

## Case Report

# An *En Plaque* Meningioma of the Temporal Bone, Complicating with a Cholesteatoma, Chronic Otitis Media and an Intracranial Abscess

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## Abstract

We describe an initially misdiagnosed case of a *meningioma en plaque* of the temporal bone. The tumor had obliterated the external ear canal, induced a chronic inflammation and a cholesteatoma of the external and middle ear, and complicated with an external fistula and intracranial abscess formation. Massive bone involvement and scarce soft tissue component of the skull base *meningioma en plaque* often pose differential diagnostic challenges in imaging. We hereby review some typical CT and MRI characteristics that could help making the correct diagnosis.

## ABBREVIATIONS

CT: Computed Tomography; MRI: Magnetic Resonance Imaging; CPA: Cerebellopontine Angle

## INTRODUCTION

Meningiomas are mostly benign tumors that represent 13–26% of all intracranial neoplasms [1]. They have been found to be more common in the sixth and seventh decade of life and in female patients [2]. There are two well-established risk factors for meningioma development: a deletion in NF2 gene and exposure to ionizing radiation. Meningiomas developing after high dose irradiation of cerebral glioma, lymphoma, leukemia or metastases tend to have higher rate of atypism than in average population [3].

*Meningioma en plaque* is a rare clinical-biological form of meningioma, characterized by carpet-like growth along the meninges, and invasion of dura along with underlying bone hyperostosis [4]. The prevalence of *meningioma en plaque* is reported to be 2–9% of all meningiomas [4]. The most common site for en plaque meningiomas is sphenoid wing; Intraosseous meningiomas of the temporal bone make up less than 1% of all meningiomas [4,5].

The bony changes, caused by meningioma, are often called

“reactive hyperostosis”. Histological samples from meningioma en plaque, however, have revealed true tumor growth inside the bone, which is likely to induce periosteal new bone formation [6,7]. Affected bone typically becomes thick and sclerotic, narrowing the intraosseous spaces and canaliculae, and compressing cranial nerves and vascular structures. Obliteration of the external ear canal and middle ear spaces can further predispose for inflammation and cholesteatoma development [8,9].

## CASE PRESENTATION

A 76-year-old Caucasian female patient was referred to ENT surgeon because of a fistulous lesion and complaints of recurrent inflammation of the right ear. She had a previous medical history of atrial fibrillation, and a right temporal lobe low-grade astrocytoma that had been treated by surgical removal and radiotherapy 15 years ago. Years after she developed narrowing of the external ear canal and a fistulous lesion anterior and caudal to the right auricle that had been surgically excised. A postoperative CT scan showed thickening of the right temporal bone reminiscent of fibrous dysplasia or Paget’s disease. No signs of brain tumor recurrence were noted in CT.

Current physical examination revealed a hard prominence in the right mastoid process region. External ear canal was almost obliterated. Anterior and caudal to the right auricle was a fistula

that excreted pus and slightly hemorrhagic fluid. Laboratory findings were unremarkable except for the elevated C-reactive protein level (44mg/l). The patient underwent another CT scan in order to further characterize the nature and extent of the fistula, as well as its possible connection to skull base pathology and previous brain tumor. CT revealed marked sclerotic thickening of the right temporal bone, obliteration of the middle ear air spaces, and an expansile mass in the right cerebellopontine angle (CPA). In MRI, this CPA mass had cystic appearance, showed strong rim-enhancement after gadolinium administration and had restricted diffusion. It was first erroneously interpreted to be a cystic vestibular schwannoma.

Operation was performed by retroauricular translabyrinthine approach, extended with canal-wall-down tympanomastoidectomy, aiming at tumor removal, cleaning of mastoid process, and opening of the external ear canal. Surgical exploration revealed immature, highly vascularized bone, reminiscent of fibrous dysplasia. External ear canal was completely obliterated by fibrous tissue. The tympanic cavity, attic and antrum were filled by edematous mucous membrane and excessive soft tissue. Small cholesteatomas were found in the medial part of the external ear canal and in the tympanic cavity. Extended mastoidectomy towards the medial and posterior cranial fossa revealed pathological dura. Behind the posterior wall of the internal acoustic meatus, surprisingly, a capsulated collection of pus was found in the inner acoustic meatus and CPA. No soft tissue tumor additional to the abscess was noted in the intracranial space during surgical exploration. In the postoperative period, the patient received antimicrobial treatment with vancomycin 0.5 x 2 i.v. and ceftriaxone 2.0 x 1 i.m. Due to concomitant diseases and deterioration of her general condition the patient was transferred to the internal medicine department where she unfortunately died of pleuropneumonia on the 6-th postoperative day. Intraoperatively taken histological samples from the external ear canal, middle ear cavity and mastoid region showed inflammatory changes as expected. Samples from the temporal bone, however, contained Intraosseous meningotheial neoplastic growth, compatible with benign meningotheial meningioma.

Post mortem examination confirmed pleuropneumonia as the cause of death. Neuropathological examination revealed residual inflammation and post-treatment changes in the right temporal lobe and cerebellum. No signs of meningioma or recurrent astrocytoma appeared in the brain parenchyma. Cerebral pathology was excluded to be the cause for patient's death.

## DISCUSSION

Our patient had a history of previous radiotherapy for ipsilateral cerebral glioma that may have provoked the meningioma development. However, the exact extension of previous radiation field is not known to the authors, and the causal relation between previous radiation and current tumor remains hypothetical. The meningioma had an *en plaque* growth pattern inside the temporal bone, invading tympanic cavity and narrowing the external ear canal. Latter induced cholesteatoma development, which later complicated with chronic recurrent middle and external ear infection, formation of a fistula and an intracranial abscess.

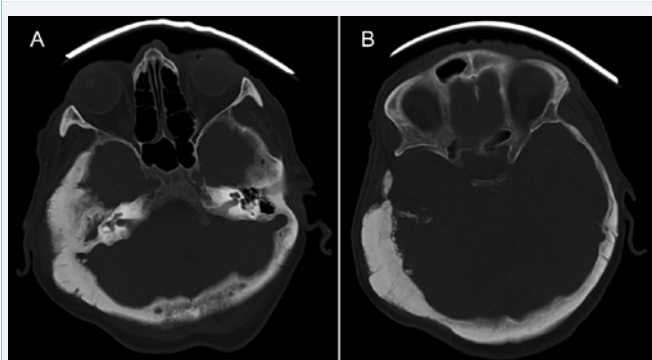
Intratemporal meningiomas can affect external ear canal, tympanic cavity and temporal bone per se or multiple subsides. Associated cholesteatoma is present in 25% of cases [5]. Histologically, intratemporal meningiomas are similar to their intracranial counterparts, exhibiting a variety of histological patterns, out of which the most prevalent is meningotheial variant [5]. Compared to the prominent bony changes, the intracranial soft tissue component of a *meningioma en plaque* is usually unremarkable, which often poses diagnostic problems for the radiologist. The diagnostic entities, that *meningioma en plaque* can be confused with, are fibrous dysplasia, osteoma, Paget's disease, chronic osteomyelitis, and sclerotic metastases of the skull - fibrous dysplasia likely being the most common misdiagnosis. In our case, radiological studies were performed elsewhere and bony changes were initially misdiagnosed as fibrous dysplasia, and CPA tumor as a cystic vestibular schwannoma.

Kim et al. have proposed some typical CT features suggestive for *meningioma en plaque* that could help discriminating it from fibrous dysplasia. These are: periosteal pattern of hyperostosis, inward bulging of skull vault, surface irregularity of the hyperostotic bone, and intracranial changes such as vasogenic edema of the brain adjacent to the hyperostotic bone [10].

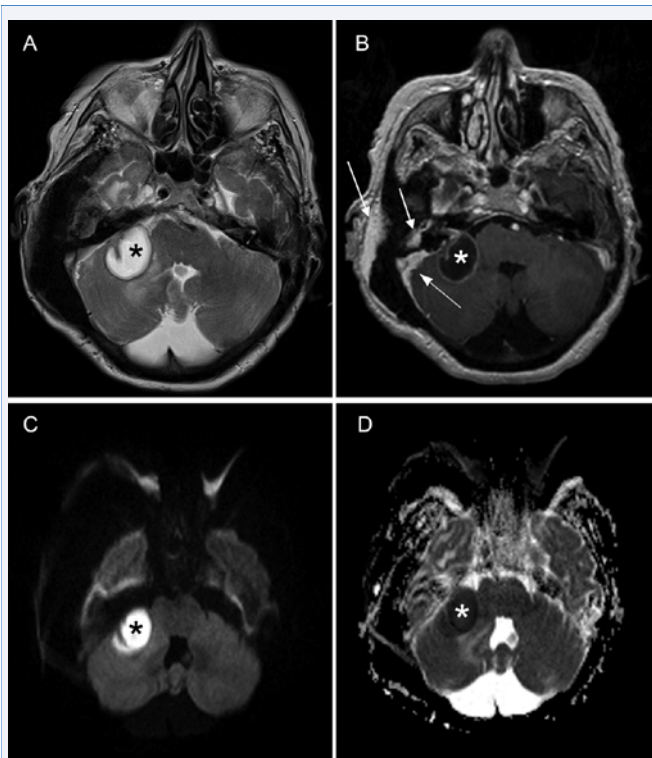
MRI features suggestive to *meningioma en plaque* are linear dural thickening and enhancing soft tissue around the affected bone, which correlate with the extraosseous soft tissue component of the tumor [11]. Latter aids differentiating meningiomas from intrinsic fibro-osseous skull base lesions which show no such extraosseous soft tissue component. The differential diagnosis of dural thickening and enhancement includes several diseases ranging from sarcoid, bacterial or granulomatous meningitis, lymphomas, gliomas, and post-irradiation changes to meningeal carcinomatosis. In actual diagnostic work-up, CT and MRI are completing each other: CT shows better details of the bone involvement whereas MRI demonstrates the dural enhancement and soft tissue components of the tumor.

Second look at our patient's CT images revealed typical features for *meningioma en plaque*, described by Kim: the irregular margins of the thickened bone, periosteal hyperostosis, and inward bulging of the inner table (Figure 1). In MRI, meningeal thickening and enhancement were apparent (Figure 2). None of these is typical for fibrous dysplasia, which in skull base usually causes bone expansion without changing its general shape and anatomical peculiarities. Another characteristic finding in fibrous dysplasia is the ground glass appearance of the affected bone that should not be confused with the bone thickening and sclerosis caused by a meningioma. In contrary to meningioma, fibrous dysplasia does not cause irregularities of bone margins nor does it cause extraosseous soft tissue growth around the affected bone [11].

Another confusing finding in our case was the mass-lesion at the CPA, initially misdiagnosed as a cystic vestibular schwannoma. Vestibular schwannomas with large cystic component may cause differential diagnostic difficulties, as opposed to typical solid and strongly enhancing counterparts. In this case, however, diffusion weighted imaging was performed and showed strong diffusion restriction inside the lesion, which is not characteristic for cystic vestibular schwannomas. Non-enhancing, extra-axial CPA lesion with restricted diffusion is typically an epidermoid



**Figure 1** Axial CT images of the skull base show thickening of the right temporal bone posterior to the old craniotomy defect. The middle ear space in (A) is obliterated by soft tissue. Typical bony CT-features for *meningioma en plaque* – irregular margins of the thickened bone, periosteal hyperostosis, and inward bulging of the inner table – are visible in (B).



**Figure 2** Axial MR images from the same level as shown in **Figure 1**. The CPA abscess is marked with an asterisk and shows high signal on T2-weighted image (A) and low signal as well as enhancing wall after gadolinium-based contrast material injection on T1-weighted image (B). Typical for an abscess is the high signal in DWI, b=1000 (C) and low signal in ADC map (D) which indicate restricted diffusion inside the lesion. In MRI, also the enhancing soft tissue component of the meningioma becomes visible along the inner and outer surface of the temporal bone and in the tympanic cavity and mastoid antrum (arrows in B).

with practically no other differentials [12]. Now, with this case, an otogenic intracranial abscess can be added to this differential diagnostic list. In radiological community, brain abscesses are generally known to present with such diffusion characteristics. This time, the unusual location of the abscess might have been the reason for misdiagnosis.

In conclusion, in the diagnostic work-up of sclerotic expansive skull base lesions a *meningioma en plaque* should be kept in mind. In the temporal bone, bone-invasive meningioma may lead to serious otological complications. By using some helpful imaging criteria, provided in the literature and summarized in this article, misdiagnoses could possibly be avoided.

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