Cancer Treatment and Research Communications 42 (2024) 100854

Contents lists available at ScienceDirect

Cancer Treatment and Research Communications

journal homepage: www.sciencedirect.com/journal/cancer-treatment-and-research-communications

En plaque meningioma of the temporal bone: A systematic review on the imaging and management of a rare tumor

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ARTICLE INFO

Keywords:
Meningioma
En plaque
Meningioma en plaque
Temporal bone
Lateral skull base

ABSTRACT

Objective: To review the published cases of meningioma en plaque of the temporal bone (TB-MEP), to gather evidence on the clinical assessment and management of this rare entity.
Methods: Following PRISMA statement recommendations, 383 abstracts were screened independently by two authors. Inclusion criteria were articles of human patients affected by TB-MEP; English or Italian language; availability of the abstract articles unrelated to TB-MEP, guidelines and systematic reviews were excluded. Only full-text articles reporting the diagnostic work-up and the management of the TB-MEP were considered for analysis.
Results: A total of 12 articles were included, for a total of 25 patients with a mean age of 52 years (range: 31–71). The average time elapsed between the onset of symptoms and the actual diagnosis of TB-MEP was 36.5 months (range: 2–120). In most cases, the pathology presented with hearing loss (80 %), often accompanied by effusive otitis media (52 %), aural fullness (32 %), and tinnitus (32 %). The main Computed Tomography (CT) findings were hyperostosis (76 %), hairy appearance of bony margins (16 %), involvement of the mastoid and middle ear (48 %). Magnetic Resonance Imaging (MRI) revealed dural enhancement (28 %), temporal hyperostosis (20 %), a clearly enhancing extra-axial mass (28 %), compression of the surrounding vasculo-nervous structures (8 %) and the possible involvement of the temporal lobe (8 %). Forty percent of patients underwent various medical and surgical treatment before reaching the diagnosis. Forty-four percent of patients were sent to definitive surgical treatment, 44 % to follow-up while 8 % received radiotherapy.
Conclusions: Meningioma en plaque (MEP) is a rare tumour, particularly when it originates within the temporal bone. Appropriate imaging in patients complaining of audiological sign and symptoms is mandatory to avoid diagnostic delays, avoid inappropriate surgical procedures, and adopt the appropriate treatment.

Introduction

The term "meningioma" was originally introduced by Harvey Cushing in 1922 to refer to the typically benign tumors that arise from the lining cells of the arachnoid. He described two distinct entities: the globular and the en-plaque types [1].

Meningioma en plaque (MEP), a rarer subtype (2–9 % of all meningiomas), is characterized by a “carpet-like” growth along the dura mater, with diffuse and extensive dural involvement, in contrast to the mass-like pattern of the globular type [2] While it most commonly affects the spheno-orbital region, MEP could be also found along the cerebral convexity, the foramen magnum, and the temporal bone [3].

MEP involving the temporal bone (TB-MEP) represents a unique clinical entity, due to its peculiar radiological features and frequent diagnostic delays, often resulting from the inadequate work-up following nonspecific signs or symptoms, such as hearing loss and tinnitus. Moreover, TB-MEP poses significant surgical challenges, given its potential for extension to adjacent foramina and neurovascular structures, leading to significant morbidity [3].

This tumor is thought to originate from arachnoid cells of the

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https://doi.org/10.1016/j.ctarc.2024.100854

Available online 26 November 2024

posterior or middle cranial fossa with secondary invasion of the TB, while even rarer cases of primary MEP arise from arachnoid cell nests located within the TB itself [4,5].

Despite the clinical importance of TB-MEP, current literature on this tumor is based on case reports and small case series, and no overviews about the diagnosis and management of this clinical entity have been published. This gap has led to ongoing debates about optimal management strategies, including surgery, radiotherapy, and watchful waiting with periodic scans.

This systematic review aims to summarize current evidence on clinical assessment, imaging, and treatment strategies for TB-MEP, offering clinicians guidance for the management of this insidious disease.

Methods

A systematic review of the literature was performed following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [6].

The search was run using the terms “Meningioma En Plaque” and “Temporal Bone Meningioma on PubMed/MEDLINE, Scopus and Web of Science databases, resulting in the following search string: *("enzymology"[MeSH Subheading] OR "enzymology"[All Fields] OR "en"[All Fields]) AND ("plaque s"[All Fields] OR "plaque, amyloid"[MeSH Terms] OR ("plaque"[All Fields] AND "amyloid"[All Fields]) OR "amyloid plaque"[All Fields] OR "plaque"[All Fields] OR "dental plaque"[MeSH Terms] OR ("dental"[All Fields] AND "plaque"[All Fields]) OR "dental plaque"[All Fields] OR "plaques"[All Fields]) AND ("meningioma"[MeSH Terms] OR "meningioma"[All Fields] OR "meningiomas"[All Fields])*.

After running the above search string in September 2023 and removing duplications, the 383 abstract titles and abstracts obtained were screened independently by two authors (AB, GM), who subsequently met and discussed disagreements on citation inclusion. When disagreement occurred during the screening process, it was overcome through detailed discussion between the two reviewers, each presenting their rationale. If consensus could not be reached, a third senior reviewer (GM) was consulted to reach the final decision.

The two authors independently assessed the quality of the included studies using an assessment tool for case series and case reports [7] which considers four domains (selection, ascertainment, causality, and reporting) and provides eight questions to aid quality score, as reported in Table 1. Studies were rated as having a low, moderate, or high risk of bias. Only articles with low or intermediate risk of bias were included. The inclusion of studies with an intermediate risk of bias was a shared decision among authors, since excluding such studies would have led to the loss of valuable scientific information, thus preventing a thorough literature review on this rare entity.

Exclusion criteria for abstracts were language other than English and Italian, topics unrelated to TB-MEP, systematic reviews and guidelines, and articles not involving human patients.

Afterwards, the full-text articles identified underwent a second screening by the same two authors. Full texts were included if the diagnosis of TB-MEP was confirmed. Articles whose full-text versions were not available were excluded. A further manual check of the references included in the articles was performed and the final number of articles included in the present review was defined.

Since MEP has been inappropriately referred to as an intraosseous meningioma, due to its frequent wide osseous involvement, an additional search was conducted using the following string: *("intraosseal"[All Fields] OR "intraosseous"[All Fields] OR "intraosseously"[All Fields]) AND ("meningioma"[MeSH Terms] OR "meningioma"[All Fields] OR "meningiomas"[All Fields]) AND ("temporal bone"[MeSH Terms] OR ("temporal"[All Fields] AND "bone"[All Fields]) OR "temporal bone"[All Fields])*.

The flowchart of the selection process is described in Fig. 1.

The general results regarding patients’ clinical presentation, diagnostic work-up, treatment, and follow-up data were extracted from the articles and recorded on an Excel database.

Results

General results

Running the above search strings in the selected databases and after duplication removal, 383 articles were identified. After initial check, full-text retrieval, and manual cross-checking of the references, 12 articles met the full criteria for analysis, with no further articles added from the second string.

Articles included were published between 1998 and 2023, 8 being case reports and 4 case series. The results of the included studies are summarized in Table 2.

Table 1
Bias of the included studies.

Title	First Author et al.	Selection 0 1	Ascertainment 0 1	Causality 0 1	Reporting 0 1	Bias 0 1 2 3 4
Hyperostotic En Plaque Meningioma Mimicking Fibrous Dysplasia of the Temporal Bone	Mingo et al.	0	1	1	1	4
Serous Otitis Media Revealing Temporal En Plaque Meningioma	Ayache et al.	1	1	1	1	4
Meningiomas presenting in the temporal bone: the pathways of spread from an intracranial site of origin	Chang et al.	1	1	1	1	4
Extended middle Fossa Surgery for Meningiomas within or at the Internal Auditory Canal	Breuer et al.	1	1	1	1	4
Isolated synchronous meningioma of the external ear canal and the temporal lobe	Bruninx et al.	0	1	0	1	3
Differentiating Hyperostotic Temporal Bone Menigioma En Plaque and Fibrous Dyplapsia on Computed Tomographic Imaging	Shapiro et al.	0	1	0	1	3
Increased Intracranial Pressure Due to Transverse Sinus Compression by a Meningioma En Plaque.	Aidan Healy et al.	0	1	0	1	3
Teaching NeuroImage: Syringomyelia Secondary to Posterior Fossa en Plaque Meningioma	Meng Wang et al.	0	1	0	1	3
Temporal Bone Meningiomas	Fotios D. Vrionis et al.	1	1	1	1	4
A case of temporal bone meningioma presenting as a serous otitis media	Simon Nicolay et al.	0	1	0	1	3
Otitis Media with Effusion Revealing Underlying Meningioma	Diane Evrard et al.	0	1	0	1	3
A rare infectious presentation of a temporal bone meningioma	Abid Qureshi et al.	0	1	0	1	3

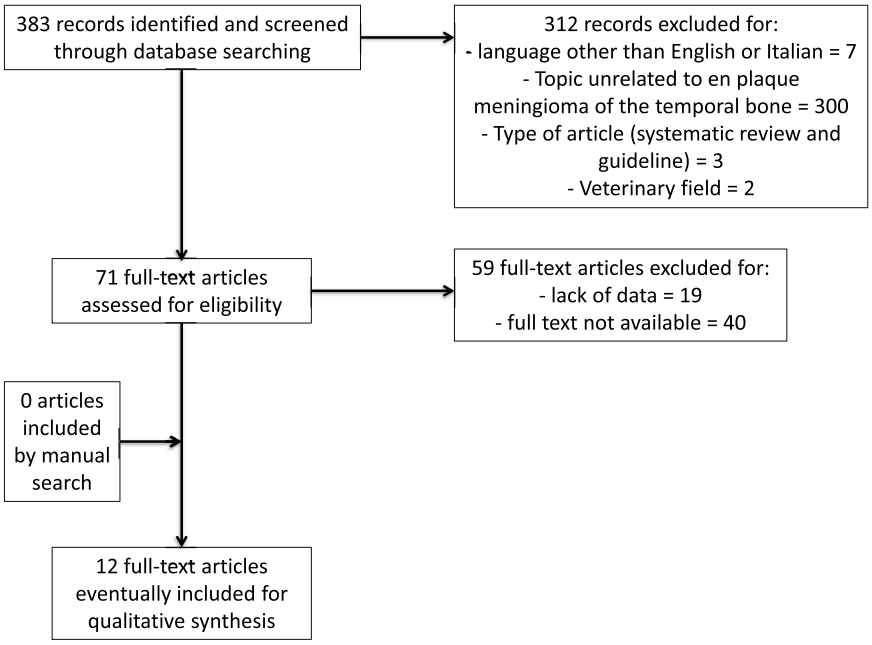

Fig. 1. Study selection process.

Patient population

The total number of patients included in the qualitative synthesis was 25, with the largest study population consisting of 10 patients [5]. The mean age of affected patients was 52 years (range: 31–71 years). Age was not reported in one article (the patient was described as a "middle-aged woman") [8]. The population consisted of 3 males (3/25, 12 %) and 22 females (22/25, 88 %), with a male-to-female ratio of 1:7 (Table 3)

The most affected side was left (12/25, 48 %). In 20 patients (20/25, 80 %), the time elapsed between the onset of symptoms and the diagnosis of MEP was specified and the average duration was 36.5 months (range: 2–120).

Clinical presentation

A summary of possible signs and symptoms of TB-MEP is reported in Fig. 2.

Auditory signs and symptoms

TB-MEP manifests predominantly with auditory signs and symptoms, mostly hearing loss (20/25, 80 %), often accompanied by effusive otitis media (13/25, 52 %), aural fullness (8/25, 32 %), and tinnitus (8/25, 32 %). In some cases, vertigo/dizziness (6/25, 24 %), otorrhea (4/25, 16 %), and otalgia (3/25, 12 %) were reported. The tympanic membrane status was intact in 72 % of cases (18/25), perforated in 4 % (1/25), and not reported in 24 % (6/25).

Neurological signs and symptoms

Occasionally, symptoms related to cranial nerve involvement, such as headache (7/25, 28 %), visual loss (2/25, 8 %), trigeminal neuralgia (1/25, 4 %), facial palsy (1/25, 4 %), dysphonia (1/25, 4 %), or weakness of the shoulder (1/25, 4 %), were observed.

Intracranial involvement

MEP occasionally manifested with intracranial involvement, including meningitis, cerebritis, brain abscess and septic thrombosis (2/25, 8 %). Less frequently reported complications included unsteady walking (1/25, 4 %), papilledema (1/25, 4 %) and nausea (1/25, 4 %).

Imaging

In most cases (15/25, 60 %), there was radiological involvement of the temporal bone only, while in the remaining cases (10/25, 40 %), there was involvement of the temporal bone along with lesser involvement of other sites, such as the parietal bone, occipital bone, sphenoid bone, posterior cranial fossa, or temporal lobe.

Most patients underwent CT and MRI (20/25, 80 %), 3 (3/25, 12 %) patients underwent only CT, while 2 (2/25, 8 %) underwent only MRI. Additionally, in one patient, the suspicion of a temporal bone pathology was raised following scintigraphy performed for another reason. The diagnosis of MEP was established based on radiological images in 13 out of 25 patients (13/25, 52 %), while in 12 cases (12/25, 48 %), additional histological examination was required to confirm the diagnosis.

CT findings

The main CT findings were as follows: hyperostosis (19/25, 76 %), a hairy appearance of bone margins (4/25, 16 %), effusion of the mastoid and middle ear (12/25, 48 %). Fig. 3 shows axial and coronal CT scan views of a left-sided TB-MEP.

MRI findings

Regarding MRI findings, 7 patients (7/25, 28 %) showed dural enhancement, 5 (5/25, 20 %) had temporal hyperostosis, while in 7 cases (7/25, 28 %), an enhancing extra-axial mass was observed, of which 1 case had a synchronous meningioma involving the left orbital roof; in 2 cases (2/25, 8 %), compression of the surrounding neurovascular structures was evident and in 2 patients (2/25, 8 %), the temporal lobe was involved.

Audiometric evaluation

Pre-operative hearing status

In 17 patients (17/25, 68 %), pre-operative audiometric assessments were documented. Of these, 11 patients (11/17, 65 %) exhibited conductive hearing loss with an average threshold of 44 dB (range: 30–90 dB). Notably, 2 patients specifically presented with only low-frequency hearing loss. In 4 patients (4/17, 23 %), a mixed hearing loss was identified, while 2 patients (2/17, 12 %) had normal hearing. Tympanometry data were available for 5 cases (5/17, 30 %) and

Table 2
Summary of the included studies.

	Author	Number of patients included	Patient's number	Sex	Side	Age (years)	Clinical presentation	Diagnosis	Treatments before diagnosis	Time between symptoms onset and diagnosis (months)	Management	FU period (months)	Additional notes
1	*Chang* et al. *1998*	1	1	M	L	45	SOM, HL	CT, MRI, Biopsy	VT (twice), Exploratory tympanotomy	36	Radiotherapy	12 (AWD)	Previous wrong diagnosis of hemangiopericytoma.
2	*Vrionis* et al. *1999*	5	1	M	R	51	HL, tinnitus, vertigo, headache	CT, MRI, Biopsy	Mastoidectomy	2	Surgery (combined MCF craniotomy and mastoidectomy)	12 (NED)	–
			2	F	n/a	37	HL	CT, MRI, Biopsy	Biopsy	96	Surgery (MCF craniotomy)	54 (NED)	–
			3	F	n/a	61	HL, tinnitus, otorrhea	CT, MRI, Biopsy	Biopsy	12	Surgery (MCF craniotomy)	79 (NED)	–
			4	F	L	31	AF with mass behind the TM, HL, tinnitus	CT, MRI, Biopsy	Biopsy	72	Surgery (combined MCF craniotomy and lateral skull base approach)	138 (AWD)	Tumor recurrence in the sphenoid sinus, left optic canal and infratemporal fossa
			5	F	n/a	61	HL with mass behind the TM	CT, MRI, Biopsy	Biopsy	6	Surgery (infralabyrintine/ suboccipital approach)	113 (AWD)	Tumor recurrence in the jugular foramen
3	*Breuer* et al. *2000*		1	F	R	70	HL, tinnitus, vertigo, headache	CT, MRI	n/a	n/a	Surgery (partial resection through extended middle cranial fossa approach)	36 (AWD)	Residual tumor size remained constant during the FU
4	*Ayache* et al. *2006*	10	1	F	L	53	SOM, AF, HL	CT, MRI	Antibiotics, Steroids, VT	48	Clinical + Imaging FU	n/a	
			2	F	R	54	SOM, AF, HL	CT, MRI	Antibiotics, Steroids, VT	72	Clinical + Imaging FU	n/a	
			3	F	R	67	SOM, tinnitus, vertigo, otalgia	CT, Biopsy	Tympanoplasty	24	Clinical + Imaging FU	n/a	
			4	F	L	55	SOM, AF, HL, headache	CT, MRI	VT	24	Clinical + Imaging FU	48 (AWD)	MRI was performed every year, with the latest showing a slight progression of the meningioma.
			5	F	R	70	SOM, HL, otorrhea	CT, MRI	Mastoidectomy, Myringoplasty, VT	96	Clinical + Imaging FU	60 (AWD)	On the last MRI, a slight progression of the right en plaque meningioma occupying the middle fossa floor and extending to the right cavernous sinus was observed (with the patient still symptom-free).
			6	F	L	49	SOM, HL, vertigo, trigeminal neuralgia, mild facial palsy, headache	CT, MRI	Antibiotics, Steroids, VT	24	Surgery (subtotal resection through combined petrosal approach)	n/a	Surgery was decided because of a disabling trigeminal neuralgia. The initial outcome was encouraging (disappearance of trigeminal neuralgia without facial or hearing complications).
			7	F	L	53	SOM, HL, otalgia	CT, MRI	Antibiotics, Steroids, VT, Mastoidectomy	24	Clinical + Imaging FU	n/a	–
			8	F	L	60	SOM, AF, tinnitus	Scintigraphy (breast	n/a	120	Clinical + Imaging FU	36 (AWD)	–

(*continued on next page*)

Table 2 (*continued*)

	Author	Number of patients included	Patient's number	Sex	Side	Age (years)	Clinical presentation	Diagnosis	Treatments before diagnosis	Time between symptoms onset and diagnosis (months)	Management	FU period (months)	Additional notes
								carcinoma), CT, MRI, Biopsy					
			9	F	L	71	SOM, AF, visual loss	MRI	VT	n/a	Radiotherapy	n/a	The patient received RT because of the progression of visual loss. The postoperative course was marked by recurrent episodes of discharge without improvement of the aural fullness.
			10	F	R	58	SOM, HL	CT, MRI, Biopsy	Mastoidectomy, Biopsy	12	Clinical + Imaging FU	n/a	MRI also revealed a second meningioma independent from the right en plaque temporal meningioma: It was a symptom-free meningioma of the left orbital roof.
5	*Bruninx* et al. *2013*		1	M	R	54	HL, vertigo	CT, MRI	n/a	n/a	Surgery (canal wall down modified radical mastoidectomy)	3 (NED)	–
6	*Nicolay* et al. *2014*	1	1	F	L	middle-aged	SOM, HL	CT, MRI	VT, Decongestants	n/a	Surgery (combined middle ear and MCF approach)	n/a	–
7	*Qureshi* et al. *2014*	1	1	F	L	35	HL, otalgia, secondary infectious processes (mastoiditis, meningitis, left sigmoid sinus thrombosis, brain abscess)	CT, MRI, Biopsy	Cortical mastoidectomy, IV antibiotics, anticoagulants	9	Clinical + Imaging FU	n/a	The patient was discharged one month after admission but has since been lost to FU.
8	*Mingo* et al. *2015*	1	1	F	L	42	SOM, HL, dizziness, headache, nausea	CT, MRI	n/a	n/a	Surgery	7 (NED)	Previously misdiagnosed as fibrous dysplasia
9	*Shapiro* et al. *2017*	1	1	F	R	40	AF, HL, tinnitus, headache	CT, Biopsy, MRI	Mastoidectomy, Canalplasty	10	n/a	n/a	Previously misdiagnosed as fibrous dysplasia
10	*Evrard* et al. *2018*		1	F	R	44	SOM, AF, HL	CT, MRI	Antibiotics, Steroids, Decongestants, VT	12	Clinical + Imaging FU	n/a	–
11	*Wang* et al. *2022*	1	1	F	L	36	Unsteady walking, hoarseness, weakness of the shoulder	CT, MRI	n/a	24	Surgery (subtotal resection)	n/a	Previously misdiagnosed as fibrous dysplasia
12	*Healy* et al. *2023*	1	1	F	L	42	Tinnitus, headache, left transverse sinus thrombosis with intracranial hypertension	MRI	Drugs for IH	7	Clinical + Imaging FU	n/a	Previously misdiagnosed as Idiopathic intracranial hypertension.

AF: aural fullness; AWD: alive with disease; CT: computed tomography; FU: follow-up; HL: hearing loss; IV: intravenous; MCF: middle cranial fossa; MRI: magnetic resonance imaging; n/a: not available; NED: no evidence of disease; RT: radiotherapy; SOM: serous otitis media; TM: tympanic membrane; VT: ventilation tube.

Table 3
Epidemiology of meningioma en plaque of the temporal bone and clinical signs and symptoms.

Total number of patients	25
Gender	N(%)
Male	3 (12)
Female	22 (88)
Affected side	N(%)
Right	9 (36)
Left	12 (48)
NA	4 (16)
Clinical Signs and Symptoms	(cumulative number of signs and symptoms)
Auditory	62
Neurological	6
Intracranial involvement	5

indicated a type B pattern in all cases.

Post-treatment hearing outcomes

Post-treatment auditory outcomes were reported for only 3 patients (3/25, 12 %): in two cases, a deterioration in hearing was observed (both cases after surgical intervention through the middle cranial fossa), with the pure tone average increasing from 16 dB to 39 dB in one case and the occurrence of conductive hearing loss in a previously functional ear in the other. In the third patient, however, the air-bone gap was successfully closed after canal wall down modified radical mastoidectomy.

Treatment

Prior to the diagnosis of MEP, 14 patients (14/25, 56 %) underwent various treatments, with each patient potentially having received multiple interventions: 6 (6/14, 43 %) patients were administered antibiotics and steroids; a trans-tympanic tube was inserted in 10 cases (10/14, 71 %), while 7 patients (7/14, 50 %) underwent alternative approaches, including exploratory tympanotomy or mastoidectomy. In one case, medications were prescribed to relieve the intracranial pressure-related symptoms. In 11 cases (11/25, 44 %), no pre-diagnostic treatment was specified.

In terms of definitive treatment, 11 patients (11/25, 44 %) underwent surgery, 11 patients (11/25, 44 %) were sent to "wait and scan", and 2 patients (2/25, 8 %) received radiotherapy. In one case (1/25, 4 %), the definitive treatment administered was not elucidated.

The primary surgical approaches employed were the following: middle cranial fossa approach (6/11, 55 %), subtotal temporal resection (2/11, 18 %), infralabyrinthine approach (1/11, 9 %), mastoidectomy (1/11, 9 %), and in one patient (1/11, 9 %), the procedure was not documented.

Follow-up

Follow-up duration was documented in 12 out of 25 cases (12/25, 48 %), with a mean of 50 months (range: 3–138 months). Among these 12 patients, at the last clinical assessment, seven (7/12, 58 %) were alive with disease (AWD): one underwent radiotherapy, two experienced recurrences after surgical resection, one underwent partial surgical resection, and three were managed with clinical and imaging surveillance. The remaining five patients in this group (5/12, 42 %) showed no evidence of disease (NED) at the last follow-up, all following surgical intervention. Imaging follow-up data were available for 13 patients, with five (5/13, 38 %) undergoing MRI exclusively, one (1/13, 8 %) undergoing CT exclusively, and seven (7/13, 54 %) undergoing both CT and MRI examinations.

Discussion

TB-MEP holds special significance for otolaryngologists due to its tendency to manifest with auditory symptoms, for which the patient seeks for medical advice from an ENT specialist.

Clinical presentation and symptoms

Otological complains are mostly those of serous otitis media, which is commonly co-existent to the tumor, as a consequence of Eustachian tube obstruction resulting from tumor extension into the middle ear cleft [2,9,10,11]. Other frequently reported clinical manifestations include headache, vertigo or dizziness, otorrhea, and otalgia [1,4,5,8,12–14,17,18]. Hearing loss may be conductive, sensorineural, or mixed, depending on the extent of tumor involvement in the middle ear, mastoid cavities, or internal auditory canal (IAC) [4,5,12–14,18]. Rarely, cranial nerve involvement with symptoms such as visual loss, trigeminal neuralgia, facial palsy, dysphonia, and weakness of the shoulder could occur. Wang et al. [15] presented a case of a 36-year-old male exhibiting a two-year history of unsteady walking, hoarse voice, and left shoulder weakness attributed to an en plaque meningioma involving the left temporal and occipital bones, along with cervical syringomyelia. Healy et al. [19] documented a patient manifesting intracranial hypertension consequent to left transverse sinus occlusion secondary to external compression by an underlying en plaque meningioma. Ayache et al. [5] reported the case of a 71-year-old woman experiencing left-sided visual impairment and serous otitis media in association with an en plaque meningioma affecting the posterior aspect of the petrous bone, clivus, and greater wing of the sphenoid. Additionally, these authors described an additional case of a 49-year-old woman with a two-year history of left-sided serous otitis media, concurrent with trigeminal neuralgia and mild facial palsy, attributed to a TB-MEP extending into the middle ear and cavernous sinus. Cranial nerve involvement is less frequent in TB-MEP compared to other meningiomas. Globular meningiomas often present a higher incidence of cranial nerve deficits due to their expansive growth, which exerts pressure on adjacent structures. In contrast, cranial nerve involvement in en plaque meningiomas, especially in the temporal bone, tends to be insidious due to slow, infiltrative tumor growth [5], leading to gradually developing symptoms that may go unrecognized. While TB-MEP primarily affects the acoustic and facial nerves, involvement of other cranial nerves (II, III, IV, or VI) may indicate a MEP involving the sphenoid bone, supported by symptoms such as proptosis, decreased visual acuity, retrobulbar pressure, visual field defects, headaches, or orbital pain [20].

As reported by Qureshi et al. [16], MEP can also manifest as an intracranial complication, for example with bacterial meningitis, brain abscess and mastoiditis with secondary septic thrombosis. These clinical symptoms are due to direct extension of the infection or hematogenous spread from the breakdown of the blood-brain barrier by the meningioma.

Diagnostic challenges in work up and delays

The variability in the clinical presentation can significantly complicate and delay diagnosis. Therefore, raising awareness about specific “red flags” may be significant for clinicians. The presence of persistent hearing loss that does not respond to conventional therapy, accompanied by otorrhea, or unilateral serous otitis media in adults, may warrant further investigation for TB-MEP. Most patients in the reviewed articles received various medical treatments or underwent non-definitive surgical procedures, such as ventilation tube insertion, mastoidectomy, and tympanoplasty, before the diagnosis of TB-MEP [1,4,5,8,16–18]. This correlates with an average diagnostic delay of 36.5 months, ranging from 2 to 120 months, consistent with literature findings [5,15].

First line investigation in case of persistent, treatment-resistant serous otitis media or auditory symptoms, is TB-CT without contrast. This

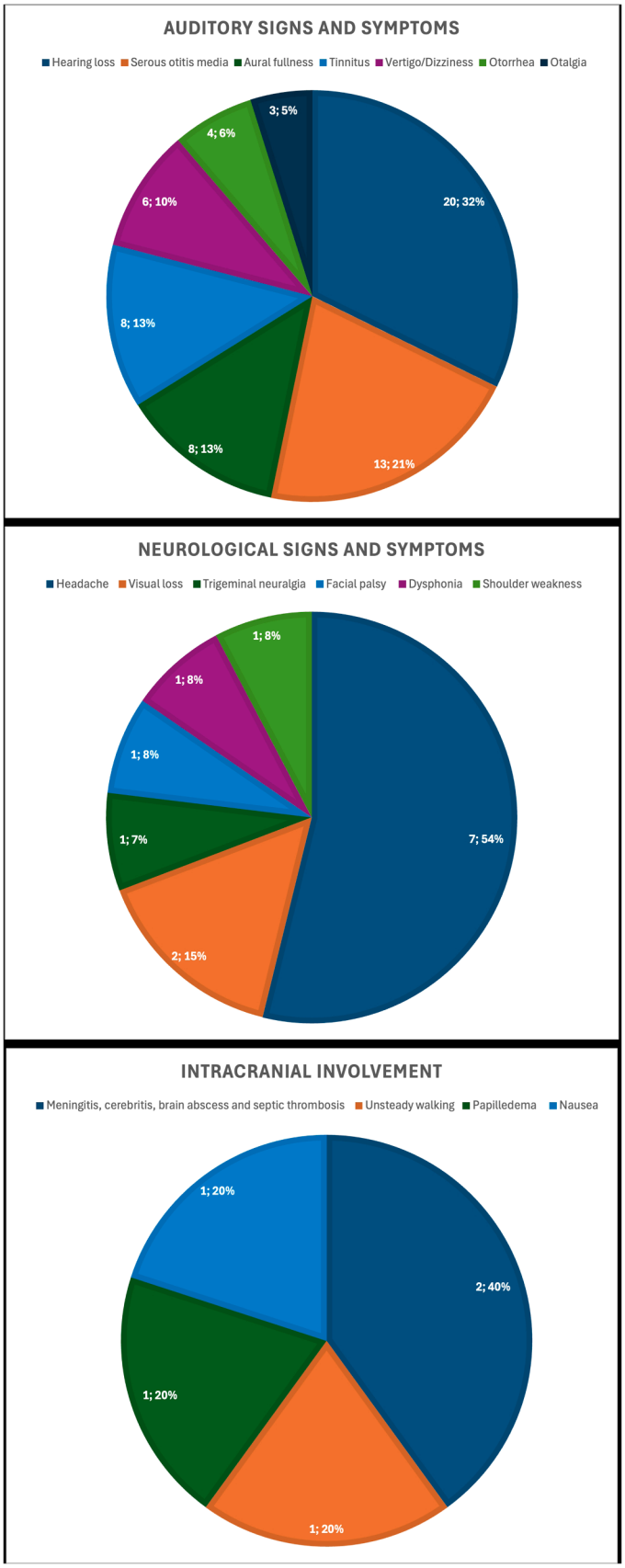

Fig. 2. Distribution of clinical manifestations of meningioma en plaque of the temporal bone.

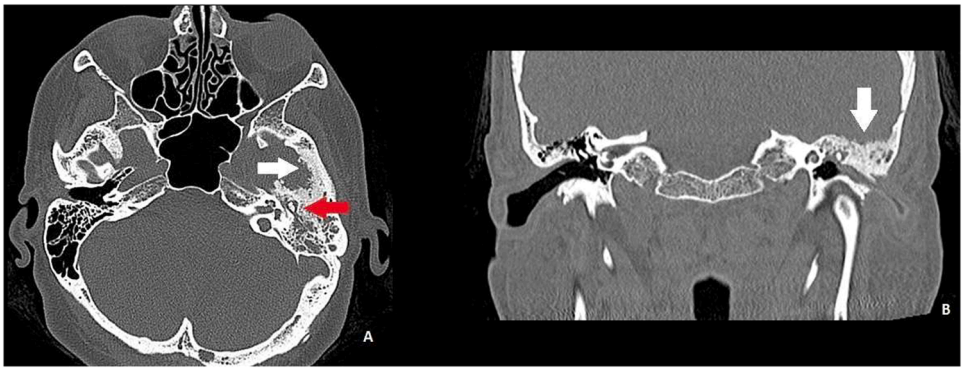

Fig. 3. Computed tomography imaging of a meningioma en plaque of the left temporal bone in a 52-year-old woman. A) Axial view revealing hyperostosis of the left temporal bone (white arrow) and left middle ear effusion (red arrow). B) Coronal view illustrating saw-like and spiculated outer and inner surfaces of the temporal bone (white arrow).

should be performed possibly before the insertion of ventilation tubes. This approach may also aid in differentiating other conditions causing serous otitis media, such as nasopharyngeal tumors [5]. Hyperostotic reactions in the affected bone, preserved trabecular architecture and a hairy, irregular aspect of the margins of the affected bone are indirect radiological features associated with changes in bone structure strongly suggesting the presence of a TB-MEP. Soft tissue opacification within the middle ear cavity is often evident, typically encasing but not destroying the ossicles [5,8,17,18].

TB-MEP could be misunderstood for primary osseous pathological conditions such as fibrous dysplasia, osteoma, osteoblastic metastases to the skull base and metabolic diseases of the bone (for example Paget's disease). Fibrous dysplasia is the condition most misdiagnosed with MEP, as evidenced by three cases included in this review [14,15,18]. The preserved bony architecture can be used to distinguish temporal bone meningioma from the typical ground-glass changes seen in fibrous dysplasia [5,8,14,18].

Although contrast-enhanced CT has proven to be nearly as sensitive as gadolinium-enhanced MRI in the detection of meningiomas, it is important to recognize that CT may have limitations in detecting certain en plaque tumors. Furthermore, its sensitivity can be diminished in regions such as the posterior fossa, where beam hardening artifacts can occur due to the skull base bone structure [4]. In such cases, MRI is preferred to confirm the diagnosis and reduce the likelihood of misdiagnoses.

If CT suggests TB-MEP, MRI is recommended as complementary radiological investigation. This approach helps avoid unnecessary surgical procedures that might otherwise be required to obtain a biopsy for a definitive diagnosis [5]. Gadolinium-enhanced MRI is indeed necessary to accurately detect the meningioma and delineate its extent. T2-weighted images demonstrate hyperintense fluid accumulation in the middle ear, as well as global thickening and hypointense sclerosis of the walls of the middle ear. Gadolinium-enhanced T1 sequences reveal enhancement of the thickened dura, indicating the meningioma's extension into the middle ear and mastoid spaces [3,8,17,18]. Additionally, MRI is preferable in certain patient situations, such as those with iodine allergies or pregnancy, where the use of contrast-enhanced CT may pose risks. In these cases, MRI provides a safe and effective alternative for diagnosis.

The diagnostic assessment of a TB-MEP should also encompass an audiometric evaluation, especially when the tumor affects the middle ear and mastoid cavities, and the ossicular chain. Only a limited number of articles included postoperative audiometric follow-up. Bruninx et al. [13] described a case of a patient who experienced improved hearing after surgery, while Breuer et al. [12] reported a case of a patient with a postoperative decline in hearing function. In this latter case, the tumor was localized in the mid-petrosal region above the IAC with extension into the IAC, and an extended middle cranial fossa approach was performed. These contrasting hearing outcomes highlight that surgical results largely depend on tumor location, the extent of invasion (such as involvement of the ossicular chain and middle ear), and the chosen surgical approach.

Table 4 summarizes the most important diagnostic criteria and management strategies for TB-MEP, providing clinicians with a practical framework for early recognition and appropriate treatment selection.

Management considerations

The most appropriate management approach for a TB-MEP remains controversial and should be individualized based on several factors. Currently, available strategies encompass surgical intervention, radiotherapy, and a watchful waiting regimen with periodic imaging. [3] Surgical intervention may be optimal for younger patients with localized tumors and good overall health, while radiotherapy might be considered for patients with significant comorbidities or high surgical risks. Watchful waiting might be suitable for slow-growing tumors in older patients or those with limited symptoms.

Although radical surgery may be considered the primary treatment approach [1], it is technically challenging due to the tumor's infiltrative nature and its proximity to neural and vascular structures. This approach carries risks of severe postoperative complications related to neural or vascular injuries, as well as an increased likelihood of cerebrospinal fluid (CSF) leaks following complex dural closure of large dura mater defects [21,22].

To optimize tumor control, both the tumor and affected dura should be resected close to the point of attachment, as suggested by Yamada et al. [23].

Table 4
Key Criteria for Diagnosis and Management of meningioma en plaque of the temporal bone.

Criteria	Details
Clinical Presentation	- Persistent, treatment-resistant auditory symptoms (e.g. hearing loss, tinnitus), especially with serous otitis media - Evaluation of cranial nerve involvement (e.g., facial palsy, trigeminal neuralgia)
Imaging Findings	- CT: Look for hyperostotic changes, irregular bony margins, and soft tissue opacification - MRI: Use Gadolinium-enhanced T1 sequences for dura thickening; T2 sequences for fluid accumulation, thickening and hypointense sclerosis of the walls of the middle ear
Management strategies	- Surgery: Recommended for younger patients and those with localized disease - Radiotherapy: Indicated in cases of incomplete resection or involvement of critical structure - Observation: Consider for asymptomatic patients or elderly individuals

Conversely, residual tumors may increase the risk of disease progression. Therefore, the decision-making process for selecting between radical and conservative surgery should consider tumor extension and invasion patterns, involvement of critical neurovascular structures, preoperative hearing status, patient age and comorbidities, growth rate on serial imaging, and the presence and severity of symptoms.

The selection of surgical approach depends on the tumor's extension, hearing preservation goals, and the patient's condition. In most cases, treatment involved a combined middle cranial fossa craniotomy/mastoidectomy approach of varying extension. This approach is believed to provide optimal aggressive tumor removal, thereby minimizing the risk of recurrence. It allows a superior and lateral view of the temporal bone, enabling the removal of all hyperostotic bone and affected dura. Moreover, it assists in the repair of dural defects through the utilization of a vascularized temporalis muscle graft to prevent CSF leaks and can be adapted to include the labyrinth based on the need for hearing preservation [1] Bruninx et al. [13] documented a case wherein a patient with an en plaque meningioma of the external ear canal underwent a canal wall-down modified radical mastoidectomy and experienced a significant improvement in hearing and vestibular symptoms, as long as no signs of tumor recurrence during follow-up. Additional surgical procedures include the infralabyrinthine/lateral skull base approach, particularly effective when the tumor affects the jugular foramen, and the petrosal or transcochlear approach used for tumors involving the petroclival region [1]. Recent advancements in surgical techniques and technologies offer promising solutions to the challenges of treating TB-MEP. For instance, image-guided surgery enhances precision in tumor localization and resection, minimizing damage to adjacent neural and vascular tissues. As reported by Danilo Silva et al. [24], imaging guidance improves the surgeon's ability to accurately identify the internal auditory canal in cases of meningiomas located in the petrous bone, which is particularly advantageous when the cranial nerve complex VII/VIII is encased and obscured by the tumor. Furthermore, imaging guidance facilitates the accurate localization of the transverse-sigmoid sinus complex, contributing to safer surgical approaches. Moreover, endoscopic techniques are increasingly integrated with microsurgery for treating lateral skull base tumors. These minimally invasive approaches not only reduce recovery times and limit surgical trauma but also enhance tumor removal. Endoscopy provides access to hard-to-reach areas within the temporal bone, offering a clearer view of the tumor and surrounding anatomy [25].

In the case of important cranial nerve involvement (i.e., facial palsy or trigeminal neuralgia), selected otoneurosurgical decompression or radiotherapy can be proposed. The cases described by Ayache et al. [5] involved a patient who underwent a subtotal resection of the meningioma through a combined petrosal approach for disabling trigeminal neuralgia and another who received radiotherapy due to progressing visual loss. Only in the first case, the patient's outcome is reported: trigeminal neuralgia disappeared and there were no facial or hearing complications.

There is no definitive consensus regarding the use and impact of radiotherapy both as a standalone treatment and as an adjuvant therapy for these tumors. Common indications for radiation therapy in cases of MEP include incomplete resection associated with residual tumor growth, tumor recurrence, lesions that cannot be surgically removed due to the patient's performance status, or involvement of critical structures like the cavernous sinus or internal carotid artery [3]. In this review, only one patient received isolated radiotherapy due to a progressive visual loss [5].

Different studies [21,26,27] demonstrate that doses of 10 Gy or lower can prevent damage to surrounding neurovascular structures. Although these studies focus on en plaque meningiomas in different locations, we conclude that such dosing may also be safely applied to TB-MEP due to their similar histopathology. Morita et al. [28] assessed the neurological risks and clinical benefits of gamma knife radiosurgery for skull base meningiomas, noting that the risk of trigeminal neuropathy increases with doses over 19 Gy, while the optic apparatus seemed to tolerate doses above 10 Gy. Given the cranial nerve risks associated with open surgery for similar tumors, they concluded that gamma knife radiosurgery is a valuable option for appropriately selected cases of recurrent, residual, or newly diagnosed skull base meningiomas. In cases of asymptomatic MEP or if the potential morbidity is too high, conservative management with a "wait and scan" approach is justified [5,17].

Follow-up and outcomes

The follow-up duration across studies demonstrates significant heterogeneity. While studies with follow-up periods exceeding 48 months consistently report tumor recurrence or progression [1,5], no consensus exists on optimal surveillance intervals. Inspired by other authors [29], we suggest a structured approach to enhance long-term follow-up and assess recurrence rates, consisting of clinical evaluation and CT or MRI with gadolinium every 6 months for the first two years post-treatment, followed by annual imaging for at least five years. For conservatively managed patients, annual imaging is recommended, with increased frequency if radiological progression or new symptoms arise. This schedule should be individualized based on factors such as the extent of resection, tumor location, patient age, and residual disease. Larger prospective studies are needed to validate prognostic indicators and identify additional clinical and histopathologic factors influencing outcomes. Implementing standardized follow-up protocols across institutions and systematic documentation of outcomes in prospective databases would improve the assessment of recurrence rates and enhance the quality of future studies.

Future directions and limitations

This review encounters several limitations that warrant further discussion. The limited number of cases reported in the literature and the broad span of publication dates restrict the generalizability of findings. Another limitation arises from the inclusion of studies with a moderate risk of bias, which can further impact the reliability of the conclusions drawn. To address these limitations, future research should focus on multi-center studies to increase patient populations and develop standardized diagnostic criteria and staging systems. Prospective longitudinal studies could track long-term outcomes, evaluate surgical approaches, and investigate prognostic factors. Given the diagnostic and therapeutic challenges highlighted, there is a need for evidence-based clinical guidelines specific to TB-MEP. Such guidelines would offer clinicians a structured approach to diagnosis, surgical decision-making, and post-operative management, ultimately improving patient outcomes and standardizing care for this rare condition.

This review underscores the critical need for further research to assist surgeons in determining optimal management based on current knowledge, considering the individual characteristics of tumors and patients.

Conclusion

Due to the rarity and the tricky signs and symptoms of TB-MEP, maintaining a high index of suspicion is essential to avoid mismanagement of this tumor. Appropriate early imaging and individualized treatment strategies are paramount to enhance patient's outcomes.

In cases of unilateral therapy-resistant auditory symptoms with or without neurological deficits, a CT of the temporal bone should be performed and integrated with gadolinium-enhanced MRI for confirmation of the diagnosis. Current management strategies include surgical intervention, radiotherapy, and a surveillance protocol with periodic imaging; the most appropriate management should be individualized based on several factors.

Funding disclosures

None.

CRediT authorship contribution statement

Arianna Burato: Writing – original draft, Validation, Methodology, Data curation, Conceptualization. **Giuseppe Maruccio:** Writing – original draft, Validation, Methodology, Formal analysis, Data curation, Conceptualization. **Livio Presutti:** Writing – review & editing, Validation, Conceptualization. **Ignacio Javier Fernandez:** Writing – review & editing, Validation, Methodology, Conceptualization. **Gabriele Molteni:** Writing – review & editing, Supervision. **Giulia Molinari:** Writing – original draft, Validation, Methodology, Formal analysis, Data curation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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