MIDDLE EAR MENINGIOMA ASSOCIATED WITH COCHLEAR IMPLANT: SPECIAL ATTENTION

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ABSTRACT

Meningiomas arising in the middle ear or presenting as a middle ear lesion are extremely uncommon and represent a diagnostic challenge. We report the case of a 79-year-old man, previously submitted to left ear cochlear implantation, treated at our Department for a suspected left chronic otitis media. Intraoperative findings and histology suggested a diagnosis of isolated left middle ear meningioma. The main radiological and pathological features of these lesions are discussed.

Keywords: meningioma, middle ear, temporal bone, differential diagnosis, surgery, histology

INTRODUCTION

Meningiomas arising in the middle ear or presenting as a middle ear lesion are extremely uncommon and represent a diagnostic challenge since they often mimic middle ear fibrous tissue or chronic otitis media (1-13). These tumors have non-specific clinical and radiographic features and consequently an accurate diagnosis requires histologic evaluation.

We report an isolated left middle ear meningioma in a patient submitted to homolateral cochlear implantation 10 years before.

CASE REPORT

In September 2012, a 79-year-old man was referred to the Department of Otolaryngology, “Carlo Poma” Civil Hospital of Mantova, for recurrent episodes of otalgia and “wet feeling” to his left ear.

He had already been treated for bilateral chronic otitis media at other ENT Departments. He had been submitted to right radical mastoidectomy 64 years before because of a cholesteatoma, and after several years he had undergone a left closed tympanoplasty for tympanosclerosis with intact tympanic membrane.

In 2002, because of his progressive and bilateral hearing loss he was submitted to a left ear cochlear implantation, but after a cerebral stroke in 2011 he lost any auditory benefit from it.

For the last 3 months the patient referred a numb feeling and an “unpleasing” sensation on his left temporo-parietal area. He referred otalgia and some episodes of otorrhea. Facial nerve function was preserved.

Otomicroscopy on that side revealed an opaque tympanic membrane with a posterior microperforation and a normal radical cavity on the right ear; au-
diometry showed a right severe mixed hearing loss and a left dead ear.

A CT demonstrated the previous mastoidectomy, a correct positioning of the implant array in the cochlear turns, and some soft-tissue density material within the middle ear; it revealed the absence of the ossicular chain and no dehiscence of the tegmen tympani and tegmen antri. No erosion of the inner ear or facial nerve canal was noticed. After clinical and blood evaluations, the patient was scheduled for cochlear implant removal surgery.

During the surgical procedure, the middle ear segment of the array appeared to be surrounded by poorly-bleeding fragmenting tissue adherent to the middle ear medial wall and the round window niche. The array was cautiously removed together with the soft tissue. The middle ear was accurately cleaned from the tissue. The round niche was filled with a muscle plug and the surgical access was closed.

Intraoperative frozen section histology ruled out the diagnosis of benign non-epithelial tumors such as glomus tumor. Histological study of permanent sections (hematoxylin & eosin staining) demonstrated nests and whorls of uniform small cells with moderate cytoplasm, fine nuclear chromatin and inconspicuous nucleoli. The stroma was densely sclerotic. Psammoma bodies were conspicuous. No mitotic activity, atypical features, nuclear pleomorphism or necrosis were noticed (Figure 1). Immunohistochemical analysis showed positive staining with vimentin and epithelial membrane antigen (EMA). S -100, Cam 5.2 and MNF-116 were negative (10). The morphological features were consistent with meningioma (WHO grade 1).

Patient’s postoperative recovery was excellent. No complication was noticed and the patient was discharged on the third postoperative day. He also referred complete resolution of the temporo-parietal “numb-unpleasing” feeling he reported preoperatively. Magnetic resonance follow-up has not shown any sign of meningioma recurrence so far.

**DISCUSSION**

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. Many cases of middle ear meningioma were not isolated middle ear meningiomas, but had primary intracranial lesions with involvement of the middle ear. The most common locations of temporal bone meningiomas are the tegmen tympani, jugular foramen, and internal auditory canal (3). Ectopic meningiomas may arise from ectopic meningothelial cells (4).

The presumed mechanism of extracranial meningioma formation is migration of meningothelial cells through potential routes such as the tegmen tympani, posterior fossa plate, and jugular foramen (2).

Embryologically, ectopic meningiomas are postulated to arise from either “punched off” embryonal arachnoid cells outside the skull and vertebra that lie along the line of fusion of the primitive nerve or bone sheaths, or it may arise as a result of differential maturation of pluripotent mesenchymal cells (2,7). The first theory is more widely accepted.

Histologically, meningiomas can be classified as globular ovoid (more common) and flat en-plaque
(more invasive) (2, 9). The WHO classifies four types of meningiomas: (1) meningotheliomatous or syncititial, (2) transitional or psammomatous, (3) fibrous and (4) angioblastic. The extracranial group of meningiomas is mainly the first or second variety (7, 9).

Nager (5) 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 (6). Type I tumors include lesions that are purely extradural 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 extra-cranially. 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.

Primary meningiomas of the temporal bones are more common in middle-aged females (9, 11). In spite of its tendency to invade the dura and the bone, meningiomas are considered benign and their symptoms are primarily caused by compression of the neighbouring neural structures.

Patients most frequently present with hearing changes, either sensorineural or conductive. Others features included otitis, polyp in the external ear, headaches, dizziness, unsteadiness, vertigo, disequilibrium, tinnitus, otalgia, bleeding, “metallic taste,” chronic cough, facial palsy or facial spasm, temporomandibular joint dysfunction or a neck mass (7).

Conventional meningioma does not usually cause diagnostic difficulties and its histological features are distinctive. However, rare variants in atypical sites can represent a diagnostic challenge for the pathologist. Biopsy may sometimes be extremely dangerous for the patient in certain histopathologic types of middle ear disease. Paragangliomas are highly vascularized lesions and a biopsy must be avoided because of the risk of dramatic hemorrhage. Biopsy of an endoauricular schwannoma of the facial nerve could result in a facial palsy.

The differential diagnosis includes schwannoma, paraganglioma, middle ear adenoma, mucoepidermoid carcinoma and adenocarcinoma. Most of these conditions are easily excluded by immunohistochemical profile and ultrastructural appearance (8).

Imaging procedures are performed in the majority of patients with suspected middle ear diseases and include conventional computed tomography (CT), angiography, and magnetic resonance imaging (MRI). In our case, the CT showed a middle-ear mass with diffuse cloudiness suggestive of severe mastoiditis or otitis media. Bone destruction or displacement can be noticed sometimes, together with focal bone remodeling by sclerosis or hyperostosis. CT scan may also reveal the leakage pathway through a posterosuperior defect of the mastoid bone near the sigmoid sinus as a result of destruction by the temporal meningioma (2).

An MRI scan with a T1-weighted, gadolinium-diethylenetriamine penta-acetic acid (Gd-DTPA) en-
hancement can be used to assess the nature of the lesion and the extent of the tumor in the temporal bone region.

Treatment of primary ear meningiomas is surgical excision (7). The nature of surgery depends on the extent of the tumour, which needs careful preoperative assessment with MRI scans. Although surgery is the treatment of choice, there is a number of challenges because of the invasiveness of the tumors and the complexity of the anatomy of the ear and temporal bone.

Leakage of cerebrospinal fluid after resection of the tumor is a potential risk when removing middle ear meningiomas, especially those tumors with intracranial involvement (11).

Radiotherapy has been proposed as a possible treatment but its role is yet to be proved. As meningioma is a slow-growing tumour, a follow-up of up to 10–15 years with yearly CT or magnetic resonance (MRI) scans along with clinical and audiological examinations has been suggested to rule out any recurrence. The overall prognosis is good.

The recurrence rate for meningiomas after total excision varies from 7 to 84%, depending on the number of years of follow-up (1-9).

Therefore, meticulous surgical extirpation of primary ear and temporal bone meningiomas is important to minimize the recurrence rate, without the need of adjuvant therapy.

CONCLUSION

No definite aetiopathogenesis of middle ear meningioma can be defined in our case. A possible role of cochlear implantation surgery in favoring meningioma development can be proposed, even though no definite conclusion can be drawn. Fibrous tissue is a common finding in the middle ear of patients submitted to CI. However, our experience suggests considering all possible differential diagnosis in case of soft tissue in the middle ear in patients submitted to cochlear implantation.

In our patient the tumor had a well-defined boundary and was confined to the middle ear without extension to the mastoid or skull base. A complete excision was carried out and no intraoperative CSF-leak was noticed. No complication was seen after surgery. Even though the progression of middle ear meningioma can be extremely slow, thus not requiring immediate surgical treatment, such disease should be considered in the differential diagnosis of soft tissue filling the middle ear in patients previously submitted to CI surgery.

REFERENCES
