

Temporal bone Cholesterol Granuloma

Abstract

Cholesterol granulomas are cystic lesions that typically arise in the petrous apex as a result of an inflammatory giant-cell reaction to cholesterol crystal deposits that are formed when normal aeration and drainage of temporal bone air cells become occluded, it could be asymptomatic but aggressive CG could produce audiologic, vestibular and neurologic symptoms, diagnosis done by MRI, and surgery indicated for symptomatic patients with need for long follow up due to the possibility of recurrence.

Review Article

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Omar Ramadan*

ENT Registrar, Al Adan Hospital, Kuwait

*Corresponding author: Omar Ramadan, ENT Registrar, Aladan Hospital, Kuwait, Email: omarram982@hotmail.com

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Introduction [1-7]

Manasse was first reported cholesterol granuloma of the middle ear in 1917. Shambaugh in 1929 described idiopathic hemotympanum as a cause to the development of cholesterol granuloma of middle ear. Cholesterol granuloma have been seen in multiple area of the human body where cholesterol crystals may exist such as the lungs, breasts, peritoneum, mediastinum, liver, spleen, thyroid, kidneys, lymph nodes, testis, facial skeleton, skull, and the temporal bones. It has also been seen the skull, in the paranasal sinuses mostly in the frontal sinus. The most common site of Cholesterol granuloma is temporal bone. There is some literatures reported intracranial Cholesterol granuloma in temporal lobe, and CPA. Bilateral middle ear cholesterol granuloma was also reported, it is rare in children.

Incidence [1-8]

Cholesterol granuloma affects young and middle-aged adults mostly, the incidence of petrous apex CG is not certain, Sabin et al., reported six cases of cholesterol granuloma among nine cases of petrous apex cystic lesions seen over 20 year period. Middle ear CG has reported in 12% of chronic otitis media patients with intact TM and in 21% with perforated TM. CG was also reported in TM, EAM and Mastoid Cavity.

Experimental Cholesterol Granuloma [9-19]

- In 1957, Ojala [9] introduced cholesterol granuloma in chick humeral by obstructing the ventilation.
- In 1959, Fridmann [10] injected sterile suspension of cholesterol into bulla of guinea pigs; he found that cholesterol granuloma developed, but that choleostoma did not.
- In 1963, Dota et al. [11] produced cholesterol granulomas in the tympanic cavity of the rabbit by repeated injections of 1 per cent of oxalic acid solution.
- In 1966, Beaumont [12] induced cholesterol granuloma following the experimental exclusion of air from pneumatized bones using the pneumatized hummers of the domestic fowl.
- In 1970, Main et al. [13] created experimental cholesterol granulomas in squirrel monkeys by obstructing the Eustachian tubes for 6-12 months.

- In 1979, Goycoolea et al. produced cholesterol granulomas in cats. In the same year, Kuijpers et al. experimentally induced middle ear effusions and cholesterol granulomas 20 months after tubal obstruction in germ-free rats.
- In 1980, Hiraide et al. [14] produced experimental cholesterol granuloma in the middle ear of guinea pigs by injection only one dose, transtympanically, of a saturated solution of cholesterol in absolute alcohol.
- In 1982, Hermann et al. produced experimentally induced middle ear effusions and cholesterol granuloma in chicken hummers. In the same year, Hiraide et al. [14] experimentally induced middle ear effusions and cholesterol granuloma ≥ 1 month after Eustachian tube obstruction in squirrel monkeys and Paparella et al. [8] created cholesterol granuloma from long-term Eustachian tube obstruction in cats, monkeys, guinea pigs and chinchillas.

Summarizing, there are three primary factors responsible for the formation of a CG, local hemorrhage which can happen during an inflammation, obstruction of middle ear ventilation and poor drainage of middle ear cavity [15-19].

Pathology [20]

Gross Pathology CG looks like slow growing expansible benign cystic lesion that contains brownish-yellow debris, those lesion can be found in any body part that contain cholesterol crystals. Microscopically Cholesterol Crystal induces the accumulation of giant cells which is responsible for the tissue reaction electron Microscopy; giant cells display large numbers of mitochondria and smooth endoplasmic reticulum surrounding the small needle or dart-shaped cholesterol crystals. Large numbers of lysosomes are present (Figure 1).

Clinical Presentation of Temporal Bone Cholesterol Granuloma [21-24]

Temporal bone CG can appear in three forms associated with chronic otitis media (especially cholesteatoma), idiopathic hemotympanum, or a localized disease in the middle ear cavity, antrum of mastoid, external auditory canal, and petrous apex. CG can happen without any history of infection. Within the temporal bones, cholesterol granuloma is happening commonly in the petro

us apex. A petro us apex cholesterol granuloma (PACG) is the most common lesion of the petro us apex. Muscle et al. The clinical presentation of petros apex cholesterol granuloma depends on the extension.

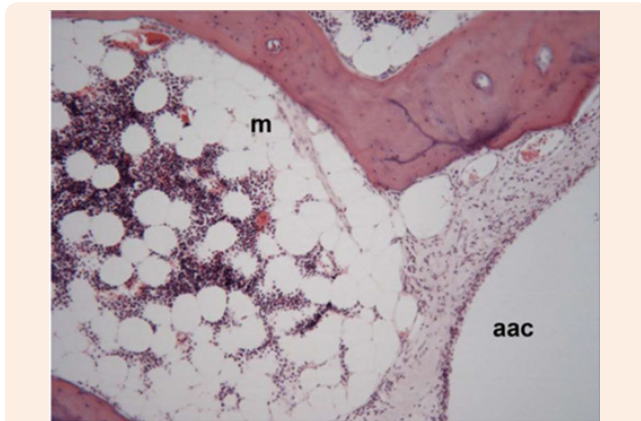


Figure 1: Hematopoietic marrow (m) adjacent of an apical air cell (aac) in an unrelated case (H&E X100).

The most common presenting symptom of PACG was hearing loss (64.7%) vestibular symptoms reported in (56%), tinnitus reported in (50%), headache reported in (32.3%), facial twitching (23.5%), and facial paresthesia reported in (20.6%), otorrhea (11.8%), diplopia (5.9%), and facial weakness reported in (2.9%). Aggressive CG of petro us apex divided:

- First type: due to vestibulocochlear involvement sensor neural hearing loss and tinnitus are the commonest symptom, followed by vertigo and dizziness.
- Second type is related to cholesterol granuloma spread to the upper part of the petro us apex causing to middle and posterior fossa dural irritation, headache and facial pain are the commonest symptoms.
- Third type: due to the compression of trigeminal or the abdomens nerve, signifying compression of the Meckler's cave region. Rarely, recurrent otitis media can be a happen as a result of, the Eustachian tube compression.

Conductive hearing loss and a blue eardrum are the symptom of Cholesterol granuloma (CG) of the middle ear middle ear Cholesterol granuloma is less aggressive than petro's apex but it may erode into the middle ear, the mastoid bone and the petro us apex. However, aggressive spread into the middle fossa is extremely rare EAC Cholesterol granuloma: present as a mass that obstruct the external ear canal (EEC) is rare and it is difficult to be diagnosed.

Intramembranous tympanic membrane CG occurs rarely and it look like bluish mass in tympanic membrane. Mastoid cholesterol granulomas are often accompanied by middle ear lesions, contrasting petro us apex cholesterol granulomas, mastoid cholesterol granulomas tend to be benign and are usually nonaggressive and no progressive.

Audiometric and vestibular evaluation [25]

The audiogram could be normal or reveal conductive, sensorineural, or mixed hearing loss according to site of lesion.

Vestibular testing can show canal weakness from inner ear involvement. We usually obtain an audiogram but rarely an electronystagmogram.

Radiology [26]

Temporal bone CT reveals an expansile, sharply defined, and often rounded mass of the petro us apex with cortical thinning and trabecular breakdown. The general appearance is that of a slowly progressive benign process. There is central soft-tissue density without an internal matrix, a calcification, or residual deputations. If the lesion is sufficiently enlarged, frank bony dehiscence is observed.

On Temporal bone MRI, cholesterol granulomas are typically hyper intense on both T1 and T2 sequences because of the accumulation of blood breakdown products and proteinaceous debris (Figure 2). Small lesions may be relatively homogeneous, whereas large lesions show more heterogeneity. Often cholesterol granulomas have a distinct hypointense peripheral rim on T2-weighted images due to hemosiderin deposition. After contrast administration, there may be subtle peripheral enhancement secondary to inflammatory response but no central enhancement that would indicate solid tissue (Table 1).

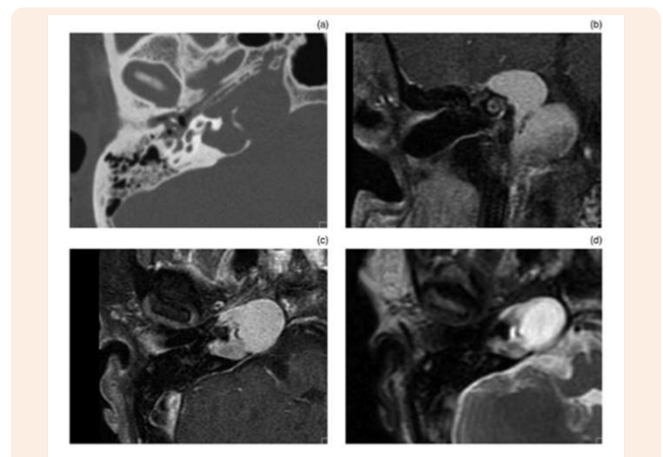


Figure 2: Temporal bone MRI.

Preoperative assessment [25]

Preoperative evaluation depends on of their symptoms. Patients complaining hearing loss are evaluated initially with audiometric assessment. Electronystagmography is reserved for patients who complain of imbalance or vertigo. Patients with asymmetric sensorineural hearing are next evaluated with auditory brainstem response testing. If results of ABR are abnormal, an MRI scan should be done, MRI also should be done In patients with cranial nerve involvement other than the eight nerve, Patients who have normal hearing abilities but also complain of cranial nerve deficits that may be preferable to the petro us apex may be screened with either a high-resolution, thin-section CT of the temporal bone or an MRI with gadolinium.

Treatment [27]

Management of cholesterol granuloma could be conservative or surgical depending on its size and the symptoms it produces.

Asymptomatic Small lesions are managed conservatively clinical and radiological follow up. An aggressive cholesterol granuloma require surgery that: (1) Drain or resects the disease, (2) preserve

hearing if it is possible and (3) preserve cranial nerve and carotid artery injury and CSF leak.

Table 1: Classification of petrous apex lesions

| Lesion | Computed Tomography | MRI T1 | MRI T2 | Enhancement |
|---------------------------|---|-------------------------------------|--------------|-------------|
| Mucus | Normal bone, nonenhancing | Hypointense | Hyperintense | No |
| Mucocele | Hypo dense, expansible smoothborder,nonenhancing | Hypointense | Hyperintense | No |
| Asymmetric Pneumatization | Normal bony architecture, nonenhancing | Hyperintense | Hypointense | No |
| Cholesteatoma | abnormal air cells, nonenhancing, isointense with CSF | Hypointense | Hyperintense | No |
| Cholesterol Granuloma | Expansile smooth border, rim enhancement, isointense with brain | Hyper intense | Hyperintense | No |
| Metastatic lesion | Destructive, unclear border | Isointense | Hyperintense | Yes |
| Chordoma | Aggressive bone destruction, calcification | Isointense: 75% Hypointense: 25% | Hyperintense | Yes |
| Chondroma | Aggressive bone destruction, calcification | Hypointense to isointense | Hyperintense | Yes |
| Chondrosarcoma | Aggressive bone destruction, calcification | Hypointense to isointense | Hyperintense | Yes |

Fenestration or classic tubes were the formerly used to drainage but it did not provide enough drainage because of the hindrance caused by pathophysiology and the content of the granuloma. Generally, the surgical route to reach the lesion is dependent on patient’s hearing acuity, tumor size, tumor location in petro us bone and the relationship to the jugular bulb. If the hearing ability is to be preserved, a middle fossa, infralabyrinthine, transsphenoidal, infracochlear approach can be used.

Middle cranial fossa approach is recommended for aggressive cholesterol granulomas spread towards the middle fossa, it is used to complete removal rather than simply drain the cyst, it is the procedure of choice when the location of cyst and poor hypo tympanic pneumatization make the infracochlear approach difficult, this approach provides good access to remove all the cyst in most cases except those cysts that spread inferiorly or those that encircle the carotid artery.

The infralabyrinthine and infracochlear approaches are used commonly in normal hearing patients. But the patients with high jugular bulb are not good candidate for the infralabyrinthine approach. The infracochlear-hypo tympanic approach is most common approach the petro us apex, it is a more conservative procedure to provide draining, decompression, and ventilation of the cyst, This procedure is safer and more simplest than the middle fossa approach because it avoids the injury to the GSPN, facial nerve, and Eustachian tube and preserve more the carotid artery and inner ear. Preoperatively, a CT scan should be obtained to show good pneumatization between the cyst wall and the hypo tympanum.

The transcochlear approach is the best approach to the petro us apex in deaf patients, it provides good exposure and control of the carotid artery for large lesions I. The transsphenoidal endoscopic surgery approach can be used for cysts reaching o

the sphenoid sinus and there is space between medial wall of cyst and lateral wall of pararegal ICA. It is contraindicated in patients where cysts are hidden behind ICA. The great disadvantage of this route is recurrent stenosis is occurring at its opening. Treatment for middle ear, mastoid, TM and EAC CG is Tympanmastoidectomy with appropriate reconstruction.

Outcomes [24]

- i. With open surgery outcomes: successful treatment achieved in 82-90 % of patient, symptoms improvement in 90%, recurrence in 12, 5%, complication rate 24.3 % (most common hearing loss).
- ii. Endoscopic outcomes: successful treatment seen in 98.6% of cases at follow-up (mean follow-up 20 months) Complications were seen in 13.2% of cases have complications, epistaxis is the most common complication. Restenosis occurred in (20.0%). The rate of recurrence was 10.7% in cases without using a stent compared with 4.3% in cases with stent using.

Follow up [26,28-30]

Based on clinical examination, including audiometric and vestibular evaluation and imaging. After surgical drainage, a CT scan can reveal air with the petro us apex; indicating good aeration of the cyst has been achieved. The cavity is sometimes refilled with serous fluid or scar tissue, which a CT scan not distinguishes from recurrent cholesterol granulomas. On MRI scans, the decrease in signal intensity on T1 weighed images and reduced size of the lesion confirms its successful evacuation. Moreover, technique of choice for the follow-up evaluation of cholesterol granuloma after surgical treatment.

- Radiologic characteristics of successful long term drainage:
- 1. Stable size-maximal diameter
 - 2. Presence of cyst aeration
 - 3.

Placement of drainage stent (Systemic recurrence can occur with re-accumulation within cysts w/o enlargement; therefore stable size may not be best). Follow up MRI of cholesterol granuloma shows hyper intense T1 that is suggest inadequate drainage in a symptomatic lesion. 52 the most common cause of CG recurrence is fibrous tissue obstruction at he drainage site.

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