Isolated Congenital Mastoid Cholesteatoma with no Involvement of Aditus Ad Antrum and Middle Ear

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ABSTRACT: Cholesteatoma is a non-neoplastic, keratinized squamous epithelial lesion that affects the temporal bone. The middle ear is the most frequent, while the isolated cholesteatoma of the mastoid is rare. The aim of this study was to describe a rare case of isolated mastoid cholesteatoma with no involvement of aditus ad antrum and middle ear including a literature review of the topic. This case report describes the case of a 58 years old female with a cholesteatoma isolated in the mastoid region, evidenced by imaging (computer tomography and magnetic resonance). A mastoidectomy was performed: mastoid process was completely involved, but antrum was not reached. Moreover, it reached the soft tissue of stylomastoid foramen as well as the posterior belly of the digastric muscle. In the literature few articles described cases of cholesteatoma isolated in the mastoid region. Research was conducted using PubMed and reference list and there were considered only reports about cholesteatoma exclusively located in the mastoid process without involvement of antrum or middle ear. Fourteen articles were included in this review, with a total number of 23 cases of cholesteatoma isolated in the mastoid region. All papers analyzed reported the cases of isolated mastoid cholesteatoma that presented a congenital origin. Its diagnosis is difficult, therefore, imaging evaluation is mandatory and surgery is the treatment of choice. Mastoid cholesteatomas without involvement of aditus ad antrum and middle ear are rare and only 23 cases are reported in literature. Our case is in line with all clinical and diagnostic features of this rare disease, but it is the only one that evidenced an exposure of the soft tissue of stylomastoid foramen as well as the posterior belly of the digastric muscle. The treatment of choice was the surgical one, avoiding damaging of important anatomo-functional structure.

KEYWORDS: Cholesteatoma, mastoid, mastoidectomy, review

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Introduction

Cholesteatoma is a non-neoplastic, keratinized, squamous epithelial lesion that affects the temporal bone. Its annual incidence is 3 per 100.000 in children and 9.2 per 100.000 in adults.1 It involves different temporal bone areas: middle ear, petrous apex, mastoid, external auditory canal. The middle ear is the most frequent site while isolated cholesteatoma of the mastoid is rare.²

The aim of this study was to describe a rare case of isolated mastoid cholesteatoma with no involvement of aditus ad antrum and middle ear, including a literature review of the topic.

Materials and Methods

Case report

A 58-year-old woman came to our Department with a one-year history of pain in the region of the right mastoid process and upper neck. She denied previous otorrhea, ear disease, perforation of the tympanic membrane or ear surgery. Otoscopy showed normal tympanic membranes and facial nerve function was grade I HB (House-Brackmann grading system). Pure-tone audiometry evidenced minimal symmetric sensorineural hearing loss.

Computed Tomographic (CT) scan of the temporal bone revealed an expansive destructive lesion in the right mastoid process. The middle ear, attic, aditus, and antrum were disease free (Figure 1).

On magnetic resonance imaging (MRI), the lesion appeared hypointense on T1- weighted images and hyperintense on T2-weighted images with a size of $20 \times 31 \text{ mm}$ (Figure 2). Diffusion weighted imaging (DWI) sequences demonstrated a restricted diffusion in the mastoid region.

A mastoidectomy was performed: the mastoid process was completely involved, but the antrum was not reached.

The bone covering the middle fossa dura, presigmoid, and retrosigmoid posterior dura as well as that of the sigmoid sinus (SS) was completely eroded (Figure 3). The cholesteatoma passed under the mastoid portion of the Fallopian canal without any involvement of the facial nerve, reaching the jugular bulb, and the jugular foramen. It was in close contact with the soft tissue of the stylomastoid foramen as well as the posterior belly of the digastric muscle (Figure 4). Surgical manipulation allowed us to identify a cleavage plane between the matrix and all of the previous described anatomical structures. Hence, complete removal was achieved respecting these structures.

Immediate post-operative facial nerve function remained grade I HB, and hearing maintained its preoperative level. After 6-month follow-up the patient was disease-free.

Review

In the literature there are few articles describing cases of isolated cholesteatoma in the mastoid region. Research



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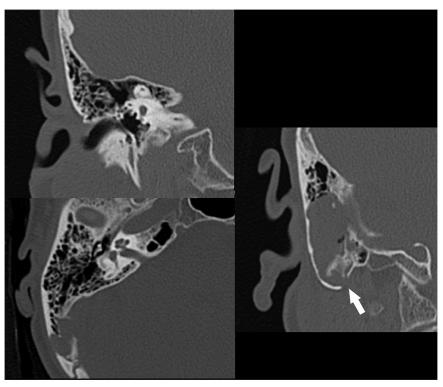


Figure 1. Computed Tomography in axial and coronal view gave evidence of the extention of the mastoid cholesteatoma without involvement of middle ear and aditus ad antrum. Erosion of the mastoid process at the level of digastric ridge, and the exposed stylomastoid foramen (white arrow).



Figure 2. Magnetic resonance imaging. T2-weighted images showed hyperintense lesion with a size of 20×31 mm (arrow).

was conducted using PubMed and reference list, identifying articles according to inclusion anatomy, histology, embryology and congenital pathology of the ear. The keywords used for research were: mastoid, cholesteatoma. Only reports describing cholesteatoma exclusively located in the mastoid process without involvement of the antrum or middle ear were considered.

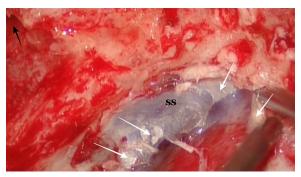


Figure 3. Intraoperative view. Aditus ad Antrum (black arrow). Matrix of Cholesteatoma (white arrows). Abbreviation: SS, sigmoid sinus.

Results

Fourteen articles were included in this review.²⁻¹⁵ To the best of our knowledge, the total number of cases of cholesteatoma isolated in the mastoid region to be reported in the literature, regarded 23 patients (Table 1). All the papers analyzed described cases of isolated mastoid cholesteatoma that presented a congenital origin. The mean age at diagnosis was 51 years old (range 13-87): 22 patients underwent surgical treatment, 1 was submitted to watchful waiting.

Discussion

Cholesteatoma is a non-neoplastic, destructive, locally invasive mass composed of keratinized squamous epithelial lesions. It is usually unilateral and may affect different areas of the temporal bones: middle ear, mastoid, petrous apex, external auditory channel.¹

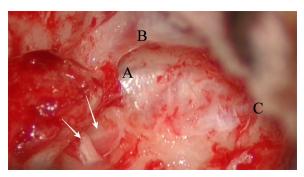


Figure 4. Intraoperative view. Matrix of Cholesteatoma (arrows). (A) Soft tissue separating the temporal bone from the neck after matrix removal. The bone was completely eroded by cholesteatoma, (B) Stylomastoid Foramen. (C). Digastric ridge.

In the literature only 14 papers describing isolated cholesteatoma of the mastoid process are reported and all were considered to have a congenital origin.

Congenital cholesteatoma of the mastoid process (MCC), is defined as cholesteatoma with all the features of a CC (normal tympanic membrane, no previous ear surgery, no history of ear discharge) and no involvement of the middle ear, attic, or aditus as confirmed by radiologic and intraoperative findings.² Our case presented all these characteristics.⁴

Origin

The origin of congenital cholesteatoma is still a source of debate. The most popular explainations for the presence of epidermal tissue behind an intact TM are: the migration theory; the amniotic fluid contamination theory; the inclusion theory and the epidermoid formation theory.¹⁷⁻²⁰

Previously Warren et al⁹ and later Hong et al,¹³ compared all these theories and stated that none of them provided an exhaustive explanation of MCC origin. In accordance with these authors, we think that the theory defined "Implantation theory," is the most suitable one for these cases. During the fontanel closure process, in fact, the persistence of squamous epithelium trapped within the suture could generate MCC.²¹

Epidemiology and clinical presentation

The mean age of CC presentation in the overall population is 6.58 years (range 3 months to 77 years),²² while the mean age of presentation in cases of isolated mastoid congenital cholesteatoma, without involvement of aditus ad antrum and middle ear, is 51 (range 13-87) with a predominance in elderly age. This difference could be associated with the absent or late onset of the symptomatology. In contrast, the diagnosis of middle ear cholesteatoma is usually made during childhood. In fact, our case presented the first symptoms at the age of 58 years.

Moreover, as reported in Table 1, three patients did not present any symptoms and the diagnosis was incidental. The case reported by Sepehri and von Unge¹⁵ of an 87 year- old man is emblematic.

In the symptomatic stage, signs are non-specific with retroauricular and/or neck pain, retroauricular swelling, or dizziness.

The origins of retroauricular and/or neck pain and retroauricular swelling are related to mastoid erosion and periosteum involvement with possible consequent inflammation of the attached muscles. In the case reported, the patient presented a not well localized pain that involved not only the mastoid process but also the latero-cervical region. This could have been attributable to involvement of the posterior belly of the digastric muscle.

Dizziness is another symptom of isolated mastoid cholesteatoma, despite our patient not presenting it. The etiopathogenetic theory relates it to the compression of ipsilateral cerebellar hemisphere or endolymphatic sac.^{2,6}

Stenosis of the external auditory canal (EAC), ear pain or hearing loss were present in a few cases. 10,12,13

Diagnosis

Since MCC diagnosis is difficult, imaging evaluation is mandatory. Otoscopy as well as pure-tone audiometry do not show any particular alterations, as in the present case.

Luntz et al,⁴ proposed 3 characteristic features for diagnosis: an initial presentation of neck pain; CT findings of a cystic, expansive lesion occupying the mastoid process without involving the middle ear; MRI findings of hyperintensity on T2-weighted images with little or no peripheral enhancement on post-contrast T1-weighted images.

In all the papers analyzed including ours, CT scan presented non-specific expansive destructive lesions in the mastoid process that had no communication with the middle ear. Different structures may be involved: the facial nerve, sigmoid sinus, jugular bulb, posterior cranial fossa dura, and the endolymphatic sac. This nonspecific evidence could be confused with epidermoid cyst, intradiploic cyst, cholesterol granuloma or anomalous sigmoid sinus. MRI and subsequent histological examination are fundamental.²

On MRI cholesteatoma is hypointense or isointense on T1 weighted images and hyperintense on T2 weighted images, without enhancement after gadolinium infusion.⁷

Other important investigations are MRI angiographic sequences for estimating the patency of the sigmoid sinus and the jugular bulb on both sides. This is useful for identifying preoperatively which side is dominant if it becomes necessary to obliterate the SS during surgery without any complications.

On the contrary, temporal bone tumors enhance after gadolinium infusion, whereas cholesterol granulomas presented a bright signal intensity on both T1-weighted and T2-weighted images and DWI sequences, with a restricted diffusion and high signal intensity. Epidermoid cyst and intradyploic cyst have a low signal intensity on T1-weighted images, high signal intensity on T2-weighted images and high signal intensity on diffusion-weighted images.

Table 1. Case Summaries.

AUTHORS	°2	AGE	SYMPTOMS	STRUCTURES INVOLVED	\$ INVOLVED					SURGERY
	L.			SS	POSTERIOR WALL OF EAC	POSTERIOR FOSSA DURA	MILLE FOSSA DURA	FACIAL NERVE	JUGULAR BULB	
Delacki and Clemis ³	-	24	Pain and swelling	NA	Eroded	Exposed	Exposed	NA	NA	Atticomastoidectomy
Luntz et al ⁴	-	54	Neck pain, instability	Exposed	Intact	Exposed	Exposed	NA	NA	CWU
Cüreoglu et al⁵	-	70	Neck pain, retroauicolar swelling	NA	Eroded	Exposed	NA	NA	NA	Modified radical mastoidectomy
Mevio et al ⁶	-	36	Dizziness	Occluded	AN	Exposed	Exposed	NA	NA	Modified radical mastoidectomy
Thakkar et al ⁷	-	NA	NA	Exposed	NA	NA	NA	NA	NA	Surgical approach NA
Lee et al ⁸	-	57	Dizziness, swelling, pain, headache, othorrea	Exposed	Eroded	Exposed	NA	NA	NA	Mastoidectomy
Warren et al ⁹	e	30	Incidental findings	Exposed	NA	Exposed	Exposed	Exposed	NA	Transmastoid aprroach
		13	Neck mass	NA	Eroded	NA	NA	Exposed	NA	CWD, neck dissection
		28	Ear pain	Dehiscent	NA	NA	NA	NA	NA	Surgical approach NA
Hidaka et al¹ ⁰	-	65	Swelling and pain	Intact	NA	Exposed	NA	NA	NA	Atticomastoidectomy
Giannuzzi et al²	ю	71	Mastoid and neck pain	Occluded	Intact	Exposed	NA	Exposed	NA	Mastoidectomy + SS packing + neck dissection
		77	Dizziness	Exposed	Intact	Exposed	NA	Exposed	NA	Mastoidectomy
		60	Incidental findings	Exposed	Intact	Exposed	NA	Exposed	NA	Mastoidectomy + SS packing
Granato et al ¹¹	-	67	SHL, tinnitus	Exposed	Intact	NA	NA	NA	NA	Surgical approach NA
Nagato et al ¹²	-	10	Stricture of EAC	Intact	Intact	Intact	NA	NA	NA	Mastoidectomy
										(Continued)

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Table 1. (Continued)

AUTHORS	°N	AGE	SYMPTOMS	STRUCTURES INVOLVED	INVOLVED					SURGERY
	Ы			SS	POSTERIOR WALL OF EAC	POSTERIOR FOSSA DURA	MILLE FOSSA DURA	FACIAL NERVE	JUGULAR BULB	
Hong et al ¹³	-	59	Ear discharge	Intact	Eroded	Intact	NA	NA	NA	Mastoiidectomy
Cvorovic et al ¹⁴	Q	AN	Retroauricolar pain and mild hearing loss	Eroded	Eroded	Exposed	Exposed	Exposed	AA	Retroauricular transmastoid perilabyrinthine mastoidectomy wall down
		NA	Ear discharge	Eroded	Eroded	Exposed	Exposed	Intact	NA	Retroauricular mastoidectomy wall down
		Child	Hearing loss in both ears	Intact	Intact	Intact	Intact	Intact	NA	Retroauricular mastoidectomy wall up
		Child	Hearing loss	Intact	Intact	Intact	Intact	Intact	NA	Retroauricular mastoidectomy wall up
		NA	Ear discharge	Intact	Eroded	Intact	Intact	Intact	NA	Retroauricular, mastoidectomy wall down
		60	Headache, earache, fever	Eroded	Intact	Exposed	Exposed	Intact	NA	Retroauricular mastoidectomy wall down
Sepehri et al ¹⁵	-	87	Incidental	NA	NA	NA	NA	NA	NA	Watchful waiting
Pace et al. (2021) ¹⁶	-	58	Neck pain	Exposed	Intact	Exposed	Exposed	Intact	Exposed	Enlarged mastoidectomy

Abbreviations: pt, patients; SS, sigmoid sinus; EAC, external auditory channel; M, male; F, female; NA, data not available.

Moreover, granulation/fibrous tissue, cholesterol granuloma, and serous fluid show low signal intensity on DWI.

Histological classification differentiates: dermoid cysts composed of both ectoderm and mesoderm layers; cholesteatomas of only ectodermal origin.²³

Isolated CC of the mastoid should also be differentiated with tympano-jugular paragangliomas, tumors of the endolymphatic sac, and meningiomas. Paragangliomas, in fact, are osteolytic and invade the sigmoid sinus, but they more frequently involve the infralabyrinthine region rather than the retrolabyrinthine 1 and are easily identified by contrast-enhanced imaging.

On MRI-DWI sequences, cholesteatomas all manifest as significant high signals, far higher than brain parenchyma: it is very striking owing to the low signal of the surrounding cerebrospinal fluid, making it very easy to be found. Therefore, it is easy to distinguish it from non-restricted lesions such as arachnoid cyst or craniopharyngeal tumor. Tumors of the endolymphatic sac are rare and show up as a heterogeneous gadoliniumenhanced mass on T1-weighted magnetic resonance images due to a remarkable vascularization among the foci of tumoral tissue. Finally, meningiomas do not usually present increased bone density with osteolysis.

Surgical key point

Surgery is the treatment of choice for cholesteatoma and mastoidectomy is the treatment most indicated for MCC.²

As previous described, cholesteatoma is a destructive disease that may erode the bony covering of important anatomical structures such as dural walls (posterior and middle fossa), sigmoid sinus, facial nerve and jugular bulb.

In our case, the bone covering the middle fossa dura, presigmoid, and retrosigmoid posterior dura was completely eroded, but an intact dura was exposed and no detectable CSF leakage was detected. A complete dissection of the matrix from the dura wall surface was performed, preserving it. When dural involvement is suspected, bipolar coagulation of the suspected dural portions is recommended for devitalization of the cholesteatoma matrix.²⁴

The bone covering the sigmoid sinus was also eroded and during surgery every effort was made to keep damage of this structure to a minimum. It is known, in fact, that veins of the brain have no muscular tissue in their very thin wall, meaning that the risk of rupture is very high.²⁵

In the literature, cases with thrombosis or/obstruction of the SS caused by MCC which required packing, are reported.^{2,6} In our opinion, it is important to remember that some contraindications exist for resection/packing of the sinus: absence or significant asymmetry of the contralateral transverse and sigmoid sinuses, with the affected side being the dominant flow system; limited thrombosis of the sinus that does not require aggressive management; presence of an intracranial complication that raises intracranial pressure (meningitis, brain abscess, subdural abscess). Furthermore, since the matrix of cholesteatoma may be very

adherent to the venous wall, in some cases it is better to leave some residual components in order to reduce risks for the patients' life.

In the literature, six cases in which the facial nerve was exposed in its third portion were reported. Patients did not usually present any preoperative facial palsy and the nerve was anatomically and functionally preserved. In our case, the cholesteatoma had passed under the mastoid portion of the Fallopian canal without any involvement of the facial nerve, reaching the jugular bulb and the jugular foramen. At the same time, it was in close contact with the soft tissue of the stylomastoid foramen as well as the posterior belly of the digastric muscle and, to the best of our knowledge, it is the only case in which this relationship is reported.

In only a few cases removal of the cholesteatoma was associated with a neck dissection. It was performed by Giannuzzi et al² to control the jugular vein during the removal of a cholesteatoma with massive involvement of the sigmoid sinus and the jugular bulb. On the other hand, Warren et al⁹ accomplished a neck dissection and parotidectomy to remove a cholesteatoma that presented a neck mass. In our case, despite the large size of the MCC, it was totally detachable via mastoidectomy enlarged to the mastoid tip.

Conclusions

Mastoid cholesteatomas without involvement of aditus ad antrum and middle ear are rare and only 23 cases are reported in literature. Our case is correspondent to all the clinical and diagnostic features of this rare disease. In fact, despite its large size it remained within the mastoid process without any involvement of the aditus ad antrum and middle ear. However, it is the only case reported in which there was exposure of the soft tissue of the stylomastoid foramen as well as the posterior belly of the digastric muscle. The treatment of choice was surgery, paying the utmost attention to avoid the damage of the important anatomo-funcional structures encountered during dissection.

Author Contributions

Giuseppe Magliulo: made a substantial contribution to the concept and design of the work.

Annalisa Pace: Interpretation of data; Drafted the article

Giannicola Iannella, Daniela Messineo: Acquisition and analysis of data

Valeria Rossetti, Alessandro Milani: revised it critically for important intellectual content.

Irene Claudia Visconti, Roberta Polimeni: Approved the version to be published.

Each author had participated sufficiently in the work to take public responsibility for appropriate portions of the content.

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