Mini Review Article

The Auricular Wen-Cholesteatoma

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Cholesteatoma is a benign aggregate of keratinized squamous epithelial cells confined to the middle ear. Although designated as a cholesteatoma, the lesion is additionally denominated as keratoma or epidermal inclusion cyst of the middle ear wherein 'keratoma' may be considered as an apt terminology.

INTRODUCTION

The lesion is essentially configured of stratified squamous epithelium with admixed keratin flakes articulating a sac-like aggregation within the middle ear. The non neoplastic lesion may engender progressive erosion of bony ossicles and circumscribing bone. Aural polyp is composed of granulation tissue intermingled with keratin flakes configuring nodules which are generally associated with cholesteatoma.

Disease Characteristics

Cholesteatoma of the temporal bone and middle ear can be subdivided into:

Congenital Cholesteatoma: Accounting for around 2% instances congenital cholesteatoma is principally derived from remnants of stratified squamous epithelium which are entrapped within temporal bone during embryogenesis. Adjoining tympanic membranes are intact. Lesion is devoid of concurrent infection. Congenital variant is predominantly unilateral, generally obstructs the eustachian tube, initiates fluid accumulation within the middle ear and engenders conductive deafness(1,2).

Acquired Cholesteatoma: Comprises of nearly 98% lesions. Acquired cholesteatoma is devoid of accompanying embryologic phenomenon although appears due to pathological modifications which engender an uncontrolled growth of keratinized squamous epithelium within the middle ear. Acquired cholesteatoma is additionally classified into primary variant; wherein lesions are devoid of preceding chronic otomastoiditis. Primary acquired cholesteatoma is associated with retraction of tympanic membrane. secondary variant; which configures a majority of lesions and is associated with pars flaccida, pars tensa and conditions such as cholesteatoma of the external auditory canal, mural cholesteatoma or petrous apex cholesteatoma.

Secondary acquired cholesteatoma arises due to injury to tympanic membrane. Of undefined incidence, acquired cholesteatoma appears at an average age of 9.7 years. Generally, male subjects between 20 years to 30 years are incriminated. A male predominance is observed and acquired cholesteatoma demonstrates a male to female proportion of 1.4.

Cholesteatoma associated with pars flaccida originates within the Prussak space and exhibits posterior expansion. Cholesteatoma associated with pars tensa originates within the posterior mesotympanum and enunciates posteromedial expansion. Although benign, cholesteatoma can erode into the central nervous system with occurrence of severe complications. Cholesteatoma is devoid of disease- associated mortality.

Disease Pathogenesis

Of obscure pathogenesis, acquired middle ear cholesteatoma is posited to arise from inflammation within the middle ear with consequent proliferation of mucosal epithelial layer of the middle ear- a phenomenon designated as "squamous metaplasia" theory. Alternatively, squamous epithelium constituting extraneous layer of tympanic membrane is hypothesized to migrate through a perforation within the eardrum into the middle ear.

Else, basal cells of tympanic membrane proliferate and displace medially through the basement membrane in order to ingress the middle ear- a phenomenon denominated as "basal hyperplasia" theory. Additionally, primary, acquired cholesteatoma is surmised to arise on account of configuration of a retraction pocket- a phenomenon termed as "retraction pocket" theory.

Primary acquired cholesteatoma can be engendered due to retraction of tympanic membrane. Thus configured, cholesteatoma can injure middle ear ossicles and erode into aditus ad antrum or the mastoid. Persistent eustachian tube dysfunction may engender retraction pockets. Antecedent placement of tympanostomy tube eliminates negative pressure within the middle ear, decreases possible configuration of a retraction pocket and circumvents the articulation of a cholesteatoma. Infrequently, the facial nerve may be exposed.

Secondary, acquired cholesteatoma is posited to arise due to infection, trauma or surgical manipulation with consequent cutaneous implantation within the middle ear through a discontinuity of the eardrum. Nevertheless, genesis of cholesteatoma is hypothesized to comprehensively integrate and involve diverse categories of aforesaid mechanisms. Irrespective of the etiology, configuration of middle ear cholesteatoma is accompanied by perpetual proliferation, evolution and migration of the lesion with consequent damage to circumscribing anatomical structures of the middle ear.

Clinical Elucidation

Accumulation of stratified squamous epithelium within the middle ear may injure adjacent anatomical structures with consequent emergence of cogent clinical symptoms. The lesion may be asymptomatic or manifest conductive deafness, dizziness or otorrhea. Classic clinical representation of cholesteatoma is painless otorrhea. Deafness is common, permanent and severe, especially when associated with injury to or erosion of middle ear ossicles. Dizziness may ensue and pain may be debilitating. Cholesteatoma engenders an inflammatory response with activation of lytic enzymes, growth factors and cytokines which consequently recruit osteoclasts in order to initiate destruction of the bone. Infected cholesteatoma progresses rapidly

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and engenders significant bony erosion. The lesion is accompanied by a chronic, foul smelling ear discharge. Untreated cholesteatoma can erode bony margins of the middle ear with expansion into adjacent zones as the face, brain or neck. Destruction of localized bone and soft tissue is contingent to accompanying inflammation and release of proteolytic enzymes. However, incumbent pressure from the tumefaction appears to be ineffective and inadequate.

Cholesteatoma of external ear is disparate from middle ear cholesteatoma wherein the lesion may extend into the mastoid or middle ear. Also, facial nerve canal or tegmen tympani may be incriminated. The lesion is composed of compact, keratinous aggregates of squamous epithelial cells configuring a lamellar pattern. Cutaneous surface of external auditory canal demonstrates diffuse acanthosis and hyperkeratosis along with accumulation of subjacent, chronic inflammatory cell infiltrate composed of small lymphocytes and macrophages. However, osteonecrosis or focal decimation of superimposed stratified squamous epithelium is absent.

Histological Elucidation

Upon gross examination, greyish/white, cystic masses or nodules of variable magnitude are incorporated with waxy, off-white, granular substance. On morphological assessment, cholesteatoma appears as an epidermoid cyst. The nodular lesion is layered with desquamated, keratinizing stratified squamous epithelium. Intrinsic layering of the tumefaction is constituted of perpetually evolving stratified squamous epithelium with eventual emergence of cellular desquamation into the tumour mass.

Cholesterol content of the tumefaction may or may not be discernible. Essentially, cholesteatoma is layered by keratinized; non-dysplastic stratified squamous epithelium admixed with abundant granulation tissue and keratinous debris. The lesion is admixed with a chronic inflammatory infiltrate comprised of lymphocytes and histiocytes. Additionally, epithelioid cell granulomas, foreign body giant cell reaction, cholesterol clefts and foci of hemosiderin pigment deposition may be exemplified. Cholesteatoma arising within external ear is constituted of a nodule or mass layered with keratinized stratified squamous epithelium superimposed upon foci of bony sequestration confined within the external auditory canal.

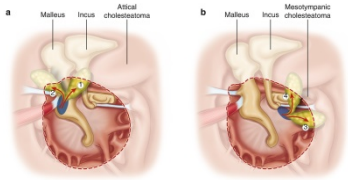

Figure1: Cholesteatoma demonstrating diverse location in relation to the middle ear ossicles

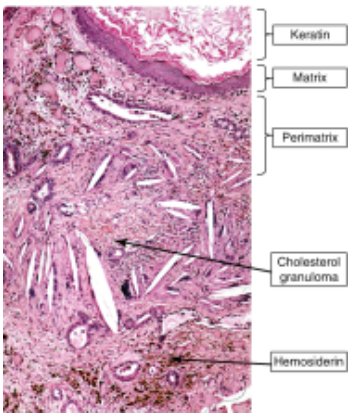

Figure 2: Cholesteatoma exhibiting diverse layers of keratin, intrinsic matrix and cholesterol clefts with hemosiderin pigment

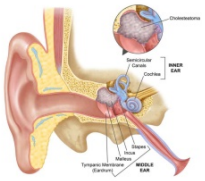

Figure 3: Cholesteatoma exemplifying the precise location adjoining middle ear ossicles and an intact tympanic membrane

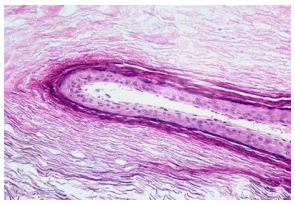

Figure 4: Cholesteatoma enunciating a stratified squamous epithelial cell lining along with keratin flakes, hemosiderin pigment and cholesterol clefts

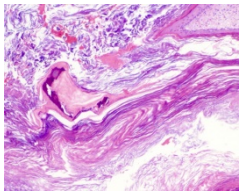

Figure 5: Cholesteatoma displaying a stratified squamous epithelial cell layer, cholesterol clefts, keratin flakes and inflammatory cell infiltrate

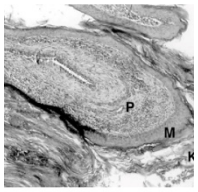

Figure 6: Cholesteatoma depicting convoluted strips of stratified squamous epithelium admixed with keratin flakes, cholesterol clefts and hemosiderin pigment

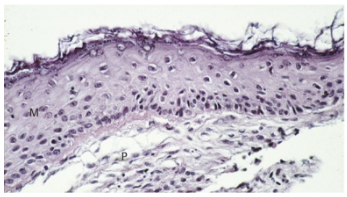

Figure 7: Cholesteatoma delineating strips of stratified squamous epithelium, keratin flakes and cholesterol clefts

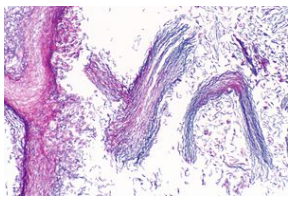

Figure 8: Cholesteatoma demonstrating strips of stratified squamous epithelium intermingled with cholesterol clefts and keratin flakes

DIFFERENTIAL DIAGNOSIS

Cholesteatoma requires segregation from conditions such as cholesterol granuloma, suppurative otitis media and inflammation or abscess of the middle ear, conditions which depict a diverse clinical representation and imaging features identical to a cholesteatoma. Also, distinction is required from a cerumen situated in the external auditory canal demonstrating

imaging characteristics simulating a cholesteatoma. Additionally, neoplasms such as middle ear osteoma, atypia or desmoplasia of superimposed stratified squamous epithelium and squamous cell carcinoma necessitate a demarcation.

Investigative Assay: Comprehensive, fundamental clinical history with details of present illness or repetitive ear infection, ear discharge and pertinent antibiotic therapy is required. Clinical manifestations such as subjective deafness, tinnitus, otorrhea, otalgia, ear pressure and vertigo require assessment. Comprehensive physical examination mandates the evaluation of head and neck with inspection of head, eyes, nose, oral cavity, oropharynx, neck and ears. Cholesteatoma can be adequately discerned upon naked eye examination. Middle ear cholesteatoma can be aptly examined with an otoscope. Microscopic evaluation of the middle ear depicts aggregates of greyish/white, keratinaceous debris confined to the postero-superior quadrant of tympanic membrane. Preoperative and postoperative audiograms are necessitated. Audiometry prior to pertinent surgical intervention is mandated in order to establish baseline levels. Parameters such as speech reception threshold, speech discrimination and air or bone conduction scores require assessment.

Upon cogent imaging, cholesteatoma demonstrates subtle defects such as labyrinthine fistula, scutal erosion, ossicular erosion or injury and defects within the tegmen. Computerized tomography (CT) is preferred for evaluating possible occurrence of cholesteatoma and precise bony configuration of the lesion can be obtained for cogent preoperative assessment, reconstruction of ear ossicles and excluding perforation of bony tegmen. Cholesteatoma appearing within sinus tympani of the middle ear requires assessment in order to circumvent residual disease. Computerized tomography (CT) along with imaging of thin slices of temporal bone in the absence of administration of contrast medium is an optimal modality for evaluating cholesteatoma.

Precise imaging is pertinent for appropriate planning of preferable surgical techniques. Upon computerized tomography, manifestations of a cholesteatoma which require evaluation are erosion of the scutum, ear ossicles or lateral semicircular canal, dehiscence of facial nerve canal or tegmen tympani and integrity of epitympanum, aditus ad antrum and mastoid antrum along with oval and round window. However, the manoeuver is nonspecific for ascertaining repetitive or residual cholesteatomas. Conventional, non-contrast magnetic resonance imaging (MRI) and diffusion-weighted magnetic resonance imaging (MRI) is recommended for ascertaining cholesteatoma. MRI is a precise technique for discerning reoccurring or residual lesions and may be optimally adopted in individuals subjected to previous surgery for cholesteatoma. Absence of tumefaction upon MRI may obviate a "second look" surgery. Ideally, magnetic resonance imaging necessitates appropriate preparation with an unobstructed external auditory canal or a postoperative cavity in order to circumvent false-positive diagnoses.

With MRI, acquired middle ear cholesteatoma can appear isointense upon T1 weighted imaging and hyper-intense upon T2 weighted imaging. Image enhancement with administration of gadolinium contrast is absent. MRI is an optimal technique adopted for imaging surveillance of cholesteatoma following surgery. Miniature, reappearing cholesteatomas of few millimetres magnitude can be discerned with almost 100% specificity.

Therapeutic Options

Aim of therapy is a safe, dry, infection- free ear. Comprehensive surgical excision of the neoplastic components is mandated. Frequently, surgical extermination is performed in combination with procedures such as tympanoplasty, mastoidectomy or ossicular reconstruction, contingent to extent of disease. Surgical extermination of cholesteatoma is a definitive treatment strategy. However, surgical intervention may not restore the hearing to normal and hearing may decline following surgery. Surgical procedure adopted is contingent to category and location of the cholesteatoma. Nevertheless, tympanomastoidectomy is a frequently performed procedure which ensures eradication of a cholesteatoma in its entirety.

An estimated 5% to 50% of surgical procedures are followed by reoccurrence of the lesion. Inadequate surgical resection may initiate widespread destruction of adjoining bone. Treatment of infected cholesteatoma may be challenging. As the lesion is devoid of vascular perfusion, systemic antibiotics are unable to ingress the tumefaction. Topical antibiotics are appropriate for controlling superficial infection, especially in instances where an ear discharge is persistent or recurrent. Although multiple surgical procedures are necessitated, cholesteatoma can be completely eradicated. Repetitive tympanomastoidectomy is required in an estimated 5% individuals. Also, deafness may be permanent. Cholesteatoma associated with papillary epithelial hyperplasia and significant koilocytosis is indicative of aggressive disease and possible infection with human papilloma virus (HPV).Complications arising from cholesteatoma are exceptional and are comprised of sigmoid sinus thrombosis, conductive hearing loss, meningitis and epidural abscess. A preliminary, close monitoring is crucial as delayed intervention is associated with inferior outcomes.

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