ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 11, Issue, 11, pp.8189-8192, November, 2019

DOI: https://doi.org/10.24941/ijcr.37176.11.2019

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

REVIEW ARTICLE

PRIMARY EXTERNAL AUDITORY CANAL CHOLESTEATOMA – OUR EXPERIENCE

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ARTICLE INFO

Article History:
Received 14th August, 2019
Received in revised form
08th September, 2019
Accepted 25th October, 2019
Published online 26th November, 2019

Key Words:

Cholestaatoma,
External auditory canal.

ABSTRACT

Introduction: External auditory canal cholesteatoma is a rare condition. We are reporting a rare case of primary auditory canal cholesteatoma causing facial canal erosion with mastoid tip involvement and jugular vein thrombophlebitis. **Case Report:** We are presenting two cases 75years and 50 years female who presented to us with unilateral otorrhoea with hearing loss. Otomicroscopy revealed external canal wall cholesteatoma with intact Tympanic membrane. High resolution CT scan confirmed the findings of otomicroscopy and also revealed further extensive disease and erosion of fallopian canal and bone over jugular bulb. Both the patients were posted for mastoidectomy surgery & cholesteatoma was removed in toto without any complications. **Conclusion:** The rare occurrence of external auditory canal cholesteatoma makes it difficult to establish its incidence rates. However its possibility should be kept in mind while treating patients with Otorroea and presenting as otitis externa cases. High resolution CT scan is an important diagnostic tool for external canal wall choesteatoma. Prompt surgical intervention is the key to successful outcome with less morbidity and mortality in extensive external canal wall cholesteatoma.

Citation: Dr. Nilam U. Sathe, Dr. Shampa Mishra, Dr. Swapnal Sawarkar and Dr. Swapna Patil. **2019.** "Primary External Auditory Canal Cholesteatoma – Our experience", *International Journal of Current Research*, 11, (11), 8189-8192.

INTRODUCTION

External auditory canal cholesteatoma is a rare condition with an estimated incidence of one in thousand cases (Anthony, 1982). It causes ulceration of canal skin and necrosis of underlying bone. Early reports of this disease have been made by Toynbee (Toynbee, 1850) and Scholefield (Scholefield, 1893) in 1850. It has propensity to invade adjacent structures like mastoid, middle ear, temporomandibular joint, facial canal, labyrinth, sigmoid sinus etc (Smith, 1978; Sismanis, 1986; Garin, 1997; Martin, 1999). Due to its rare occurrence, many of previous studies report few cases. We are reporting a rare case of primary auditory canal cholesteatoma causing facial canal erosion with mastoid tip involvement and jugular vein thrombophlebitis.

CASE REPORT

Case 1: A 75 years old lady, non diabetic non hypertensive, presented to our hospital with left otalgia, scanty mucopurulent ear discharge and moderate conductive hearing loss from two months. There was no history of facial palsy, recent trauma, recent ear syringing, or previous surgery. She did not have any complaints in right ear.

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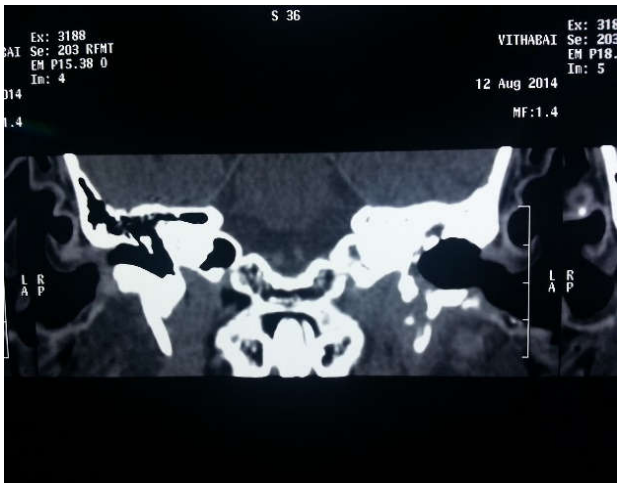
Examination under microscope revealed granulations in inferior canal wall lateral to annulus with posterior canal wall erosion. There was no polyp or cholesteatoma seen. Tympanic membrane was intact but dull in appearance. Her right ear was normal. High resolution computed tomography scan showed soft tissue density involving left external auditory canal, middle ear and mastoid air cells with erosion of posteroinferior canal wall including jugular bulb, facial canal, mastoid part of left temporomandibular joint and left styloid process (Figure 1, 2). A peripherally enhancing collection was seen medial to tip of left mastoid process with non opacification of internal jugular vein. We decided to explore her left ear and she was posted for Mastoidectomy surgery. It revealed mastoid tip cholesteatoma with its erosion. Facial nerve was dehiscent in its vertical segment. Middle ear and ossicles were found intact. She was subsequently discharged on oral antibiotics. Patient was followed for 6 months and recovered without complications.

Case 2: A 5o years old lady, non diabetic, hypertensive, presented to our hospital with left otorrhoea since 3 months which was scanty, fowl smelling, purulent ear discharge and moderate conductive hearing loss from two months. There was no history of facial palsy, recent trauma, recent ear syringing, or previous surgery. She did not have any complaints in right ear.



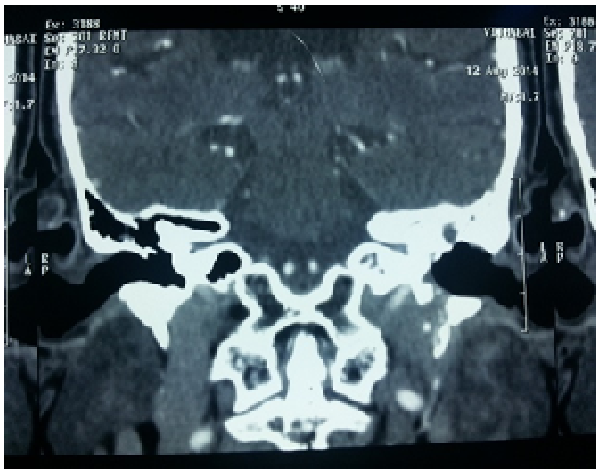


Figure 1 and 2. High resolution computed tomography scan showing soft tissue density involving left external auditory canal, middle ear and mastoid air cells with erosion of posteroinferior canal wall including jugular bulb, facial canal, mastoid part of left temporomandibular joint and left styloid

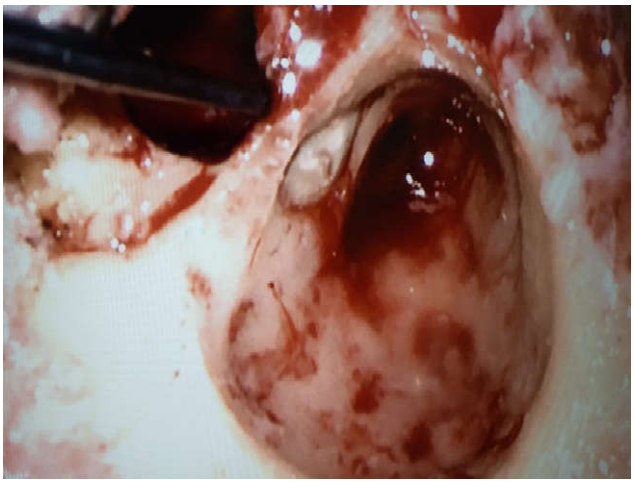
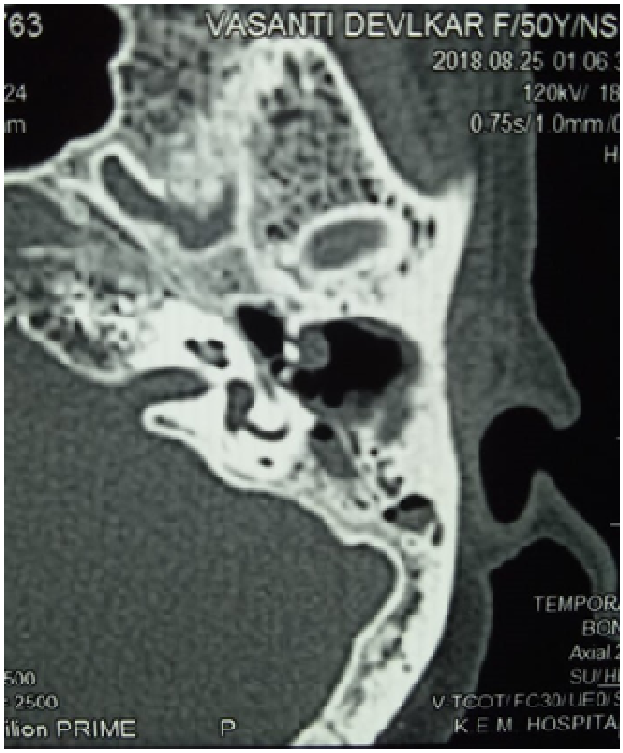


Figure 3. Showing cholesteatoma and granulations in inferior canal wall lateral to annulus with erosion of posterior canal wall and floor of external auditory canal.

Examination under microscope revealed cholesteatoma and granulations in inferior canal wall lateral to annulus with erosion of posterior canal wall and floor of external auditory canal (Figure 3). There was no polyp or cholesteatoma seen. Tympanic membrane was intact but dull and thickened in appearance. Her right ear was normal.



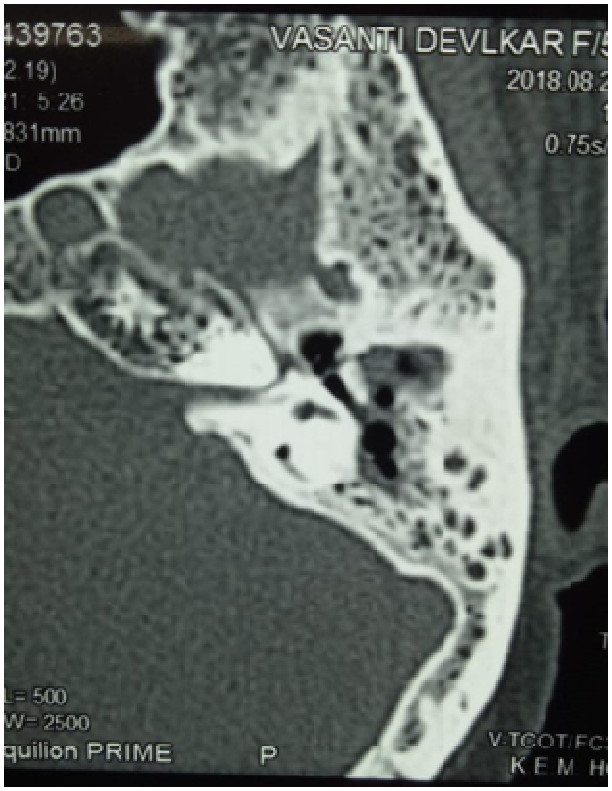


Figure 4 a & b. High resolution computed tomography scan showed soft tissue density involving left external auditory canal, middle ear and mastoid air cells with erosion of poster inferior canal wall , facial canal

Pure tone audiogram was suggestive of moderate conductive hearing loss in left ear and normal hearing in the right ear. High resolution computed tomography scan showed soft tissue density involving left external auditory canal, middle ear and mastoid air cells with erosion of poster inferior canal wall, facial canal, and (Figure 4 a & b). Patient was posted for surgery. Canal wall down mastoidectomy was done.

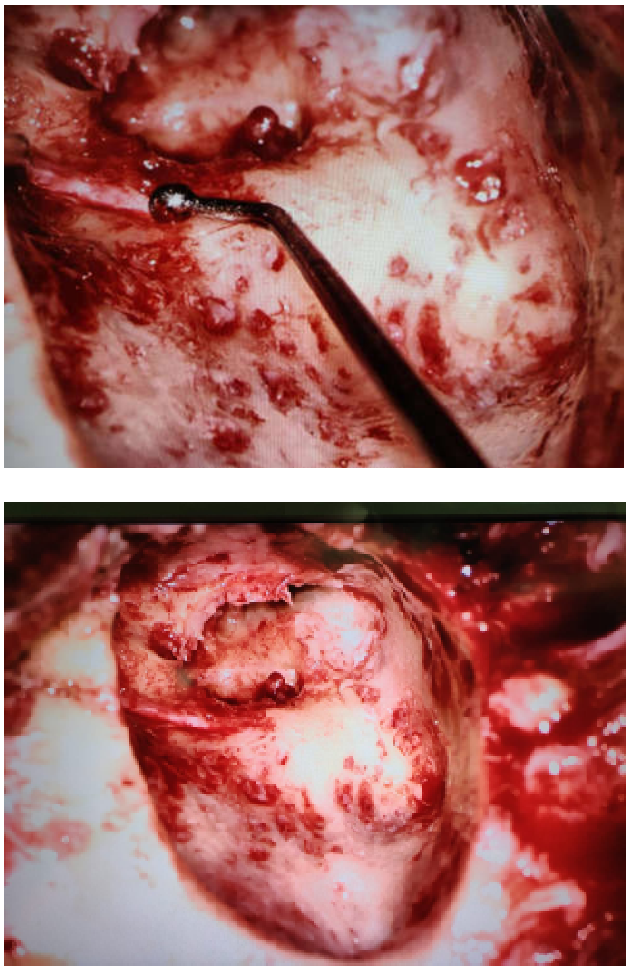


Figure 5 a& b. Showing Cholesteatoma extending from external auditory canal eroding posterior wall of external auditory canal and it was draping Vertical part of facial nerve with complete erosion of fallopian canal

Cholesteatoma was found extending from external auditory canal it had eroded posterior wall of external auditory canal and it was draping Vertical part of facial nerve with complete erosion of fallopian canal Figure 5 a& b). Partial necrosis of head of malleus and linticular process of incus was seen. Chorda Tympani nerve was also inserted abnormally to the vertical segment of facial nerve towards the tip of vertical segment. Entire cholesteatoma was removed by canal wall down mastoidectomy and facial nerve was covered with double layer of Temporalis fascia graft. Type III tympanoplsty done and Temporal is fascia graft graft placed over stapes head to preserve the hearing. On follow up the patient has recovered without any complication.

DISCUSSION

The rare occurrence of external auditory canal cholesteatoma makes it difficult to establish its incidence rates. From Anthony and Anthony incidence of 1.2 primary cases per 1000 new ontological patients can be estimated.[1]It is defined as invasion of squamous tissue from the canal into a localized area of bony erosion. It was historically confused with keratosis obturans which is characterized by abnormal epithelial migration leading to accumulation of keratin in the medial portion of ear canal (Naiberg, 1984; Persaud, 2004). Although the two terms were long used interchangeably, they are now generally held to separate disease entities (Naiberg, 1984; Persaud, 2004). External auditory canal cholesteatoma has been generally classified into primary and secondary by Tos (Tos, 1997). Primary cases have an unknown etiology whereas secondary cases are generally postsurgery, posttraumatic or postradiotherapy (Brookes, 1984; Venkatraman, 1997).

It causes skin ulceration and osteoradionecrosis at the inferoposterior bony ear canal lateral to annulus. It can invade mastoid, middle ear, facial nerve, dura, sigmoid sinus etc (Smith, 1978; Sismanis, 1986; Garin, 1997; Martin, 1999). Patients with external auditory canal cholesteatoma generally present with features of external otitis including otorrhoea and pain (Heilbrun, 2003). Generally there is no hearing deficit. But many cases can be remarkably silent or even asymptomatic. On examination, granulation tissue may be seen at the edge of a bony crater filled with keratin debris (Heilbrun, 2003). Generally it is treated with cleaning, removal of debris and medications. Surgery involves removal of cholesteatoma matrix and necrotic bone followed by grafting to promote healthy reepithelization (Farrior, 1990; Vrabec, 2002).

Conclusion

- The rare occurrence of external auditory canal cholesteatoma makes it difficult to establish its incidence rates. However its possibility should be kept in mind while treating patients with Otorroea and presenting as otitis extrna cases.
- High resolution CT scan is an important diagnostic tool for external canal wall choesteatoma.
- Prompt surgical intervention is the key to successful outcome with less morbidity and mortality in extensive external canal wall cholesteatoma.

Financial Support and Sponsorships: Nil

No conflict of interest

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