medicinska revija
medical review

UDK: 616.289-006

Labus M. et al. ■ MD-Medical Data 2018;10(4): 213-216

Prikaz slučaja / Case report

OTOLOGIC DIAGNOSIS OF TEMPORAL BONE MENINGIOMA

OTOLOŠKA DIJAGNOZA MENINGEOMA TEMPORALNE KOSTI

Milica Labus[1], Milan Erdoglija[1], Biserka Vukomanović Đurđević[2]

Correspondence to:

Milica Labus, MD
Department of Otorhinolaryngology
Military Medical Academy
Crnotravska 17, 11000 Belgrade, Serbia
Phone: +381 64 1840219
E-mail address: krstic.milica@yahoo.com

[1] Department of Otorhinolaryngology, Military Medical Academy, Belgrade, Serbia
[2] Institute for Pathology, Faculty of Medicine, Military Medical Academy, Belgrade, Serbia

Ključne reči

Meningeom; Ektrakranijalni tumor; Slabljenje sluha; Tinitus; Glavobolja

Key words

Meningioma; Extracranial tumors; Hearing loss; Tinnitus; Headache

Sažetak

Meningeomi su klinički i histološki benigne, solitarne mekotkivne promene, najčešće lokalizovane intrakranijalno. U ovom izlaganju, predstavljamo neuobičajene slučajeve ekstrakranijalnih meningeoma lokalizovanih u temporalnoj kosti kao svojevrstan dijagnostički izazov. Pacijenti su bili hospitalizovani radi razjašnjenja etiologije levostrane glavobolje praćene vrlo specifičnim otološkim simptomima: istostranim gubitkom sluha, zujanjem i osećajem punoće u uvu. Tokom dijagnostičkih procedura, neočekivani nalazi su otkriveni kontrasnom kompjuterizovanom tomografijom (CT) temporalne kosti i potvrdjeni histopatološkom analizom kao meningeomi. U dodatku će biti razmotrene kliničke, radiološke i patohistološke karakteristike ekstrakranijalnih meningeoma.

INTRODUCTION

Meningiomas are frequent intracranial tumors, with incidence of 13-19% among all primary brain neoplasms. This tumor is mostly encapsulated and benign, a wide range of symptoms it causes is associated with a different tumor localization. Extracranial meningiomas, which include the temporal bone meningiomas, are rare with less than 1% of the prevalence among all meningiomas [1, 2].

Temporal bone meningiomas are more frequent in women than in men (2: 1). The average age in which they occur is 49.6 years [3]. Clinically, patients are complaining of some kind of deafness, either sensorineural or conductive, otalgia, purulent otorrhea, bleeding, facial nerve palsy, tinnitus, headache, vertigo and instability which are present for a few years back. The average time from the appearance of symptoms to the final diagnosis is 24.6 months. However, it is described a statistically significant difference between the average duration of symptoms and the anatomic site of distribution: external auditory canal (47.3 m); middle ear (24.7 m); temporal bone (20 m); and mixed locations (7.6 m) [2, 3]. Temporal bone meningeoma clinically looks like a chronic suppurative otitis more than temporal bone tumor. (Figure 2a)

Diagnostic imaging procedures include conventional skull radiographs, computed tomography (CT), angiography and magnetic resonance imaging (MRI). Given the site of origin, spreading of the tumor and specific CT / MR findings, meningiomas are divided into three subgroups: those that originate from tegmen tympani, jugular foramen (JF) and internal auditory canal (IAC) [4]. Characteristic CT features of tegmen tympani meningioma includes thickening of the tegmen tympani; a middle ear cavity soft-tissue mass, without erosion or destruction whereas MR imaging findings are linear dural enhancement along the floor of the middle cranial fossa and homogenous soft-tissue enhancement. JF meningioma CT features includes a centrifugal pattern of spread to the regional skull base, without bone thickening, while MR imaging showes centrifugal skull base infiltration with more intense enhacement of the intratympanic soft-tissue mass than tumor below the skull base. IAC MR imaging findings are uniform enhancement of all intralabyrinthine structures. Common to all temporal bone meningiomas is the presence of an intracranial enhancing dural-based component and preserved internal architecture of involved bone [4]. Obviously, CT and MRI are completing each other: CT shows better details of the bone involvement while MRI demonstrates the dural enhancement and soft tissue components of the tumor.

Correct diagnosis of ear and temporal bone meningiomas cannot be made without histologic evaluation. As well as intracranial, meningiomas of temporal bones shows differ-

ent histological patterns. The immunohistochemical profile of ear and temporal bone meningiomas is identical as in intracranial lesions, with vimentin, epithelial membrane antigen and S-100 protein immunoreactivity. According to the WHO classification, there are 15 subtypes of meningioma [5], although most cases belong to meningothelial or psammomatous meningiomas.

Surgical excision is a treatment of choice if the tumor is located so that it can be safely and completely removed. However, a multidisciplinary approach that involves radiotherapy, applied after surgery for temporal bone tumor forms or radiotherapy is primary kind of therapy if the surgical treatment is not applicable, is often necessary because of the size and extension of temporal bone tumor process [6].

Five-year survival rate is 80%, while five-year survival rate without relapse (for operatively treated patients) is 77.1%. Ten-year survival rate is 68.6%. Recurrences are recorded in less than 20% of cases. Generally speaking, the prognosis of primary meningiomas of the ear and temporal bone looks great with a survival of 15.5 years [3, 7].

There is no correlation between clinical, radiological or pathohistological characteristics with a prognosis of disease development.

CASE REPORTS

Case report No I

A 50-year-old female patient, previously healthy, complained of a progressive, one year present, headache. The headache was consistently left-sided, treated effectively with medications and was not accompanied by nausea or vomiting. As the symptoms progressed, she noticed a left sided gradual hearing loss and an ear fullness, followed by tinnitus on the same side. Primarily patient was observed by neurologist who recommended diagnostic imaging of the head. Computerized tomography (CT) of the head showed an expansive mass that completely filled the left temporal bone with middle fossa brain extension (Figure 1a).

Otomicroscopic examination revealed a soft-tissue mass arising from the superior and anterior wall of the left external auditory canal, significantly narrowing it down (Figure 1b).

Pure tone audiometry showed a conductive hearing loss on the left while tympanogram was type B on the same side.

Histopathological examination showed meningothelial tumor cells with regular, vesicular nuclei and nuclear pseudoinclusions; with syncytial architectonics and tendency to form whorls. Immunohistochemical analysis were positive for vimentin and epithelial membrane antigen (EMA) (Figure 1c).

Due to the size and extension of the tumor, surgical treatment was not possible, so the patient was referred to the radiotherapy. Two-years follow up showes reduction and stagnation of the tumor.

Case report No II

A 52-year-old male patient, with hypertension and history of stroke with left-sided weakness that occured 6 years before presented to ORL specialist because of the impaired hearing. He also complained on occasional purulent, fetid otorrhea, frequent left-sided headaches and occasional dizziness. Otomicroscopic examination showed thickened, sclerotic tympanic membrane on the left side (Figure 2a). Computerized tomography (CT) of the head showed a sclerotic mastoid processus on the left. In 2013, canal-wall down tympanoplasty was performed, the histopathological examination of the harvested material confirmed the presence of a cholesteatoma. Two years after surgery, patient complained of headaches in the left temporal region, same sided tinni-

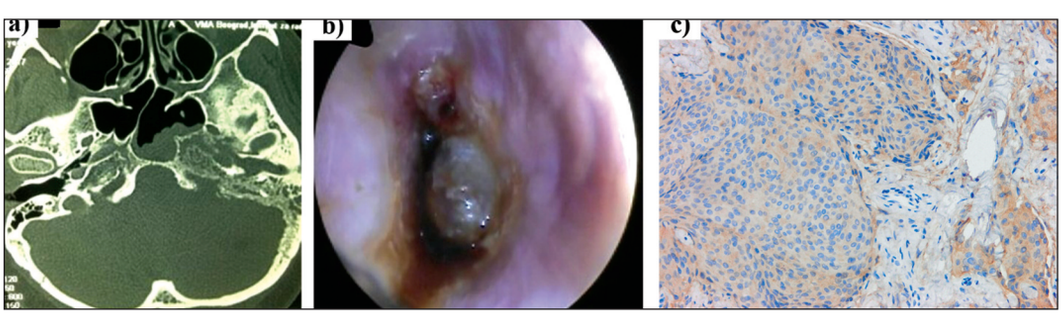

Figure 1: *a) CT scan of a temporal bones: expansive mass that completely filled the left temporal bone with middle fossa brain extension; b) Otomicroscopic examination: a soft-tissue mass arising from the superior and anterior wall of the left external auditory canal, significantly narrowing it down; c) EMA x20 - tumor cells show expression on the Epithelial Membrane Antigen*

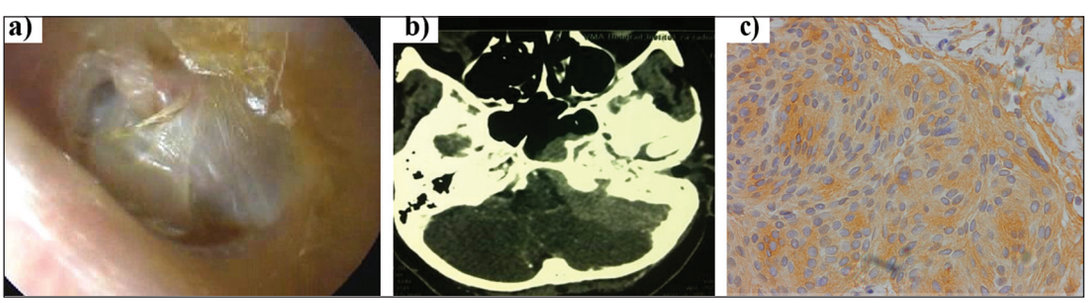

Figure 2: *(a) Otomicroscopic examination shows sclerotic tympanic membrane on the left; b) CT scan of temporal bones: hyperostosis of the sphenoid and temporal bone on the left side; c) Vimentinx40- Tumor cells express an immunohistochemical marker*

tus, dizziness and slight balance disorder. Otomicroscopic examination revealed wet pretympanic secretion and attic perforation on the tympanic membrane. CT scan of temporal bones showed soft-tissue attenuation in middle ear and antrum on the left and hyperostosis of the sphenoid and temporal bone at the same side (Figure 2b).

Pure tone audiometry showed mild perceptive hearing loss on the right, while left was present mixed hearing loss (70-80 dB).

Retympanoplasty of the left ear was performed in 2015. Intraoperatively, a severe hyperostosis of the middle ear cavity had been seen, so CT scan with contrast was performed. Images revealed soft-tissue mass at the floor of the posterior cranial fossa

Histopathological examination of tumor biopsy revealed an ectopic intraosseous meningioma. Immunohistochemical analysis were positive for vimentin, Epithelial Membrane Antigen and PR8 (Figure 2c).

The patient was then referred to a neurosurgeon for further diagnostic and treatment.

DISSCUSION

Otologic diagnosis of the temporal bone meningiomas can be quite a challenge. The literature is limited to isolated case reports with a few literature reviews. The largest one is retrospective study of 36 cases of primary ear and temporal bone meningioma [3]. Symptoms presented were non-specific, with the hearing loss as the most frequent complaint, followed with otitis, otalgia, headache and tinnitus, similar to our patients. The duration of symptoms was from 2 weeks to 192 months, 24.6 in average, resembling our data.

Roentgenographic procedures used in the majority of patients included conventional skull radiographs and computed tomography (CT); most frequent findings were diffuse opaqueness with fluid levels without destroying bone structures (common for mastoiditis or middle ear infections) and remodeling of the bones in the direction of sclerosis or hyperostosis. These findings may be differently diagnosed as primary bone pathological changes, most often as fibrotic dysplasia, osteoma, Paget's disease and skull-based metastases [8]. MRI scans with Gd-DTPA enhancement was used in some patients to underline the nature of the mass-features suggestive to meningioma were linear dural thickening and enhancing soft tissue around the affected bone. The differential diagnosis of these findings includes several diseases ranging from sarcoid, bacterial or granulomatous meningitis, lymphomas, gliomas, and post-irradiation changes to meningeal carcinomatosis [9]. Fourteen patients had radiographic studies interpreted as normal.

Taking into account the rarity of these tumors-only 2.0% of the benign or malignant ear and temporal bone tumors, and their nonspecific clinical and radiographic features, it is not surprising that the mean duration of clinical symptoms is 2 years [3].

Once it is correctly diagnosed, a meticulous surgical extirpation seems to be a treatment of choice. Given the average time needed for the diagnose which leads to the extension of the disease and the complex anatomy of the temporal bone, it is clear that surgical excision that would preserve cranial nerve fuction and hearing can be quite a challenge. Multidisciplinary aproach that includes radiotherapy is often needed. One of our patients was surgicaly treated by neurosurgeons, and as far as we know, is still without relapse. Other one was treated with radiotherapy because of the invasiveness and extension of the tumor. Thompson at al. reported that all of their patients were treated by partial or complete surgical excision, so none of them received radiation therapy. Follow-up showed a recurrence rate of 28%, with no metastatic dissemination of meningiomas recorded.

According to available literature data, none of the clinical, radiographic or pathological features is in correlation with the prognosis of the disease, although recurrences have been more frequent in women. Thompson at al. reported that deaths occurred as a result of involvement of the vital structures of the ear and region of the temporal bone or the complications of the operation, rather than the aggressive nature of the tumor.

CONCLUSION

We have presented rare cases of temporal bone meningioma with their clinical, radiological and pathological characteristics. Ear and temporal bone meningiomas are rare, but the otorhinolaryngologist should consider it in the differential diagnosis of middle ear pathology. Presenting symptoms are non-specific, which can lead to wrong diagnosis and delayed treatment. Radiological visualization combined with pathohistological analysis with immunohistochemical staining is essential for diagnosis. Periodic monitoring of the patient is necessary in order to prevent relapse.

Abstract

Meningiomas are clinically and histologically benign, solitary and soft tissue neoplasms, most commonly localized intracranially. In this report we present unusual cases of extracranial meningiomas localized in temporal bone as a sort of diagnostic challenge. Patients were addmited in order to clarify etiology of their left-sided headaches, which were followed by very specific otologic symptoms: same sided gradual hearing loss, tinnitus and feeling of ear fullness. During the diagnostic procedures, unexpected findings were revealed by contrasted computerized tomography (CT) of temporal bones and confirmed by histopathological analysis as meningiomas. In addition clinical, radiological and pathohistological characteristics of extracranial meningiomas and their differential diagnosis will be considered.

LITERATURE

1. Wiemels J, Wrensch M, Claus EB. Epidemiology and etiology of meningioma. J Neurooncology 2010 Sep; 99 (3):307-14.

2. O'Reilly RC, Kapadia SB, Kammerer DB. Primary extracranial meningiomas of the temporal bone. Otolaryngolgy Head Neck Surgery 1998; 118(5):690–694.

3. Thompson LD, Bouffard JP, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: a clinicopathologic study of 36 cases with a review of the literature. Modern Pathology 2003 Mar; 16(3):236-45.

4. Hamilton BE, Salzman KL, Patel N et al. Imaging and clinical characteristics of temporal bone meningioma. AJNR 2006 Nov-Dec; 27(10):2204-9.

5. David N. Louis, Arie Perry, Guido Reifenberger, Andreas von Deimling, Dominique Figarella-Branger, Webster K. Cavenee et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. Acta Neuropathologica, 2016 Jun; 131(6):803-20.

6. Burger PC. Intracranial Meninges. In: Burger PC, Scheithauer B, Vogel FS. Surgical Pathology of the Nervous System and its Coverings. 4th ed. Philadelphia: Elsevier; 2002:p 49–71.

7. Mirimanoff RO, Dosoroetz DE, Linggood RM, Ojemann RG, Martuza RL. Meningioma: analysis of recurrence and progression following neurosurgical resection. Journal of Neurosurgery 1985 Jan; 62(1):18–24.

8. Shapiro S, Cassis A. Differentiating Hyperostotic Temporal Bone Meningioma En Plaque and Fibrous Dysplasia on Computed Tomographic Imaging. JAMA Otolaryngology— Head & Neck Surgery 2017 Mar 01; 143(3):316-17.

9. Watts J, Box G, Galvin A, Brotchie P, Trost N, Sutherland T**.** Magnetic resonance imaging of meningiomas: a pictorial review**.** Insights Imaging 2014 Feb; 5(1):113–22.

■ Rad primljen: 06.11.2018. / Rad prihvaćen: 06.11.2018