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Temporal bone meningioma involving the middle ear: A case report

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Abstract. Meningioma is a common intracranial tumor involving the meninges. The localization of this type of tumor is rarely extracranial due to its typically low invasive properties. Furthermore, invasion of the middle ear is exceptional. The present study reported a case of meningioma extending into the middle ear from the middle cranial fossa through the tegmen tympani. The clinical and pathological characteristics, as well as the outcome of the patient, were described.

Introduction

Meningioma is a common intracranial tumour, accounting for 13-19% of all primary brain tumors (1). This tumor is generally encapsulated and benign (2). The symptoms of meningioma depend on the pressure of the tumor on the brain or spinal cord and the intracranial location of the tumor (3). Symptoms, such as seizures, single or multiple muscle twitches, spasms, loss of control of body functions, change in sensation and partial or total loss of consciousness, are associated with the different locations of the mass (1,4). Meningiomas accounted for ~33.8% of all the primary brain and central nervous system tumors reported in the United States between 2002 and 2006 (2). Furthermore, in recent decades, increased exposure to risk factors has determined an increase in the incidence of primary brain tumors, inclusive of meningioma, in several countries (5). The predominant risk factor identified is the exposure to ionizing radiation; however, other risk factors may be associated with the risk of meningioma, including elevated estrogen and/or progesterone hormone levels (2,6,7), head trauma (2), cell phone use (2,8), breast cancer (2), occupation (2), diet (2). Notably, a significant inverse correlation has been identified between meningioma and allergies (2,9); Linos *et al* (10) revealed that individuals with a history of allergy exhibited a lower risk of developing brain tumours than individuals with no history of allergy. Meningiomas have a higher incidence rate among female individuals, with a female to male ratio of ~2:1 (5). In addition, age-specific incidence rates indicate that risk increases with age. Prevalence rates for non-Hispanic individuals of African descent are marginally higher (6.67 per 100,000 persons) compared with Caucasian non-Hispanic and Hispanic individuals (5.90 and 5.94 per 100,000 persons, respectively) (2,4). The primary treatment strategy for meningiomas is total resection surgery if the tumor is benign and in an area of the brain where it can be safely and completely removed. Subsequently, radiation therapy is applied for the most malignant cases of meningioma or when surgery is not feasible due to the meningioma location.

Extracranial meningiomas are uncommon (accounting for <2% of meningiomas) (11), particularly those extending into the middle ear. The present study described a rare case of meningioma extending into the middle ear from the middle cranial fossa through the tegmen tympani. Written informed consent was obtained from the patient.

Case report

In May 2004, a 56-year-old woman presented to the Ear Nose and Throat Unit of Santa Maria delle Grazie Hospital (Naples, Italy) with an intense headache associated with vertigo that was treated with intravenous administration of 10% glycerol and 8 mg dexamethasone, with no results. After approximately two months, the patient had developed generalized seizure episodes. Brain magnetic resonance imaging (MRI) with gadolinium revealed the presence of a large (37x40x25 mm), T1-hypointense lesion widely invading the right middle cranial fossa with compression of the adjacent structures and dislocation of the middle cerebral artery.

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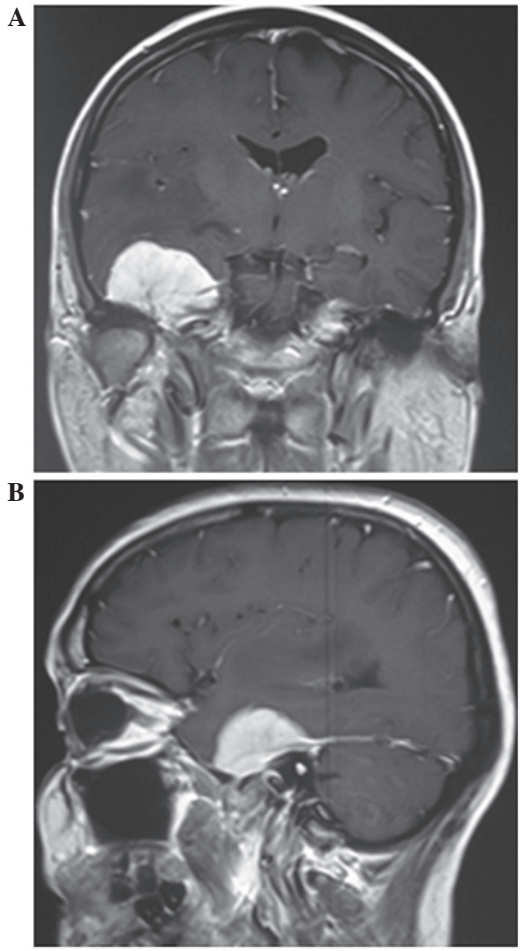

Figure 1. (A) Coronal and (B) sagittal brain magnetic resonance imaging with gadolinium performed in July 2004 revealing a right temporal mass widely involving the middle cranial fossa.

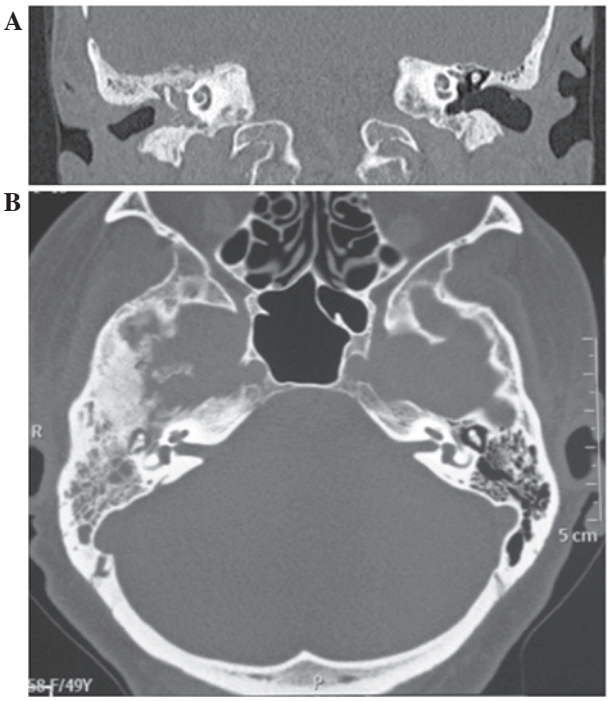

Figure 2. (A) Coronal and (B) axial computed tomography scans performed in November 2009 demonstrating an intratympanic mass enveloping the ossicular chain, without erosion.

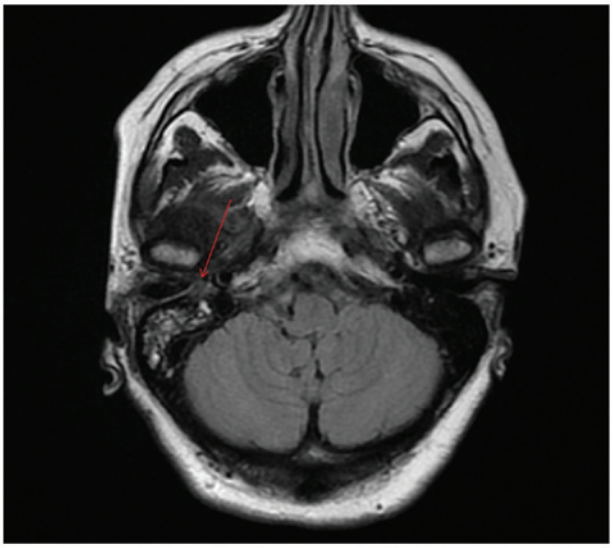

Figure 3. Pre-operative T2 brain magnetic resonance imaging performed in January 2010, showing a hyperintense tissue mass occupying the tympanic cavity. The arrow indicates the presence of the hyperintense mass.

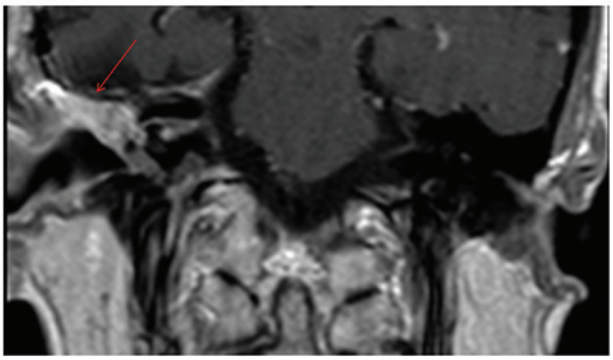

Figure 4. Early post-operative brain magnetic resonance imaging with gadolinium revealing the absence of the intratympanic mass in January 2010. The arrow indicates the area from which the intratympanic mass was removed.

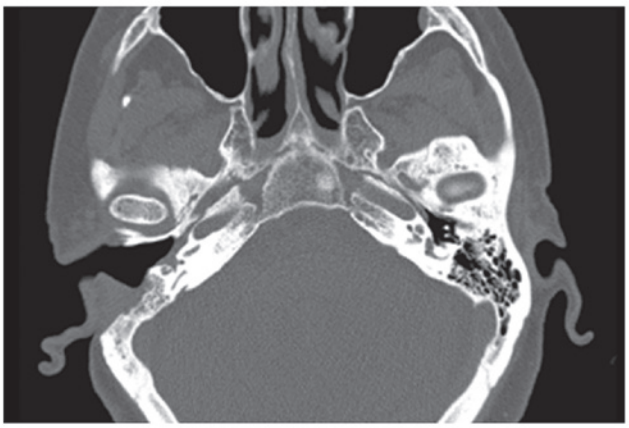

Figure 5. Axial temporal computed tomography scan performed in January 2015 demonstrating the absence of the middle ear tumor.

In addition, the lesion exhibited high contrast enhancement and a dural tail (Fig. 1A and B). Based on these radiological features of the lesion, a diagnosis of meningioma was deter-

mined. A right pterional craniotomy with partial extirpation of the lesion was performed, since the tumor presented a large and deep implantation base on the tentorial dura of the petrous bone roof.

The postoperative course was uneventful. Histopathological examination of the excised tumor determined a diagnosis of transitional meningioma. Subsequently, the patient underwent postoperative stereotactic radiotherapy (200 cGy, six days a week) for five weeks, to treat the residual tumor, and clinico-radiological follow-up.

In March 2006, two years after surgery, the patient underwent appositional transtympanic drainage in the right ear due to right chronic otitis media with effusion at the Complex Unit of Otolaryngology and Neck Surgery of Monaldi Hospital (Naples, Italy); however, no substantial clinical improvement occurred. The patient experienced progressive worsening of right-ear hearing loss, otorrhea and otalgia, despite frequent antibiotic therapies (with quinolones and cephalosporins). Therefore, the patient underwent petrous bone computed tomography (CT). The CT scan identified an intratympanic mass that enveloped the ossicular chain without erosion and thickening of the tegmen tympani (Fig. 2A and B).

In January 2010, the patient was admitted to the Ear Nose and Throat Unit of Santa Maria delle Grazie Hospital with right otalgia, otorrhea, vertigo and increasing right hypoacusis, which had occurred intermittently for the past 20 years. A right otomicroscopy identified granulation tissue in the drainage tube and the hyperemic, bulging and everted tympanic membrane. A pure tone audiogram determined right anacusis and left-sided sensorineural hearing loss. In addition, pre-operative brain MRI confirmed the presence of a hyperintense tissue mass, occupying the tympanic cavity (Fig. 3).

Consequently, a right canal-wall-down (CWD) tympanoplasty was performed. A granulomatous-like mass invading the tympanic cavity, mastoid antrum and oval window was removed (Fig. 4). Histological analysis confirmed the lesion to be transitional meningioma with identical histology to the previous tumor and thus, it was considered to be a section of the tumor that was resected in 2004.

The patient underwent clinico-radiological follow-up and, five years later, exhibited no evidence of middle ear tumor recurrence (Fig. 5).

Discussion

Meningioma is a common intracranial tumor, accounting for 13-19% of all the primary brain tumors (1). Meningiomas are slow-growing and originate from the arachnoid villi of the meninges (11). Extracranial meningioma are uncommon (accounting for <2% of meningiomas) (11), particularly those extending into the middle ear. Furthermore, the majority of intracranial meningiomas are benign (90%), with atypical (6%) and malignant forms (2%) occurring less frequently (12).

Meningiomas are typically diagnosed in adults aged >60 years and the incidence rate increases with age. In addition, these tumors occur more often in women than in men (13). Depending on the location of the tumor, patients can present with a variety of neurological symptoms, including headaches, speech problems, visual disturbances, cognitive deficits, motor deficits and epilepsy (4). In cases of temporal bone meningioma, otalgia is the most common feature, although hearing loss (conductive, mixed or sensorineural) (14), facial palsy and tinnitus are also common (15). A lower proportion of patients present with other cranial nerve palsies or vestibular symptoms (16).

In the current case, the patient exhibited a middle fossa meningioma that presented with epileptic seizures and symptoms of chronic otitis media. Chronic otitis media has been reported to occur in 16% of intratympanic meningiomas (17), possibly due to the obstruction of the Eustachian tube upon extension of the tumor into the middle ear cavity. Temporal bone meningioma scans follow multiple extension pathways: The tegmen tympani, the jugular foramen and the internal auditory canal (16). Based on their imaging characteristics, meningiomas are stratified into three groups with different primary locations and vectors of spread. The jugular foramen and tegmen tympani meningiomas are characterized by spread to the middle ear cavity. By contrast, internal auditory canal meningiomas spread to the intralabyrinthine structures (18). In the present case, meningioma primary to the tegmen tympani arose from the floor of the middle cranial fossa and spread inferomedially into the middle ear cavity. A number of radiological features (including thickening of the tegmen tympani, absence of bone and ossicular erosion and contrast enhancement of the lesion) (18) helped to distinguish meningioma from cholesteatoma in the current study. Furthermore, the jugular foramen, internal acoustic canal and labyrinthic area appeared healthy and free of disease.

Four histological subtypes of meningioma have been reported thus far: Meningotheliomatous, transitional, fibrous and angioblastic. Extracranial meningiomas are more often transitional or meningotheliomatous (19). In the present case, the extracranial lesion was diagnosed as a transitional meningioma.

Surgery is the treatment of choice for meningioma, with overall survival rates of 85, 75 and 70% at 5, 10 and 15 years, respectively. However, complete resection is often not considered to be possible, particularly in large tumors (20). Furthermore, in contrast to their intracranial counterparts, meningiomas involving the temporal bone do not often exhibit well-demarcated tumor margins (21).

The role of radiotherapy in the treatment of meningiomas remains controversial and is generally reserved for the adjuvant treatment of non-radically removed or recurrent meningiomas (19). Thus, in the present case, the patient underwent neurosurgical therapy with successive radiotherapy. Due to the persistence of the residual tumor in middle fossa for a number of years, the patient underwent a CWD tympanoplasty for the middle ear tumor.

In conclusion, temporal bone meningioma involving the middle ear is uncommon. Due to presenting similar symptoms with chronic otitis media, diagnosing meningioma is often difficult. However, imaging techniques can facilitate distinguishing between meningioma tumors and other diseases of the middle ear. In particular, in cases of hyperplastic chronic otitis media in patients with a previous meningioma, a temporal bone CT and MRI with gadolinium should be performed. Furthermore, treatment typically requires sequential neurosurgical and otosurgical approaches, followed by postoperative radiotherapy in cases of partial exeresis.

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