); the left IJV is seen behind and lateral to the left internal carotid artery (LICA).



[B] T1-weighted-images with gadolinium demonstrating invasion of the lesion to the right clivus medially (large arrows) and extending to the sigmoid sinus laterally (small arrows).

meningioma. On T2-weighted images the lesion had a moderate intensity of signal.

Comment

When the otoscopic findings are somewhat unusual for chronic otitis media, a suspected tumor in the middle ear cleft should be considered to be any of the following: benign tumor such as glomus tumor, schwannoma or meningioma (primary or extending from the immediate adjacent area). Such unusual features may include an intact tympanic membrane with solid fullness behind it, not resembling primary cholesteatoma. Malignant metastasis to the middle ear cleft is a very rare condition; however, in the case presented here it was considered a realistic possibility due to the patient's known multiple primary malignant tumors. Breast cancer metastasis to the middle ear was suspected during assessment of the patient, even though it was reported as a very rare site for relapse and was eventually ruled out by histological study.

Meningiomas comprise the second largest group of primary brain tumors after gliomas, accounting for 18% of all primary intracranial neoplasms. Only an estimated 2% of meningiomas occur extracranially [1]; middle ear or temporal bone involvement of meningioma is extremely rare. Thompson et al. [2] reported 36 cases of ear and temporal bone meningiomas diagnosed between 1970 and 1996. The majority of patients were adults, most of them women (female/male ratio = 2/1). The mean age at diagnosis was 50 years. Hearing loss was found in 20 patients on presentation, as in our case. However, tumors affecting both the temporal bone and middle ear were reported in only seven patients [2].

The incidence of meningioma is reported to be relatively high among women with breast cancer. The case presented here supports this possible association. Reports of the possible expression of progesterone receptor in meningiomal tissues raise the possibility of a pathophysiological hormonal relationship [3].

Nager [4] in 1966 delineated two types of temporal bone meningiomas: type 1, being an extension of an intracranial meningioma with an extradural component, and type 2, which do not originate within the cranium but are primary extradural meningiomas. Primary extracranial meningiomas (type 2) were further classified according to their position relative to the cranium [5]. Type I tumors include lesions that are purely extracalvarial with no attachment to bone. Type II tumors are purely calvarial, being located entirely within the bone of the skull. Type III tumors correspond to calvarial tumors with extracalvarial extension, i.e., a tumor that is located within the skull but also has a soft tissue component that extends extracranially. Type II and type III tumors are subdivided into skull base (B) or convexity (C) tumors. The tumor in the presented patient can be classified as a type 2IIIB, namely, primary extradural skull base calvarial meningioma with extracalvarial extension.

Lang et al. [5] reported that patients with benign type IIB or IIIB tumors were much more likely to experience recurrence than those with benign type IIC or IIIC tumors. In their series, the recurrence rate was 26% in the former two categories and there was no recurrence in the latter two.

Treatment objectives for meningiomas of the cranial base include relief of neurological disability, prevention of clinical progression or recurrence, and minimizing morbidity. Based on the slow-growing nature of meningiomas in general, the long clinical history of the disease in the particular patients discussed (hearing loss in the affected ear was known in this patient for many years), and the lack of other neurological disability related to the disease, together with the history of the two malignant tumors the patient had already been diagnosed with (breast and uterine carcinoma), which were supposed to affect her life expectancy more significantly than the superficially invading nature of her meningioma in this case, it was decided to adopt an observational management approach based on serial future MRI studies. After 3 years of follow-up there was no significant deterioration in her clinical status or any changes in the tumor extension on the MRI studies.

In conclusion, in cases with no obvious otitis and/or suspicious otoscopic findings the physician should reduce their threshold for imaging and even for immediate surgical middle ear exploration.

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No one means all he says, and yet very few say all they mean, for words are slippery and thought is viscous

Henry Brooks Adams (1838-1918), American novelist, journalist, historian and academic