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CASE REPORT

Primary temporal bone secretory meningioma presenting as chronic otitis media

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Abstract We report an extremely rare case of a secretory meningioma primarily involving the temporal bone. A 56-year old female patient presented to us with a history of a chronic otitis media and unilateral hearing loss. Diagnostic investigations revealed a tumor arising from the temporal bone without signs of intracranial involvement. Histopathological examination showed a meningioma of the secretory type. The tumor was partially resected and serial imaging at follow-up revealed no extension of the tumor. No new symptoms developed 1 year after surgery. Secretory meningioma is a rare meningioma subtype and extracranial presentation in the temporal bone is very unusual. We present the first case of a primary temporal bone secretory meningioma in the otorhinolaryngological literature. As radical as possible surgical excision with serial imaging at follow-up is recommended.

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Introduction

Primary extracranial meningiomas of the temporal bone are rare tumors. In a review of the literature by 'O Reilly et al. in 1998 [1], only 26 cases of a primary temporal bone meningioma were identified. Secondary extracranial meningiomas are relatively more common, and they usually have a large intracranial component that causes neurological symptoms. However, primary extracranial meningiomas can exist with few or no symptoms and frequently present with unilateral conductive hearing loss or chronic otitis media.

Among meningiomas, secretory meningiomas represent a rare histological subtype. They are characterized by a unique differentiation of meningothelial cells resulting in the production of hyaline inclusions [2–4, 9]. This article describes a patient with a primary temporal bone secretory meningioma presenting as chronic otitis media.

Case report

A 56-year-old woman reported a 5-year history of otitis media of the right ear and subjective hearing loss. She was previously treated elsewhere with ventilation tubes and topical antibiotics. The patient presented to us complaining of intermittent purulent otorrhoea of the right ear, with otalgia. She denied any vertigo, imbalance or tinnitus. Her past medical history was unremarkable.

Physical examination revealed purulent otorrhoea of the right ear, with the ventilation tube in situ. Rinne testing was

negative on the right. Neurologic examination, including evaluation of the cranial nerves, and head and neck examination were normal.

Audiometry showed a mixed hearing loss of the right ear with a 40–50 dB conductive component. During the follow-up there was no significant improvement of signs and symptoms, and therefore, additional diagnostic imaging was suggested.

High resolution Computed Tomography (CT) in the axial plane with coronal reconstructions of the petrous bones revealed on the right side a soft tissue mass filling up the entire middle ear cavity and mastoid cells (Fig. 1). The ossicular chain was intact and not displaced. There was no evidence of destruction of the petrous bone or mastoid. These findings were primary compatible with chronic otitis media in which a cholesteatoma could not be ruled out.

A middle ear and mastoid exploration was performed via a combined aproach tympanotomy (CAT). After mastoidectomy, a soft tissue mass was found filling the antrum and middle ear, which bled easily on touch. A clear boundary between bone and tumor mass was not observed, suggesting infiltration. The tumor was removed as radical as possible, with preservation of the ossicular chain and facial canal. After surgery, the patient recovered promptly and improvement of hearing was seen.

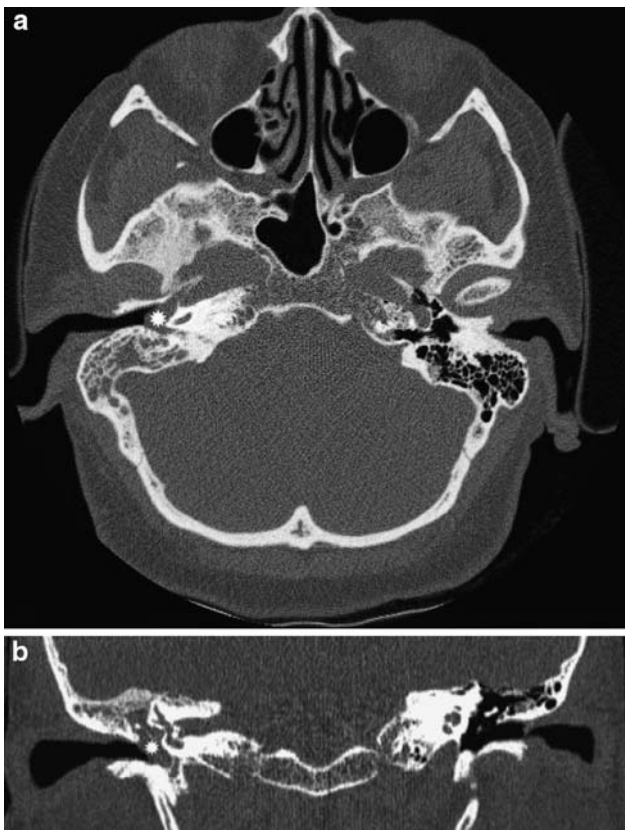

Fig. 1 High resolution Computed Tomography (CT) in the axial plane (**a**) with coronal reconstruction (**b**) of the petrous bones shows in the right ear middle ear and mastoid cells a soft tissue mass (*asterisks*). The ossicular chain was intact and not displaced. There was no evidence of destruction of the petrous bone or mastoid

Histopathological examination of the tumor revealed a meningioma with numerous intracellular pseudolumina with round eosinophilic periodic acid Schiff (PAS), epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) positive bodies, so-called pseudopsammomabodies (Fig. 2). This entity is known as a secretory meningioma.

Magnetic resonance imaging (MRI) of the head was performed to assess the extent of the tumor. In the middle cranial fossa on the right side, a homogeneous hyper-intense mass on the axial contrast enhanced T1-WI could be appreciated, in close contact to the temporal bone and with extension into the sphenoid sinus (Fig. 3a). Thickening and hyper-intense signal intensity on the T1–WI of the tentorium on the right side represent a so-called “dural tail”. On the coronal and sagittal T1–WI the extension of the mass into the petrous bone with spread to the epitympanum could be seen (Fig. 3b, c). The brain parenchyma was unremarkable.

At 6 months follow-up, MRI showed a slight increase of tumor volume in the temporal bone without any change of tumor enhancement of the tentorium. Follow-up at 1 year showed no changes in tumor volume or extension on MRI. No new symptoms developed 1 year after surgery. The patient will be followed for symptoms and signs at regular intervals with six monthly repeat of MRI.

Discussion

Meningiomas constitute about 18% primary intracranial tumors [5]. Extracranial extension is seen commonly, but

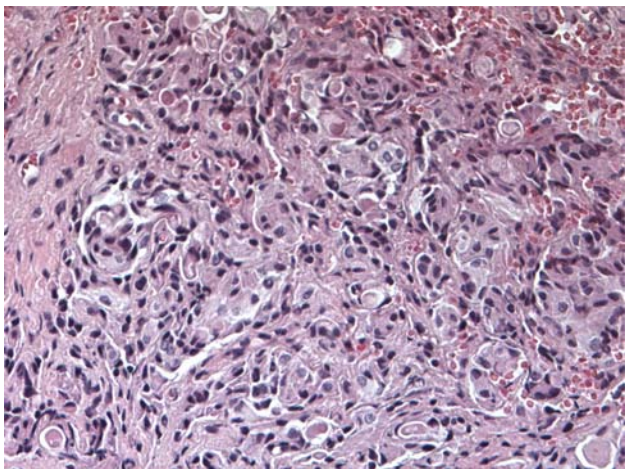

Fig. 2 Secretory meningeoma with many round pseudo-psammoma bodies (HE, original magnification ×200)

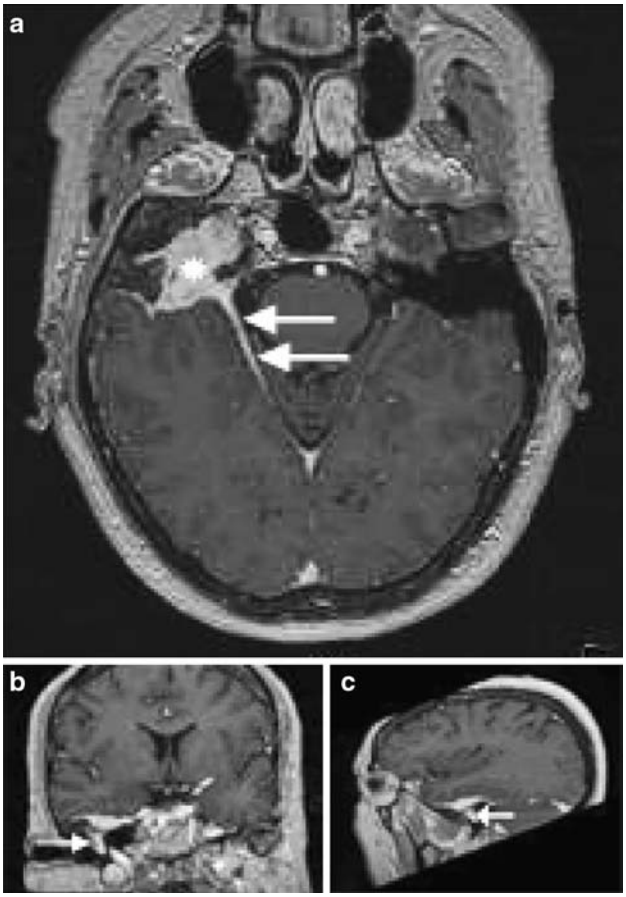

Fig. 3 **a** Post-contrast T1–WI MRI of the head. In the middle cranial fossa on the right side, a homogeneous hyper-intense mass on the axial plane can be seen (*asterisk*), in close contact to the temporal bone. Also thickening and hyper-intense signal intensity of the tentorium (*arrows*) represent the "dural tail". **b**, **c** On the coronal and sagittal images the extension of the mass into the petrous bone with spread to the epitympanon can be seen (*small arrows*)

primary extracranial meningiomas are rare. They usually present in the orbit, nasal cavity, paranasal sinuses and neck [6]. The temporal bone is an unusual site of presentation. The most common symptoms are sensorineural hearing loss, headache and vertigo [7]. In our case, the patient presented with chronic otitis media, which occurs in 16% all intratympanic meningiomas [7]. This is presumably the result of obstruction of the Eustachian tube due to tumor extension into the middle ear cavity. The differential diagnosis of otological symptoms caused by middle ear tumors includes cholesteatoma, adenoma, paraganglioma, adenocarcinoma, lymphoma or metastatic carcinoma [8].

Temporal bone meningiomas are slow growing tumors and do not metastasize. After complete resection, the prognosis is good, with 5-year survival rates of more than 80% [9]. However, as in our case, complete resection is often considered not possible and would have resulted in significant functional impairment of hearing and balance. As radical as possible surgery based on optimal improvement of clinical symptoms seems reasonable.

Meningiomas often infiltrate bone along the Haversian canals, and clear margins are rarely apparent. The recurrence rate of ear and temporal bone meningiomas was 28% in a large patient cohort by Thompson et al. [9]. Therefore it is essential to follow these patients with serial imaging studies to detect recurrence and to exclude intracranial involvement.

There is still debate about the place of radiotherapy in the treatment of meningiomas. For intracranial meningiomas, radiotherapy is frequently recommended as a safe and reliable adjunctive treatment for partially resected meningiomas [10, 11]. The role of radiotherapy for primary extracranial meningiomas has not been established.

According to the WHO classification there are 15 histologic subtypes of meningiomas (Table 1) [12]. Of these subtypes meningiothelial, fibrous and transitional (mixed) are the most common. The prognostic significance of these subtypes is very low, although some subtypes like clear cell meningiomas and papillary meningiomas are clinically aggressive [13].

Meningiomas of the secretory type are rare. Immunohistochemical analysis reveals epithelial differentiation

Table 1 [12] World Health Organisation (WHO) classification of meningiomas

	WHO grade
Meningiomas with low risk of recurrence or aggressive growth	
Meningothelial	I
Fibrous (fibroblastic)	I
Transitional (mixed)	I
Psammomatous	I
Angiomatous	I
Microcystic	I
Secretory	I
Lymphoplasmacyte-rich	I
Metaplastic	I
Meningiomas with greater likelihood of recurrence and/or aggressive behavior	
Atypical	II
Clear cell (intracranial)	II
Chordoid	II
Rhabdoid	III
Papillary	III
Anaplastic (malignant)	III
Meningiomas of any subtype or grade with high proliferative index and/or brain invasion	

features reflecting the pluropotential of cap cells. Intracellular lumina containing secretory lobules and eosinophilic (hyaline) inclusions are their distinguishing histological features [2–4]. In 1986 Alguacil-Garcia et al. [2] has determined the secretory features of these meningiomas thus suggested the name secretory meningioma. Formerly, these inclusions were referred to as "pseudopsammoma bodies" after a publication by Kepes et al. [3].

The frequency of secretory meningiomas is reported in the literature as approximately 1.6–3% [2, 4, 9]. Only 119 cases have been reported until present [14]. All were located intracranially. Only one case of a secretory meningioma primarily involving the temporal bone has been recently reported by Ereno et al. [15]. They presented a patient with a history of increasing left hypoacusis and sporadic vertigo. CT scan revealed a tumor occupying the mastoid, middle ear and external auditory canal. The tumor preserved the bone integrity without disruption of the temporal inner table, and was surgically removed. A typical secretory meningioma was diagnosed. The main emphasis in their article was on histopathology. Treatment and follow-up were not discussed.

Clinically, intracranial secretory meningiomas are associated with a good prognosis. In a large case study by Probst et al. [4], 31 patients with an intracranial secretory meningioma were followed after surgical removal of the tumor, with an average observation time of 23 months. No recurrences occurred during the follow-up, which ranged from 1 month to 8 years.

To conclude, meningiomas arising from the temporal bone are rare. And the secretory type is extremely rare. As radical as possible surgical excision with serial imaging at follow-up is recommended.

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