A PROSPECTIVE STUDY OF CHOLESTEATOMA IN PAEDIATRIC AGE GROUP AND ITS MANAGEMENT

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ABSTRACT

INTRODUCTION

A cholesteatoma (sometimes called a keratoma) is an abnormal growth of squamous epithelium in the middle ear and mastoid.¹ It may progressively enlarge to surround and destroy the ossicles, resulting in conductive hearing loss. Hearing loss also may occur if the cholesteatoma obstructs the eustachian tube orifice, leading to middle ear effusion. Surgical therapy is required for most cholesteatomas. The extent and effectiveness of surgery depends upon the size of the cholesteatoma. Early diagnosis is crucial to an optimal outcome

In children this disease is more aggressive and complex and leads to many complications and the treatment is mostly surgical.² This study is chosen because the incidence of recurrence and residual disease is higher in children and its management is very challenging.

OBJECTIVE

To study the clinical features, disease process. spread and management of cholesteatoma in paediatric age group (less than 16 years).

MATERIALS AND METHODS

A prospective study was conducted at Govt. ENT Hospital over a period of two years that included 50 cases of cholesteatoma in paediatric age group.

RESULTS

Surgical treatment by Mastoidectomy was done in all cases. Out of 50 cases after surgery 33 patients had dry ear and improvement in hearing 7 patients had recurrence of disease 8 patients had cavity problems like discharge from the ear and 2 patients developed neurological complications.

CONCLUSION

Childhood cholesteatoma has an aggressive nature with tendency for complications hence its treatment is very challenging. Eradication of disease and restoration of hearing should be the primary goal of treatment. Long term follow up is important to monitor the disease recurrence.

KEYWORDS

Cholesteatoma, Mastoidectomy, Otorrhoea, Deafness.

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INTRODUCTION: Cholesteatoma is a destructive and expanding growth consisting of keratinizing squamous epithelium in the middle ear and/or mastoid process.³

Submission 30-12-2015, Peer Review 16-01-2016, Acceptance 29-01-2016, Published 08-02-2016. Corresponding Author: Dr. K. Nagaraj, H. No. 11-5-127, Flat No. 2B, Sai Sudha Apts, Red Hills, Nampally, Hyderabad. E-mail: drnagarajkarna@gmail.com DOI: 10.18410/jebmh/2016/77 Although cholesteatomas are not classified as either tumours or cancers, they can still cause significant problems because of their erosive and expansile properties resulting in the destruction of the bones of the middle ear (ossicles), as well as their possible spread through the base of the skull into the brain. They are also often infected and can result in chronically draining ears.

The majority (98%) of patients with cholesteatoma have ear discharge or hearing loss or both in the affected ear.

Cholesteatoma is a persistent disease. Once the diagnosis of cholesteatoma is made in a patient who can

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tolerate a general anesthetic, the standard treatment is to surgically remove the growth.⁴

The challenge of cholesteatoma surgery is to permanently remove the cholesteatoma whilst retaining or reconstructing the normal functions of the structures housed within the temporal bone.

Surgical techniques: The primary goal of the surgical treatment was the eradication of disease followed by restoration of hearing. Among the numerous surgical techniques described, the two techniques which are commonly performed are the canal wall down mastoidectomy (CWD) and the other canal wall up (CWU) mastoidectomy.^{5,6}

CWD is done preferably to obtain an adequate anatomical overview and safe eradication while CWU technique preserves anatomical structures and hearing restoration is better. The incidence of recurrence of disease is more common in CWU mastoidectomy. The CWD technique involves removal of posterior canal wall which provides better visualization of the epitympanum and mesotympanum hence better clearance of the disease.

MATERIALS AND METHODS: A Total 50 paediatric cases of cholesteatoma were studied after management for a period of six months.

Patients were selected after taking thorough history, clinical examination, otosopic examination, investigations like Audiometry and CT Scan of temporal bones.⁷ once the diagnosis of cholesteatoma was made the patients were subjected to surgical treatment. Mainly two methods of surgery was done. One is canal wall down Mastoidectomy and the other one was canal wall up mastoidectomy.

RESULTS AND OBSERVATIONS: This paper presents an analysis of 50 paediatric cholesteatoma cases which underwent treatment over a period of two years.

Out of 50 patients the incidence of cholesteatoma was highest in the age group of 13-16 years while second most common age group found was between 9-12 years. Males were more commonly involved than females. Left ear was more involved. Most of the patients presented with only ear discharge 21 pts whereas 10 pts presented with discharge and deafness and 5 pts presented with come complications. On Otoscopy 23 pts had attic perforation while 11 pts presented with aural polyp, 8 patients had postero superior perforation in the tympanic membrane. Incus was the most common ossicle eroded and all ossicles were eroded in 10 cases. 8 pts had erosion of mastoid cortex, tegmen plate was eroded in 2 patients, Facial canal dehiscence was seen in 4 pts, posterior canal wall dehiscence was noted in 2 patients. In most of the cases the type of hearing loss was conductive (80%) mixed hearing loss was seen in 16% and sensory neural hearing loss in 4%.

Based on the extension of cholesteatoma 35 patients underwent CWD technique of mastoidectomy, 8 patients CWU mastoidectomy 5 patients underwent inside out technique. **Follow up**: after 6 months of follow up 33 patients (66%) had a dry ear with improvement in hearing, 7 patients (14%) had recurrence of disease, discharging cavities were noted in 8 patients (16%) and 2 patients (4%) had sensory neural hearing loss.



Fig. 1: Bar chart showing age distribution



Fig. 2: Bar chart showing sex distribution



Fig. 3: Pie chart showing sex distribution



Fig. 4: Bar diagram showing involved side and its percentage

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Fig. 5: Bar diagram showing the clinical presentation of patients



Fig. 6: Bar diagram showing the otoscopic findings



Fig. 7: Showing the involvement of ossicles



Fig. 8: Bar diagram showing the erosion of bone due to disease



Fig. 9: Bar diagram showing the type of hearing loss



Fig. 10: Pie diagram showing the degree of involvement



Fig. 11: Bar diagram showing different operative procedures done in paediatric cholesteatoma cases



Fig. 12: Bar diagram showing the result of follow up

DISCUSSION: A cholesteatoma consists of squamous epithelium that is trapped within the skull base and that can erode and destroy important structures within the temporal bone. Its potential for causing central nervous system (CNS) complications (e.g., brain abscess, meningitis) makes it a potentially fatal lesion.

Generally, the following 3 types of cholesteatoma are identified:

- Congenital cholesteatoma.⁸
- Primary acquired cholesteatoma.
- Secondary acquired cholesteatoma.

Signs and Symptoms: The hallmark symptom of a cholesteatoma is a painless otorrhoea, either unremitting or frequently recurrent. Other symptoms include the following:

- Conductive hearing loss.
- Dizziness: Relatively uncommon.
- Drainage and granulation tissue in the ear canal and middle ear: Unresponsive to antimicrobial therapy.

Occasionally, cholesteatoma initially presents with symptoms of CNS complications, including the following:

- Sigmoid sinus thrombosis.
- Epidural abscess.
- Meningitis.

Unlike other cholesteatomas, the congenital type is usually identified behind an intact, normal-appearing tympanic membrane. The child often has no history of recurrent suppurative ear disease, previous otologic surgery, or tympanic membrane perforation.

Diagnosis: No laboratory tests or incisional biopsies are generally necessary for the diagnosis of cholesteatomas, because the diagnosis can be made based on physical examination and radiologic findings.

Computed tomography (CT) scanning is the diagnostic imaging modality of choice for these lesions, owing to its ability to detect subtle bony defects.⁷

Histologically, surgically removed cholesteatoma specimens demonstrate typical squamous epithelium. The histology is indistinguishable from that of sebaceous cysts or keratomas removed from any other portion of the body.

Audiometry should be performed prior to surgery whenever possible. Air and bone conduction, the speech reception threshold, and speech discrimination scores should all be determined within a few weeks of the proposed operative procedure.

Magnetic resonance imaging (MRI) is used when very specific problems, such as the following, are suspected.

- Dural involvement or invasion.
- Subdural or epidural abscess.
- Brain herniation into the mastoid cavity.
- Inflammation of the membranous labyrinth or facial nerve.
- Sigmoid sinus thrombosis.
- Meningitis.

Management: Virtually all cholesteatomas should be excised. The only absolute contraindications to the surgical removal of cholesteatomas are medical in nature.

Canal wall-down mastoidectomy: In the canal walldown (open) procedure, the posterior canal wall is removed. A large meatoplasty is created to allow adequate air circulation into the cavity that arises from the operation. Canal wall–down operations have the highest probability of permanently ridding patients of cholesteatomas.

Canal wall-up mastoidectomy⁹: In the canal wall–up (closed) procedure, q the canal wall is preserved. Canal wall–up procedures have the advantage of maintaining a normal appearance, but the risk of persistent or recurrent cholesteatomas is higher than in the canal wall-down operation.

CONCLUSION: Cholesteatoma in children¹⁰ is an aggressive disease hence eradication of the disease by surgery was the primary goal followed by restoration of hearing. Lower and middle socio economic group was affected commonly. CT scan temporal bone was diagnostic in demonstrating the extent of disease and also any intracranial extension with bony erosion. Higher recurrences were noted with CWU mastoidectomy hence long term follow up is mandatory to monitor for disease recurrence.¹¹

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