Otology & Neurotology
39:320–332 © 2018, Otology & Neurotology, Inc.

Temporal Bone Meningo-Encephalic-Herniation: Etiological Categorization and Surgical Strategy

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Objective: To study the clinical presentation, intraoperative findings and surgical management in meningo-encephalic-herniation (MEH) based on the etiology.
Study Design: A retrospective clinical study and is a follow-up on the previously published report in 2009.
Setting: A quaternary referral otology and skull base center
Patients and Methods: The inclusion criteria were intraoperatively verified MEH in patients with a minimum follow-up of 12 months, which yielded 262 operated ears. The data were extracted regarding demographics, laterality, clinical presentation, past surgeries, contralateral-ear condition, intraoperative findings, complications, recurrences, revision-surgeries, audiometric-data, and follow-up.
Results: The mean age at surgery was 49.7 years with the involvement of right-ear in 53.8% of patients. Lesions were categorized based on the etiology as chronic-otitis-media with/without cholesteatoma-MEH (COM/CHOL-MEH)-47.7%, iatrogenic-MEHs −20.9%; traumatic-MEHs −8% and spontaneous-MEHs −23.3%. At presentation, hearing loss (100 and 98.2%) and otorrhea (65.6 and 49.1%) were predominant in COM/CHOL-MEHs and iatrogenic-MEHs, respectively. On the other hand, meningitis (23.9 and 14.3%) and cerebrospinal fluid-leak (52.4 and 42.8%) were more pronounced in spontaneous and traumatic MEHs, respectively. Surgical approaches included 1) transmastoid, 2) middle-cranial-fossa-approach, 3) combined, and 4) middle-ear-obliteration (MEO) techniques. A total of 52.8% of COM/CHOL-MEHs and 49.1% of iatrogenic-MEHs underwent MEO. Middle-cranial-fossa approach was predominantly used in spontaneous-MEHs (52.5%) and traumatic-MEHs (38.1%). The defect was mostly single (75.2%). Smaller, multiple, bilateral lesions were more common in spontaneous-MEHs with tegmen-tympani involvement (57.4%).
Conclusion: Incorporating etiology into MEHs is a key-step that can be used as a guidance in choosing the right surgery. MEO is a part of armamentarium, and should be used whenever needed, if the objective is performing a definitive surgery. **Key Words:** Cerebrospinal-fluid leak—Iatrogenic-MEH—Meningitis—Meningo-encephalic herniation—Middle-ear-obliteration—Spontaneous-MEH—Surgical repair—traumatic-MEH.

Otol Neurotol **39:**320–332, 2018.

Meningo-encephalic-herniation (MEH) is defined by the presence of brain tissue with its meningeal cover descending into the temporal bone (1–3). Although described in the literature a century ago (4), diagnosis of a temporal-bone-MEH can be quite difficult, given the rarity of the condition with the incidence varying from 1 in 3,000 to 1 in 10,000 (5,6). In addition, even in patients of CSF otorrhea, a herniation does not have to exist. Radiologically investigating through computed tomography (CT) and magnetic resonance imaging (MRI) may be the only way to preoperatively make a diagnosis.

Chronic inflammation, previous surgery, irradiation, trauma, and neoplasms are well-known predisposing factors for MEHs (7). Recently, an increased number of spontaneous patients have been reported (8–14), associated with benign intracranial hypertension or pseudotumor cerebri (15–17). While in chronically draining/operated ears, MEH symptomatology is often mimicked by otological issues, the diagnosis of spontaneous-MEHs relies on a high degree of suspicion in any prolonged case of unilateral clear otorrhea (9). Consequently, delay in diagnosis can lead to neurological complications (2,9–12,18–25) (Figure 1).

Hence, once a MEH is diagnosed, a prompt surgery is mandatory and based on site and size of defect various approaches like trans-mastoid-approach (TMA), middle-cranial-fossa approach (MCFA), combined-approach (transmastoid-middle-cranial-fossa) and middle-ear-obliteration (MEO) with cul-de-sac closure of the external ear canal can be performed. Surgery is individually tailored and presently there is no consensus regarding optimal surgical management.

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The authors disclose no conflicts of interest.
DOI: 10.1097/MAO.0000000000001693

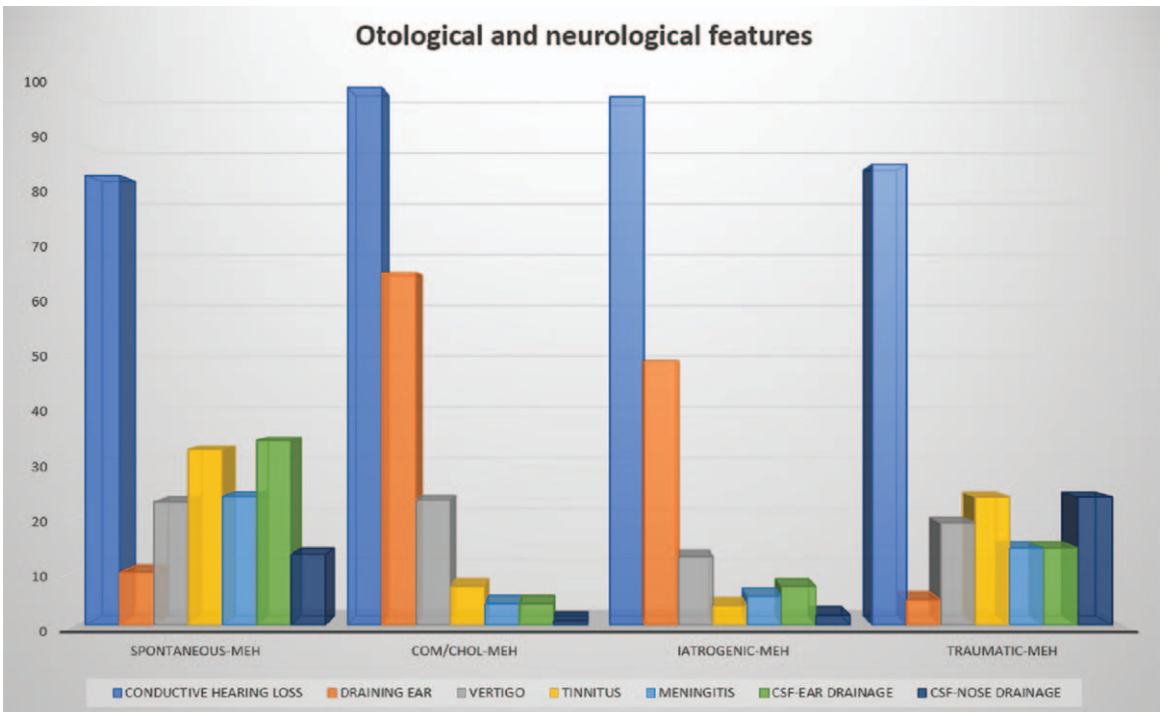

FIG. 1. Distribution of otological and neurological features in meningo-encephalic hernia according to etiology.

To date, numerous reports have highlighted on a single MEH etiology (7,9,10,26–28). In the current follow-up study of our previous report (19), additional consecutive 129 patients were added to the previous 133 patients, which enabled comparison between different etiologic groups with respect to clinical characteristics, herniation properties, and surgical strategy. Furthermore, a substantial number of patients underwent MEO procedure, providing a thorough evaluation of this technique as to be seriously taken into consideration when dealing with MEH pathology.

METHODS

A retrospective chart review of all patients operated at our center from February 1983 to June 2016 for MEHs of the temporal-bone was conducted. The inclusion criteria were intraoperatively verified MEH in patients with a minimum follow-up of 12 months. The exclusion criteria were 1) patients of dura exposure through bony defect with cerebrospinal fluid (CSF) leak with no visible herniation observed intra-operatively 2) patients with a history of temporal-bone neoplasia or head/neck radiation, and 3) patients with incomplete records or those lost to follow-up for a minimum period of 1 year.

This yielded 262 operated ears from 249 patients for analysis. The data were extracted regarding demographics, laterality, clinical presentation, past surgeries, contralateral-ear condition, intraoperative findings, complications, recurrences, revision-surgeries, audiometric-data, and follow-up.

All patients underwent complete preoperative oto-neurologic evaluation. Facial nerve function was graded pre and postoperatively by the House-Brackmann grading system from 1985 onward (29). However, the first two patients reported prior to this period were classified retroactively.

The audiometric-data obtained included pre- and postoperative air-conduction (AC), bone-conduction (BC) thresholds at four frequencies (500, 1000, 2000, and 4000 Hz), pure-tone average (PTA). A high-resolution computed tomography (HRCT) scan of the temporal-bone and MRI were obtained in all patients.

Many classifications have been reported, based on location of defect and etiology (28,30–33). However, in the present series, MEHs were classified into four categories based on the etiology: chronic otitis media/cholesteatoma-MEHs (COM/CHOL-MEHs), iatrogenic-MEHs, spontaneous-MEHs, and traumatic-MEHs. COM/CHOL-MEHs were assigned for all patients with active infection/cholesteatoma seen intraoperatively, regardless of their previous surgical history. Otherwise, all other postoperative patients without above-mentioned intraoperative findings seen during MEH surgery were categorized as iatrogenic-MEHs. Spontaneous-MEHs were assigned for patients without an obvious underlying reason.

Surgeries adopted in this series included 1) TMA, 2) MCFA, 3) combined-approach, and 4) MEO. The following were the surgical approaches employed in the present series.

Transmastoid-approach: mastoidectomy was performed and MEH was exposed, coagulated, and sectioned at the waist. The repair of the bony-defect was effected ''from below'' by inserting a piece of cartilage/bone through the defect, which may be reinforced with bone-pate and muscle/fascia.

Middle-cranial-fossa approach: a skin-incision was taken from pretragal area, curving at first posteriorly, then anteriorly, extending superiorly for 7 to 8 cm with an incision in the temporalis muscle exposing the temporal-squamous bone. A

4 × 4 cm craniotomy was performed, with its lower edge at the level of the zygomatic process. After careful elevation of the MCF dura, the MEH neck was identified, coagulated and cut. The MEH tissue was left in middle ear/mastoid. A piece of cartilage/calvarian-bone was placed between the bony defect and the dura. The defect was further reinforced with muscle/fascia (19).

Combined-approach: trans-mastoid approach was combined with a mini-craniotomy in the temporal region at the level of tegmen. The MEH was treated as explained in trans-mastoid-approach. Then, a sufficiently large piece of cartilage was inserted extra-durally through the craniotomy opening (52). The bony-defect was further repaired by bone-pate and covered by temporalis fascia from the mastoid cavity.

Middle-ear-obliteration: after blind-sac-closure of the external-auditory-canal, a subtotal petrosectomy was performed, with complete removal of mucosa, skin, tympanic-membrane, malleus, and incus. The MEH was reduced by bipolar coagulation and cut with a microscissor. Once the disease was eradicated, the middle-ear opening of the eustachian tube was plugged with soft-tissue and bone-wax. The cavity was then obliterated with abdominal fat (57).

Operating-notes were analyzed for size, number, location of the defect, repair-technique, materials used, and presence of an active CSF-leak.

The bony-defect size was categorized into three groups: ≤1 cm (small), 1 to 2 cm (medium), and >2 cm (large) and was microscopically estimated in all patients. Multiple defects were considered when the number exceeded two and this was considered to maintain uniformity with most of the previously published reports (2,9–12).

Technique of Dura/Bone Repair

Multilayered autologous tissue was employed for extradural-repair in majority of patients and solid autologous materials were added to separate the two cavities in most of them (Table 1). The exceptions were some very small, single, postero lateral tegmen, and all PCF-defects, wherein a piece of temporalis-fascia, muscle/fat and human-fibrin-glue were sufficient. In about one-fifth patients managed by MEO, the repair was not performed. In some patients, after gradual reduction and excision of a MEHs stalk in the ''waist,'' dura appeared sealed at the tegmen, presumably by bipolar-coagulation, and no further repair was needed. In others, substantial dura and bone-loss gave no possibility for a repair.

Review of the Literature

A review of the literature was done using a PubMed Search using relevant search words. The demographics, presentation, etiology, laterality, surgical approach, MEH-properties, active CSF-leak, hearing, complication, recurrence, and revision rates with follow-up time were tabulated and compared.

The present study was approved by the institutional review board of the hospital for ethical research.

Statistical Analysis

Data analysis was done using SPSS 24 (IBM, New York, NY) statistical package. Chi-square test was used to measure the significance for the nonparametric data. The significance of means was measured between groups using ''Analysis of variance'' (ANOVA). The pre- and postoperative hearings were evaluated using paired *t* test. A *p* value less than 0.05 was considered statistically significant (95% confidence interval).

RESULTS

Spontaneous-MEHs (Table 2) predominantly occurred in the older (57.1 yr), while the traumatic-MEHs occurred in younger age-group (31.5 yr). Of the 262 ears, 128 (51.5%) were males and 134 (48.5%) were females and the mean age was 49.4 years. Out of 13 bilateral patients, 50% were spontaneous-MEHs. Thirty eight (62.3%) in spontaneous-MEHs and 35 (63.6%) in iatrogenic-MEHs occurred in the right ear. While the traumatic-MEHs presented early (22.6 mo), the COM/CHOL-MEHs presented quite late (45.4 mo). Fifty-six percent of COM/CHOL-MEHs had previous ipsilateral surgeries. The contralateral-ear was affected in 26 (20.8%), 15 (27.3%), 8 (13.1%) patients of COM/CHOL-MEHs, iatrogenic-MEHs, and spontaneous-MEHs, respectively.

Clinical Features

The main presentation was conductive-hearing-loss (94.6%) followed by discharging-ear (44.3%) (Table 2). The latter was more pronounced in COM/CHOL-MEHs and iatrogenic-MEHs. Pulsatile-tinnitus was found predominantly in spontaneous-MEHs and traumatic-MEHs ($p = 0.001$).

Meningitis (23.9% and 14.3%, respectively, in spontaneous-MEHs and traumatic-MEHs) and CSF-ear drainage (39.3% and 19%, respectively, in spontaneous-MEHs and traumatic-MEHs) were the common neurological sequelae observed. However, they were uncommon in COM/CHOL-MEHs and iatrogenic-MEHs.

Iatrogenic-MEHs had previously undergone canal-wall-up, canal-wall-down, and radical-mastoidectomies in 23 (41.8%), 15 (27.3%), and 9 (16.4%) ears, respectively. Canaloplasty and subtotal-petrosectomy (STP) were performed in one ear each and 6 ears were pluri-operated (mean: 1.7 surgeries).

Surgical Approaches

Overall, MEO (46.2%) and MCFA (32.8%) were the commonly employed surgeries followed by TMA (32.8%) and combined-approach (1.9%) (Table 1) (Figure 2). Sixty-six (52.8%) of COM/CHOL-MEHs and 27 (49.1%) of iatrogenic-MEHs underwent MEO. MCFA was predominantly employed in 32 (52.5%) of spontaneous-MEHs and 8 (38.1%) of traumatic-MEHs. TMA were largely utilized for COM/CHOL-MEH in 56 (44.8%) and iatrogenic-MEH in 18 (32.7%) ears. All patients with ventilating tubes underwent myringoplasty simultaneously, with the exceptional nine MEO patients. All perforations closed successfully.

Defect Properties

Size: Overall, the small (≤1 cm), medium (1–2 cm) and large (>2 cm) defects were found in 79 (30.2%), 99 (37.8%), and 84 (32.1%) patients, respectively (Table 3).

Number: Single-defect (75.2%) was most common across all MEHs, whereas multiple was predominantly found in spontaneous-MEHs (27.9%).

Location: Tegmen-mastoid was involved in 59.2%, tegmen-antri in 46.6%, tegmen-tympani in 29.8% of

TABLE 1. *Surgical treatment with outcomes according to etiology*

Surgery Approach	Etiology n (%) Spontaneous-MEH 61 (23.3)	COM/CHOL-MEH 125 (47.7)	Iatrogenic-MEH 55 (20.9)	Traumatic-MEH 21 (8.0)	Total n (%)	*p* Value
TMA[a]	6 (9.8)	56 (44.8)	18 (32.7)	6 (28.6)	86 (32.8)	0.001
MEO	22 (36.1)	66 (52.8)[b]	27 (49.1)	6 (28.6)	121 (46.2)	
MCFA	32 (52.5)	2 (1.6)	8 (14.5)	8 (38.1)	50 (19.1)	
Combined[a]	1 (1.6)	1 (0.8)	2 (3.6)	1 (4.8)	5 (1.9)	
Repair technique and materials						
TF+C+BP+HFG	5 (8.2)	8 (6.4)	6 (10.9)	0 (0)	19 (7.3)	0.010
TF+B±M ± HFG	17 (27.9)	15 (12.0)	13 (23.6)	9 (42.9)	54 (20.6)	
TF+C ± M ± HFG	21 (34.4)	61 (48.8)	21 (38.2)	9 (42.9)	112 (42.7)	
TF+M+HFG	11 (18.0)	13 (10.4)	4 (7.3)	1 (4.8)	29 (11.1)	
TF+F+HFG	2 (3.3)	0 (0)	1 (1.8)	0 (0)	3 (1.1)	
none	5 (8.2)	28 (22.4)	10 (18.2)	2 (9.5)	45 (17.2)	
Active CSF leak	28 (45.9)	50 (40.0)	23 (41.8)	9 (42.8)	110 (41.9)	0.897
Recurrence	0 (0)	3 (2.4)	0 (0)	1 (4.8)	4 (1.5)	—
Revision surgery	1 (1.6)[c]	4 (3.2)[d]	0 (0)	2 (9.5)[e]	7 (2.7)	0.128

[a]Transmastoid approach.
[b]Out of 66 patients, 60 underwent subtotal petrosectomy and 6 underwent transotic or transcochlear approach due to preoperative deafness.
[c]Revision due to CSF leak- without MEH.
[d]One out of 4 had revision for CSF leak without recurrent MEH.
[e]Revision d\t CSF; n: number leak in one, and post-traumatic MEH entrapped cholesteatoma and MEH in external ear canal in other patient (first operation was MCF approach).

B indicates bone; BP, bone pate; C, cartilage; CHOL, cholesteatoma; COM, chronic otitis media; CSF, cerebrospinal-fluid; F, fat; HFG, human fibrin glue; M, muscle; MCFA, middle-cranial fossa approach; MEH, meningo-encephalic-herniation; MEO, middle-ear obliteration; TF, fascia temporalis; TMA, transmastoid approach.

patients. While the tegmen-mastoid was least involved in spontaneous-MEHs (13.1%), it involved others in >36% of patients ($p = 0.004$). Tegmen-tympani was involved in 18% of spontaneous-MEHs, whereas it was involved in 5 (4%), 3 (5.5%), and 2 (9.5%) of COM/CHOL-MEHs, iatrogenic-MEHs, and traumatic-MEHs, respectively ($p = 0.009$). Posterior-fossa-dura defect was observed in seven patients.

Repair of Defect

After the herniated tissue was reduced using bipolar coagulation and excised with a microscissor, the bony defect was subsequently repaired. A combination of temporalis-fascia, cartilage with/without a free-flap temporalis-muscle, human-fibrin-glue, bone, and bone-pate were used. As a solid component, cartilage was used in 112 (42.7%) and bone in 54 (20.6%) of patients. Wherever the cartilage was inadequate, bone-pate was used and was reinforced further by temporalis-fascia, temporalis-muscle, and Human-fibrin-glue as in 19 (7.3%) patients. Small-tegmen and all posterior-fossa-dura defects were usually closed with either temporalis-fascia with/without muscle and glue.

Hearing Outcomes

Preoperatively, a total of 34 (12.9%) patients had profound-hearing-loss. In the remaining 228 (87.1%) patients of measurable hearing, overall mean pre-and-postoperative data are demonstrated in Table 4. The AC-thresholds were similar between spontaneous-MEHs and traumatic-MEHs and COM/CHOL-MEHs and iatrogenic-MEHs. The BC-thresholds in all four categories ranged between 25.6 and 28.9 dB (Table 4).

Postoperatively, five patients developed profound-hearing loss. Their mean preoperative PTA-AC and PTA-BC were 77 and 53 dB, respectively. In one patient of spontaneous-MEH this occurred after a STP. In four patients with petrous-bone-cholesteatoma (three COM/CHOL-MEHs, one iatrogenic), STP was performed in three patients, of which one patient required labyrinthectomy; transotic-approach was done in the other patient. In the remaining 85 patients who had preoperatively measurable-hearing who underwent MEO, AC and BC dropped by 6.1 and 2.7 dB, respectively.

In the COM/CHOL-MEHs and iatrogenic-MEHs, there was an improvement in the postoperative ABG by 6.5 and 6.4 dB, respectively, which was statistically significant. However, in spontaneous-MEHs and traumatic-MEHs, improvement in the postoperative ABG was 2.6 and 3.1 dB, respectively.

Follow-up Evaluation

The mean duration of follow-up was 39.9 month and patients were evaluated clinically every 6 months in the first year and subsequently once-a-year upto 5 years with both HRCT and MRI. We recommend evaluation for idiopathic intracranial hypertension, especially in spontaneous MEH after repair.

Complication, Recurrence, and Revision Surgery

Postoperative complications occurred in 7 (2.7%) patients. One patient developed immediate profound-

TABLE 2. *Demographics and presentation according to etiology*

Variables	Etiology n (%)					
	Spontaneous-MEH 61 (23.3)	COM/CHOL-MEH 125 (47.7)	Iatrogenic-MEH 55 (20.9)	Traumatic-MEH 21 (8.0)	Total (n = 262) n (%)	*p* Value
Demographics						
Mean age (range)	57.1 (13–76)	49.1 (9–82)	49.9 (11–78)	31.5 (11–66)	49.7 (9–82)	0.001
Female	25 (41.0)	57 (45.6)	34 (61.8)	11 (52.4)	127 (48.5)	0.120
Male	36 (59.0)	68 (54.4)	21 (38.2)	10 (47.6)	135 (51.5)	
Right side	38 (62.3)	59 (47.2)	35 (63.6)	9 (42.9)	141 (53.8)	0.068
Left side	23 (37.7)	66 (52.8)	20 (36.4)	12 (57.1)	121 (46.2)	
Bilateral lesion[a]	13 (21.3)	10 (8.0)	3 (5.5)	0 (0)	26 (9.9)	0.005
Presentation						
Otologic						
CHL	51 (83.6)	125 (100)	54 (98.2)	18 (85.7)	248 (94.6)	0.001
Profound HL	2 (3.3)	23 (18.4)	7 (12.7)	2 (9.5)	34 (12.9)	0.036
Draining ear\otorrhea	6 (9.8)	82 (65.6)	27 (49.1)	1 (4.7)	116 (44.3)	0.001
Vertigo	14 (22.9)	29 (23.2)	7 (12.7)	4 (19.0)	54 (20.6)	0.417
+ fistula sign[b]	2 (3.3)	21 (16.8)	3 (5.4)	0 (0)	25 (9.5)	0.004
Tinnitus	20 (32.7)	9 (7.2)	2 (3.6)	5 (23.8)	36 (13.7)	0.001
Neurologic						
Meningitis	17 (23.9)	5 (4.0)	3 (5.4)	3 (14.3)	28 (10.7)	0.001
Brain abscess	1 (1.6)	1 (0.8)	2 (3.6)	0 (0)	4 (1.5)	0.498
Seizures	3 (4.9)	1 (0.8)	0 (0)	0 (0)	4 (1.5)	
FN palsy	0 (0)	5 (4.0)	4 (7.2)	2 (9.5)	12 (4.5)	0.141
CSF leakage						
CSF-ear drainage	24 (39.3)	18 (14.4)	8 (14.5)	4 (19.0)	54 (20.6)	0.001
CSF-nose drainage	8 (13.1)	1 (0.8)	1 (1.8)	5 (23.6)	15 (5.7)	0.001
Previous VT[c]	21 (34.4)	3 (2.4)	0 (0)	0 (0)	24 (9.2)	-
Mean symptom duration in months (range)	34.3 (4–135)	45.4 (1–168)	37.0 (1–125)	22.6 (1–62)	39.2 (1–168)	0.007
Hospital stay (d)	4 (1–6)	3 (1–6)	3 (1–6)	4 (1–6)		0.001
Follow-up (mo) (mean, range)	34.8 (12–108)	39.1 (12–180)	47.6 (12–312)	39.4 (12–114)	39.9 (12–312)	0.231
Previous ipsilateral surgery (besides VT)	5 (8.2)[d]	70 (56.0)[e]	55 (100.0)[f]	2 (9.5)[g]	132 (50.4)	0.001
Associated ear disease in the opposite ear	8 (13.1)[h]	26 (20.8)[i]	15 (27.3)[j]	1 (4.7)[k]	50 (19.1)	0.077

[a]Thirteen patients had bilateral MEH. Eleven patients had MEH of the same etiology. Out of two reminder patients, in first one, MEH occurred due to cholesteatoma in one and SMEH in the other ear. In the second patient, MEH occurred due to cholesteatoma in one and after ear operation in the other ear.

[b]In all patients SCC fistula/labyrinth erosion was found intra-operatively.

[c]Ventilating tubes.

[d]Three patients were twice operated by MCF approach; one patient underwent two MCF approaches, and one TM approach without success, the reminder patient had three TM approaches-all without success.

[e]Thirty-nine patients underwent CWU mastoidectomy, 28 underwent CWD mastoidectomy, 2 underwent radical mastoidectomy, and 1 canaloplasty.

[f]Twenty-three patients underwent CWU mastoidectomy, 15 underwent CWD mastoidectomy, 9 underwent radical mastoidectomy, 1 case had canaloplasty, 1 subtotal petrosectomy, and 6 cases were plurioperated.

[g]Eleven had cholesteatoma, three had COM, and one had MEH.

[h]Seven out of eight patients had bilateral MEH (six operated in our institution and one elsewhere before); one patient had COM in the opposite ear.

[i]Twenty-one ears were affected by cholesteatoma, four ears had bilateral MEH due to COM ± CHOL, the reminder patient had SMEH in contralateral ear.

[j]Both had release of parieto-temporal hematoma.

[k]Cholesteatoma in the contralateral ear; n: number

CHOL indicates cholesteatoma; COM, chronic otitis media; CSF, cerebrospinal fluid; CWU, canal wall-up; EEC, external ear canal; HL, hearing loss; MCF, middle-cranial-fossa; MEH, meningo-encephalic herniation; SCC, semi-circular canal; SMEH, spontaneous meningo-encephalic herniation; TM, tympanic membrane; VT, ventilating tube.

hearing-loss after STP for spontaneous-MEHs without any apparent labyrinth involvement as observed intra-operatively. A detailed HRCT and MRI examination failed to prove any inner-ear damage. One patient developed extradural-subtemporal hematoma and other patient meningitis; they were managed conservatively.

Postoperatively, CSF-leak occurred in three patients and was subsequently managed by revision surgery. No

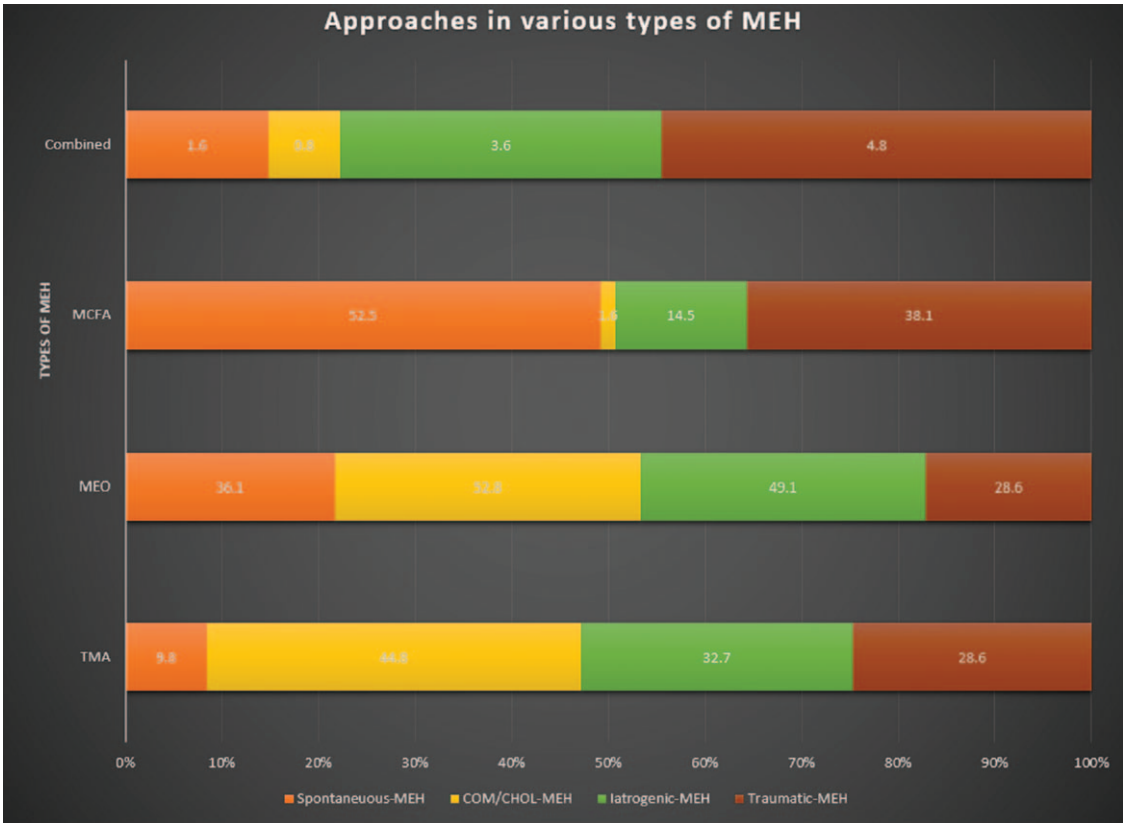

FIG. 2. Surgical approaches followed in meningo-encephalic-herniation.

TABLE 3. *MEH properties according to etiology*

MEH properties	Etiology n (%)					
Size (cm)	Spontaneous-MEH 61 (23.3)	COM/CHOL-MEH 125 (47.7)	Iatrogenic-MEH 55 (20.9)	Traumatic-MEH 21 (8.0)	Total n (%)	*p* Value
≤1 (small)	28 (45.9)	42 (33.6)	11 (20.0)	6 (28.6)	79 (30.2)	0.080
>1 to 2 (medium)	20 (31.6)	37 (29.6)	24 (43.6)	11 (52.4)	99 (37.8)	
>2 (large)	14 (23.0)	46 (36.8)	20 (36.4)	4 (19.0)	84 (32.1)	
Number						
1	36 (59.0)	98 (78.4)	47 (85.5)	16 (76.2)	197 (75.2)	0.017
2	8 (13.1)	13 (10.4)	5 (9.1)	2 (9.5)	28 (10.7)	
Multiple	17 (27.9)	14 (11.2)	3 (5.5)	3 (14.3)	37 (14.1)	
Location						
multiple site	33 (54.1)	38 (30.4)	22 (40.0)	9 (42.9)	102 (38.9)	0.019
TM[a] only	8 (13.1)	48 (38.4)	20 (36.4)	8 (38.1)	84 (32.1)	0.004
TA[b] only	7 (11.5)	29 (23.2)	9 (16.4)	2 (9.5)	47 (17.9)	0.159
TT[c] only	11 (18.0)	5 (4)	3 (5.5)	2 (9.5)	21 (8.0)	0.009
PCF[d]	2 (3.3)	4 (3.2)	1 (1.8)	0 (0)	7 (2.7)	0.813
EEC[e]	0 (0)	1 (0.8)	0 (0)	0 (0)	1 (0.4)	0.777
TM overall	29 (47.5)	80 (64.0)	35 (63.6)	11 (52.4)	155 (59.2)	0.140
TA overall	29 (47.5)	56 (44.8)	28 (50.9)	9 (42.9)	122 (46.6)	0.870
TT overall	35 (57.4)	18 (14.4)	15 (27.3)	10 (47.6)	78 (29.8)	0.001
PCF overall	5 (8.2)	16 (12.8)	4 (7.3)	0 (0)	25 (9.5)	0.463
EEC overall	0 (0)	2 (2.4)	2 (3.6)	2 (3.6)	7 (2.7)	0.127

[a]Tegmen mastoid.
[b]Tegmen antrum.
[c]Tegmen tympani.
[d]Posterior cranial fossa.
[e]External ear canal; n, number.
CHOL indicates cholesteatoma; COM, chronic otitis media; MEH, meningo-encephalic-herniation; PCF, posterior cranial fossa; TA, tegmen mastoid; TM, tympanic membrane; TT, tegmen tympani.

TABLE 4. *Comparison between preoperative and postoperative hearing status between etiology subgroups*

Etiology	Spontaneous-MEH 61 (23.3)	COM /CHOL-MEH 125 (47.7)	Iatrogenic-MEH 55 (20.9)	Traumatic-MEH 21 (8.0)	Total (n = 262)	*p* Value[a]
Preoperative hearing (dB)						
Deafness	2 (3.3)	23 (18.4)	7 (12.7)	2 (9.5)	34 (12.9%)	0.035
PTA-AC	47.6dB ± 18,2	62.1dB ± 18.4	61.2dB ± 20.2	45.3dB ± 22.9	56.7dB ± 20.4	0.001
PTA-BC	26.8dB ± 10,3	28.9dB ± 13.5	27.7dB ± 12.7	25.6dB ± 12.5	27.8dB ± 12.5	0.723
ABG	21.8dB ± 12.3	33.1dB ± 12.7	33.5dB ± 13.7	19.7dB ± 13.9	28.8dB ± 14.4	0.001
Hearing at last follow-up (dB)						
Deafness[b]	3 (4.9)	26 (20.8)	8 (14.5)	2 (9.5)	39 (14.8%)	0.033
PTA AC	46.6dB ± 21.6	57.6dB ± 21.7	56.1dB ± 20.5	42.8dB ± 20.8	53.2dB ± 22.1	0.023
PTA BC	27.9dB ± 11.1	30.2dB ± 13.6	28.4dB ± 11.9	26.3dB ± 13.2	28.8dB ± 12.7	0.568
ABG	19.2dB ± 15.1	26.6dB ± 14.7	27.1dB ± 13.8	16.6dB ± 12.6	24.3dB ± 14.8	0.015
Paired *t* test						
	n (56)	n (102)	n (48)	n (18)	Cases with the hearing preservation have been analyzed for pre-and post-operative ABG	
Pre-operative ABG	21.8	33.1	33.4	25.4		
Post-operative ABG	19.2	26.6	27.1	20.9		
p	0.001	0.019	0.001	0.348		

[a]Test of analysis of variance (ANOVA) for means and χ^2 test for proportions has been used; ABG, Air-bone gap; n, number.

[b]In all present series, five patients had profound hearing loss postoperatively. In one case transotic approach was employed due to extensive labyrinth involvement, three cases of supra-labyrinthine petrous bone cholesteatoma had some labyrinth involvement and vertigo and decision to add labyrinthectomy to STP was made. The last patient underwent STP due to huge MEH occupying EEC, middle ear and mastoid, without apparent labyrinth involvement.

ABG indicates air-bone gap; AC, air conduction; BC, bone conduction; BP, bone pate; CHOL, cholesteatoma; COM, chronic otitis media; MEH, meningo-encephalic-herniation; PTA, pure-tone audiogram.

major intracranial-complication developed in the present series. All patients were symptom-free at last follow-up.

Two recurrent MEHs and one CSF-leak patients were elaborated in our previous paper (22). Additional recurrences occurred in the follow-up period, including two MEHs and three CSF-leak patients. All eight (3.1%) patients were revisited surgically.

CSF-leak: Patient 1: A patient of COM/CHOL-MEH having leak in tegmen-antri repaired by TMA presented with CSF-leak a month later with extrusion of cartilage and was revised by MCFA using a composite-graft. Patient 2: A patient of spontaneous-MEH who had multiple-sited defects were repaired using temporalis-fascia and cartilage by MCFA. Forty-five days postoperatively, CSF-leak between small defects was observed. This was repaired using muscle, bone-pate and human-fibrin-glue through TMA, which provided a complete closure. Patient 3: Rhinorrhea appeared 15 days after STP performed for multiple-sited spontaneous-MEHs and required another STP. Intraoperatively, vertical portion of the internal-carotid canal was uncovered with a discrete CSF-leak and was successfully managed by insertion of temporalis-muscle and human-fibrin-glue. The patients were symptom-free on the last follow-up after 96, 36, and 34 months, respectively.

The first case of recurrent MEH was initially treated by MCFA for a single, small, tegmen-mastoid located defect 4-month after trauma. The repair was performed with insertion of temporalis-fascia, temporalis-muscle, and human-fibrin-glue. The same patient presented 5 years later with a large, multisited MEHs with cholesteatoma. A revision HRCT revealed a longitudinal fracture-line, passing through the posterior-external-ear-canal, attic-aditus-ad-antrum with presumed post-traumatic skin-entrapment. STP was successfully employed with the multilayered closure of the defect. In the second case, initially a multisited large MEH due to the petrous-bone-cholesteatoma was repaired by STP in a multilayered fashion. There was a recurrence of cholesteatoma 4 years later and was removed by revision surgery. A medium-sized MEH was found and was managed effectively by an additional multilayer closure with employment of another STP.

DISCUSSION

Given the rarity and the overall cumulative experience with MEHs, a prompt diagnosis and optimal surgical management poses certain dilemmas. Although tegmental-floor dehiscence has been reported in 15–34% of autopsy cases (33–36), incidence of MEH is quite rare. To support this, at our center, out of 20,393 ear surgeries performed in the last 33 years, MEHs was verified intraoperatively in 262 patient (1.3%), with similar incidence being reported by others (1,7).

Etiology

The lack of tegmen-bone alone is usually insufficient for MEHs development, as dura itself is capable to support the brain over a larger bony-defect (22,37,38).

The proof lies in the fact that we regularly perform enlarged-translabyrinthine approach by removing whole tegmen-plate and have not come across any brain herniation postoperatively (39).

Theories of spontaneous-MEHs development are well described in the literature and include aberrant-arachnoid granulations (40), decades of CSF-pulsations (41), and congenital defects (42,43) as well as underlying associated idiopathic intracranial hypertension (2,13,23). In chronic otitis media, regardless of coincidental-cholesteatoma, many authors advocate the presence of granulations, as more important factor in violating both bony-tegmen and dura (13,22,26,44, 45), while cholesteatoma can create local-ischemia and dural enzymatic-degradation (46).

Diagnosis

The diagnosis is hastened by preoperative HRCT and MRI, as both are mandatory as well as complementary to each other (37,47). Whenever there is a suspicion of MEH on HRCT, an MRI is performed at our center. HRCT alone may not distinguish cholesteatoma, granulation tissue, cholesterol granuloma or any other soft tissue masses inside the middle ear or mastoid cavity and MRI on the other hand can distinguish all these conditions. Once the diagnosis is established, treatment is surgical. Main goals are to maintain/restore dural continuity and effectively divide intracranial and temporal-bone cavities, whenever possible, thereby avoiding further complications and recurrences (21,48).

Currently, 50% of patients were of COM/CHOL-MEHs, whereas it varied tremendously across the literature from 4.3 to 93.3% (2,11–13,21–23,25). One of the possible explanations could be the confusion in categorizing COM/CHOL-MEHs and iatrogenic-MEHs, since majority of the former underwent previous mastoidectomies (49–52). Seventy patients (56%) of current COM/CHOL-MEHs were previously operated.

Iatrogenic-MEHs differed between reports according to different authors due to the dilemma involved in branding a case as ''iatrogenic.'' While some strictly relied on previous operative-notes (2,22) where absence of intraoperative-injury resulted in decreased iatrogenic-group, others defined MEHs as ''iatrogenic'' whenever the patient underwent prior mastoidectomies, thereby rising this category (21). Due to a difficulty in determining whether MEHs occurred by chronic otitis media/cholesteatoma process or by previous surgery, the present study assigns all patients with active infection/cholesteatoma seen intraoperatively into COM/CHOL-MEHs.

Clinical Presentation

Hearing-loss and draining ear/otorrhea were currently the major presenting-symptoms, which is in line with the previous studies (Table 5). MEHs symptomatology was often mimicked by chronic ear set-up delaying the diagnosis by about 4 years in COM/CHOL-MEHs and iatrogenic-MEHs. In fact, 9% of these cases in the present study had symptoms for over 10 years.

Hearing-loss and tinnitus presented more in spontaneous-MEHs, which may have prompted ventilation-tube-insertion. Prolonged clear-otorrhea through ventilation-tube has often been confirmed by many as CSF-leak (9,12,13). In fact, ventilation-tube was inserted previously in about a third of spontaneous-MEHs, assuming them to be chronic serous-otitis-media and were then referred to our center with long-lasting clear-otorrhea (Table 2).

Although it appears that spontaneous-MEHs can be managed easily as it occurs without any underlying ear-pathology (26), we believe it is quite challenging to repair, owing to the underlying intracranial hypertension (2,13,23) and an increased-risk of recurrence (20,53,54).

Higher incidence of neurological or CSF-leaks (11,12) in spontaneous-MEHs and traumatic-MEHs allowed a more classical MEH presentation leading to less delay in diagnosis, especially for traumatic-MEHs (Table 2). The beta2-transferrin test was not routinely done at our center owing to clear clinical and imaging characteristics, which was in the agreement with previous studies (10–13,28).

Intraoperative Findings

One may expect typically a single (75%), medium-to-large-sized defect that principally involves tegmen-mastoid (60%), as observed in the current study, as well with others (2,11,12,14,21,55). Such a scenario is more probable in non-spontaneous-MEHs (Table 3) as supported by others (2,11,12,14) (Table 5).

Smaller, multiple, bilateral lesions can be expected in spontaneous-MEHs, with frequent tegmen-tympani involvement (60%), and is supported by other studies (9,11,12), who reported the involvement between 44 and 58% of cases (Tables 2, 3, and 5).

In our opinion, difference between preoperative CSF-ear drainage (20.6%), and intraoperative CSF-leak (41.9%) is presumably explained by the preoperative sealing-effect of MEHs in some, acting as a ''plug'' between intracranial and mastoid-cavity.

Hearing Outcomes

In COM/CHOL-MEHs and iatrogenic-MEHs the hearing postoperatively improved significantly, whereas it was not the same in spontaneous-MEHs and traumatic-MEHs. This could be attributed to the reversal of hearing-mechanism in these cases by ossiculoplasty performed after MEH-repair in 15% of these patients. Due to nonhomogeneity and lack of hearing-data in a substantial number of studies (Table 5), the comparison was impossible.

Surgical Strategy and Decision-making

Surgical approach is generally planned according to the size (1,7,21,27,28,56), site (1,13,24,37,55,56), number of defects (24,27), associated middle-ear infection,

TABLE 5. *Literature review of MEH*

						Surgery and MEH defect properties						Audiometry					
Author, year	No. of patients No. of surgery	Age mean, (range)	Signs and symptoms at presentation (%)	Etiology (%)	Bilateral (%)	Approach (n) %	Repair technique and material	Size mean (range)\(mm) (%)	Number (%)	Location (%)	Active CSF leak (%)	Preop. (dB) (mean)	Postop. (dB) (mean)	Complications (%)	Recurrence (%)	Revision surgery (%)	Follow-up mean (mo)
Jackson et al. (15), 1997	35, 35	38.5 (1–70)	Otorrhea (68.6), HL (51.4), Tinnitus (40), Vertigo (20), Otalgia (8.5), Seizures (2.8)	Iatrogenic (77.2), COM (11.4), Spontaneous (11.4)	(0)	Various approaches-unclear MEO (11.4%)	(extradural and intradural) TF (68.4), TF + muscle (10.5), TF+C (5.2), TF+B (2.6), TF + Proplast (2.6), TF+FG (13.1), C (2.6), Lyodura (2.6), none (5.2)	TA mean 11.8 TT mean 7.2	NA	TA (57.1), TT (34.3), TA+TT (8.6),	(31.4)	SRT (45.9) (n = 30)	SRT (52.7) (n = 30)	Seizure (7.9), Stroke (5.2), Sepsis (2.6), CSF leak (5.2), SNHL (10.4), Proplast extrusion (2.6)	MEH (8.6)	(8.6)	48.7
Mosnier et al. (16), 2000	15,15	53 (25–74)	Otorrhea\HL\Vertigo (46.6), Meningitis (26.6), Seizures (20), CSF leak (20), Hemiparesis (6.6), Temporal hematoma (6.6)	COM (60), CHOL (33.3), Iatrogenic (6.6)	(0)	TMA + MCFA (73), MCFA (27)	(extradural) TF+B+FG (100)	NA	NA	TA (33.3), TA (26.7), TT (20), TT+TA (20)	NA	NA	NA	(0)	(0)	(0)	24
Wotten et al. (4), 2005	12, 12	42.8 (19–72)	TM perf. (75), Draining ear (58), Cholesteatoma (50), TM atelectasis (25), Tinnitus (8), Vertigo (8)	Iatrogenic (100)	(0)	TMA (58), TMA + MCFA (42)	TF (intradural)	NA	NA	NA	NA	ABG (33)	ABG (28.1)	CSF leak (16.6), Cholesteatoma (16.6), TMA perf. (25)	CSF leak[a] (16.6)	TMA (25)	48
Gubbels et al. (6), 2007	15, 16	56.9 (6–81)	Clear otorrhea (80), HL (6.6), Meningitis (6.6), Brain abscess (6.6)	Spontaneous (100)	(13.3)	MCFA (100)	(intradural) B ± HA C ± TF ± FG (12) TF ± FG ± muscle (4)	not clear	Single (43.7), Two (12.5), Multiple (43.7)	TT (43.7), TA (6.3), TT+TA (50)	(81.3)	NA	NA	Corneal abrasion (6.3), CSF leak+ brain abscess (6.3)	CSF leak (6.3)	TMA\MCF-A (6.3)	13
Nahas et al. (7), 2008	15, 15	58.5 (31–77)	HL (93.3), Headache (33.3), Meningitis\brain abscess (6.6)	Spontaneous (100)	(33)	TMA+MCFA (100)	NA	13.4 (5–40) (n = 9)	Single (46.6), Two (53.4)	TT (21.5), TA (35.7), TT+TA (42.8) (n = 14)	(86.6)	ABG (25.4) (n = 10)	ABG (16.5) (n = 8)	(0)	MEH (6.6)	(0)	NA
Ramalingam et al. (32), 2008	13, 13	34.6 (7–56)	HL (100), Otalgia (46.2), Imbalance (38.5), Headache (23.1), Clear otorrhea (23.1)	Iatrogenic (100)	(0)	TMA (38.4), TMA+MCFA[b] (53.8) PC[c] (7.8)	(extradural) C+TF (38.4), TF+B+TF (53.8)	15 (7–30)	NA	TT (92.3), PCF (7.7)	NA	not clear	NA	NA	MEH (7.7)	(0)	(12–48)
Semaan et al. (8), 2011	31, 31 (27 MEH)	62	HL (80), Clear otorrhea (68), Headache (22), Clear rhinorrhea (16), Tinnitus (12), Meningitis (19), Brain abscess (6.5)	Spontaneous (90), COM (6), PT (4)	(0)	TMA (96.7) MEO (3.2)	(extradural) C+TF+ tissue sealant (100)	<10: (45), 10–20: (32), >20: (23)	Single (75), Two (21.4) Multiple (3.6)	TT (57), TT+TA (43) (n = 28) PCF (10)	NA	PTA-AC (39), PTA-BC (24), ABG (17) (n = 22)	PTA-AC (36), PTA-BC (24) ABG (12) (n = 21)	(0)	(0)	(0)	30
Stucken et al. (9), 2012	20, 20	57.4 (16–80)	HL (94.4), Clear otorrhea (60), Meningitis\brain abscess (20), Clear rhinorrhea (15)	Spontaneous (55), COM (20), PT (15), Iatrogenic (5), Hydrocephalus (5)	(0)	TMA+MCFA (70), MCFA (30)	TF+F+AlloDerm (50), muscle+TF +pericranium + B (45), fascia lata (5)	NA	Single (57.9), Two (10.5), Multiple (31.6) (n = 19)	TT (57.9), TA (63.2), PA (26.3)	(61.1) (n = 18)	ABG (15) (n = 16)	ABG (12) (n = 15)	CSF leak (5)	(0)	STP (5)	27.2
Kari et al, (10), 2011	56, 56 (42 MEH)	NA	Clear otorrhea (21), Pneumocephalus (3.6), Other—not clear	Spontaneous (59), Iatrogenic (21), PT (13), COM (7)	(0)	TMA (75), MCFA (7), TMA+MCFA (9), MEO (9)	(extradural) TF+B (% not clear) None (9)	NA	NA	NA	NA	NA	NA	CSF leak (1.8)	CSF leak (1.8)	TMA (1.8)	54 (n = 37)
Kenning et al. (17), 2012	23, 23 (22MEH)	55.1	HL (100), Clear otorrhea (83), Meningitis (17), AOM (4), Tinnitus (4)	PT (22), Iatrogenic (9.1) COM (4.3), Spontaneous- not clear	(43)	TMA+MCFA (100)	(intradural) DS (96), (extradural) pericranium\TF (87) + B (48), DS (9), B (4.3)	NA	NA	NA	(83)	NA	NA	Seizures (4.3), Meningitis (4.3), DVT (4.3), Ossicular fixation (4.3), Wound infection (4.3)	CSF leak (4.3)	(0)	10.4

(Continued on next page)

TABLE 5 *(Continued)*

						Surgery and MEH defect properties						Audiometry					
Author, year	No. of patients No. of surgery	Age mean, (range)	Signs and symptoms at presentation (%)	Etiology (%)	Bilateral (%)	Approach (n) %	Repair technique and material	Size mean (range)\(mm) (%)	Number (%)	Location (%)	Active CSF leak (%)	Preop. (dB) (mean)	Postop. (dB) (mean)	Complications (%)	Recurrence (%)	Revision surgery (%)	Follow-up mean (mo)
De Zinis et al. (49), 2013	18, 18 (9 MEH)	53 (23–77)	HL (66.6), Otorrhea (55.5), Vertigo (27.7), FN paralysis (5.5)	Iatrogenic (61.1), CHOL (38.8)	(0)	TMA (100)	(extradural) BP + C + periosteum (100), addition of TF+FG prior BP (30)	<10: (27.7), 10–20: (61.1), >20: (11.1) (mean 13.1)	NA	TA (55.5), TT (38.8), TA (38.8), multiple (22.2)	(30)	ABG (30)	ABG ≤20 in 56% pts	(0)	(0)	(0)	61
Carlson et al. (18), 2013	86, 89 (85 MEH)	52.3 (9.3–76)	HL (92), Clear otorrhea (73), Tinnitus (42.7), Dizziness (17.4), Headache (14), Clear rhinorrhea (9), Meningitis (5.6), Seizures (2.2), Brain abscess (1.1)	Spontaneous (39.3), COM (27), Iatrogenic (18), PT (14.6)	(14.6)	TMA+MCFA (72), MCFA (28)	(mainly extradural) TF+C\B+TF\DS (98.9), titanium mesh (12.4), one-layer (1.1)	<10: (51.4), 10–20: (26.4), >20: (22.2)	Single (48.3), Two (15.7), Multiple (36)	TT+TA (55.1), TT (39.3), TA (5.6)	(80) (n = 85)	ABG > 10 (52 \71)	PTA ≤10 in (57.8) pts, improved (24.4) pts, worse (17.8) pts	Death of MI (1.1), Seizures (1.1), Meningitis (2.2), CSF-leak (7.9), FN palsy (1.1), Wound infection (3.4)	CSF leak (7.9)	TMA\MCF-A (1.1)	14.5
Wahba et al. (19), 2013	12, 12	35.7 (22–55)	Otorrhea (66.6), Meningitis (25), Brain abscess (8.3)	Iatrogenic (75), PT (25)	(0)	TMA (100)	(extradural), C + muscle + FG + TF (100)	5.6, (2.2–15)	NA	TA (75), TT+TA (25)	NA	NA	NA	Wound infection (8.3)	(0)	(0)	19.2
Jeevan et al. (20), 2015	32, 34	51 (5–74)	HL (78.1), Otorrhea (31.25), FN paralysis (6.2) Meningitis (6.2), Seizures (3.1)	Spontaneous (34.4), Iatrogenic (25), COM (18.8), CHOL (9.4), Tumor (6.2), PT (3.1), RT (3.1)	(0)	TMA+MCFA (100)	(extradural\intradural) dura-repair with sutures\ DS, B+TF+FG (100)	NA	NA	NA	(37.5)	NA	NA	Graft infection (3.1), SNHL (3.1)	(0)	TMA\MCF-A (3.1)	58.8
O'Connell et al. (11), 2017	26, 26 (24 MEH)	60.5 (25–85)	NA	Spontaneous (50), COM\Iatrogenic (50)	(0)	TMA+MCFA (100)	(extradural) loose areolar tissue +B \C+DS (100)	6.8 (3.1–11.7)	Single (57.7), Two\Multiple (42.3)	TT+TA (69.2), TA (19.2), TT (11.5)	(76.9)	PTA-AC (48.6) PTA-BC (24.3) ABG (24.7)	PTA-AC (39.2), PTA-BC (22.3), ABG (18.9) (n = 19)	Wound seroma (3.8)	(0)	(0)	8.3
Present study	249, 262	49.7 (9–82)	HL (94.6), Draining ear (44.3), Vertigo (20.6), Clear otorrhea (20.6), Clear rhinorrhea (5.7), Meningitis (10.7), Brain abscess (1.5), Seizures (1.5)	Spontaneous (23.3), COM ± CHOL (47.7), Iatrogenic (20.9), PT (8.0)	(9.9)	TMA (32.8), MEO (46.2), MCFA (19.1), TMA+ MCFA (1.9)	(extradural) TF+C ± M ± HFG (42.7), TF+B ± M ± HFG (20.6), None (17.2), TF+M+HFG (11.1), TF+C+BP+HFG (7.3), TF+FG+HFG (1.1)	≤10: (30.2), 10–20: (37.8), >20: (32.1)	Single (75.2), Two (10.7), Multiple (14.1)	TA (32.1), TA (17.9), TT (8.0), PCF (2.7), EEC (0.4), multiple (38.9)	(41.9)	PTA-AC (56.7), PTA-BC (27.8), ABG (28.8) (n = 228)	PTA-AC (55.6), PTA-BC (29.9), ABG (25.7) (n = 196)	Profound SNHL (0.4), Meningitis (0.4), CSF-leak (1.1), Extradural subtemporal hematoma (0.4), Abdominal hematoma (0.4)	MEH (1.5), CSF leak (1.5)	MCFA (0.4), TMA (0.8), STP (1.9)	39.9

[a]Recurrent CSF-leak from the other site, TM: Tympanic membrane.
[b]Two of combined approach underwent subtotal petrosectomy (15.4%).
[c]Posterior craniotomy.

ABG indicates air-bone gap; AC, air conduction; AOM, acute otitis media; B, bone; BC, bone conduction; BP, bone pate; C, cartilage; CHOL, cholesteatoma; COM, chronic otitis media; CSF, cerebro-spinal; DS, dural substitute; DVT, deep vein thrombosis; F, fat; FG, fibrin glue; FN, facial nerve; HAC, hydroxyapatite cement, temporal bone; HFG, human fibrin glue; HL, hearing loss; MEO, middle-ear obliteration; MCFA, middle-cranial fossa approach; MI, myocardial infarction; NA, not available; PA, petrous apex; PCF, posterior cranial fossa; PT, posttraumatic; PTA, pure-tone audiogram; pts, patients; RT, radiotherapy; TA, tegmen antri; TF, temporalis fascia; TM perf, tympanic membrane perforation; TMA, transmastoid approach; TT, tegmen tympani.

hearing (27,37,55), MEH-volume, and surgeon experience (28).

MCFA was our first-choice for spontaneous-MEHs (53%) and traumatic-MEHs (38%), as they had predominantly anteromedial defects and intact ossicular-chain (Table 1), which permitted best exploration of other possible defects. Multiple defects were seen in 40% of spontaneous-MEHs and 24% of traumatic-MEHs. (Tables 1 and 3). Furthermore, it assisted in stable extradural-repair with precise graft-placement, as observed with others (2,11,22,23,32,48). In our center, MCFA used for this purpose does not require the use of MCF-retractor. Middle-meningeal artery was usually not coagulated, as defects were mostly postero-lateral to this foramen. Postoperative prophylactic antiepileptics further minimized neurological complications.

As a quaternary-referral center, 50.4% of cases were previously operated elsewhere and 21% of them were plurioperated. Given the background of chronically infected, plurioperated cases, lack of healthy middle-ear/mastoid-cavities and maximal conductive-loss with limited possibility for hearing reconstruction, we believe that MEO serves as a viable option for such cases.

Though as a last-resort surgery in the literature, MEO was currently the preferred option in 46% of cases as well as in 38% as revision surgeries. In the literature, MEO was used in 11 cases, which accounted for 3.2 to 15.4% of the primary surgery (1,11,13,21,26) and was employed in plurioperated, inner-ear involvement cases and in one case of revision surgery (12) (Table 5).

In the present study, MEO was principally, employed in half of COM/CHOL-MEHs and iatrogenic-MEHs, wherein a large (70%) and multisited (49.5%) defects with complex otologic-history were present. In iatrogenic-MEHs, surgery could have been avoided if the previous mastoidectomies were performed maintaining dura and bone integrity (1), with immediate repair at initial surgery (22,27,28). Interesting speculation lies in the fact that iatrogenic-MEHs occurred almost twice in the right ear, perhaps due to more forceful drilling done by a right-hand surgeon, as reported also in other series (21).

When favorable middle ear conditions with small, postero-lateral defect was the scenario, TMA was a feasible choice (Table 1), rendering minimally morbid operation, shorter hospital-stay and option for concurrent-ossiculoplasty. Furthermore, MEO/TMA was used in cases of posterior-fossa plate involvement (9.5%) as well.

Spontaneous-MEHs patients principally do not hold underlying mastoid-disease and yet the fact that MEO was employed in 36% of cases, deserves special attention. All were difficult cases of medium-to-large multisited MEHs and five cases were revision surgeries (Table 2). PCF defect was present in three cases; another three had uncovered vertical carotid-canal surrounded by long-standing MEHs; in four cases, substantial dura and bony-loss gave no possibility for a repair. In one case, long-standing herniation caused labyrinthine erosion. In yet another case, a more anterior MEHs necessitated infra-temporal fossa B approach. Out of remaining five cases, one had profound-deafness and four had mixed hearing-loss.

Even though a 10-year period of follow-up with HRCT and MRI is mandatory after MEO, due to occult chance of cholesteatoma recurrence (58), it is our recommendation to perform this surgery due to various advantages. In addition to providing superior quality of life, water activities can be resumed without restrictions. The drawback of hearing-loss can be addressed with BAHA in deserving cases.

Contrary to the literature (Table 5), combined-approach was limitedly used (1.9%), as in our experience small, postero-lateral-defects were dealt with TMA, and a larger, antero-medial-defect in patients without previous mastoidectomies were managed effectively by MCFA, as supported by others (58).

Dural quality was often poor adjacent to the bony-defect, especially in a case with coexisting infection and remote edges, thereby compromising intradural-repair. Multilayered autologous tissue was employed for extradural-repair is recommended by previous authors as well (2,12,59,60). Principally in about 70% cases, solid materials were added for proper separation of the two cavities (11,26,27,48). In 17.2% cases, managed by MEO, the repair was not performed (Table 1).

Complication, Recurrence and Revision surgery

Complication rate across the literature varied between 1.8 and 58.2% with neurological complications in 6.3 and 15.7% and CSF-leak in 1.8 and 16.6% of cases. Currently, out of 2.7% complications encountered, meningitis occurred in 0.4% and CSF-leak in 1.1% of cases. Few studies have reported use of lumbar drains (10,11,13). Although not used currently, recurrence-rates in the present study were 1.5% both for MEHs and CSF-leaks, which is positively comparable with other reports (Table 5). Revision-surgery was performed in 3.1% of cases whereas it was 1.1 and 25% of cases in the other series (Table 5).

CONCLUSION

Although the current study highlights a rare skull-base problem, any active otologic-clinic is bound to encounter MEHs quite regularly. One should be vigilant in a case of prolonged clear-otorrhea in ventilation-tube-insertion for a CSOM and should keep in mind MEHs possibility in any chronic or operated ear, as the timely diagnosis minimizes future complications. Dealing with a patient in a time frame between diagnosis and treatment, incorporating the MEHs-etiology could be helpful in patient-consultation and guiding toward the right-surgical choice. MEO is a part of armamentarium of every otologist and should be used without hesitation whenever needed, if the objective is performing a definite surgery.

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