Clinics in Surgery

Case Report
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A Recurrent Cholesteatoma Presenting as a Pinna Lesion

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Abstract

Background: Cholesteatoma is a recognized destructive benign lesion that has the potential to cause serious complications when surgical intervention is delayed.

Case Report: A 23 year old female with a background of right-sided mastoid surgery for cholesteatoma, attended ENT clinic with a lesion of the right pinna. Attempted aspiration of the assumed pinna abscess was unsuccessful.

Investigations: A CT scan of the temporal bones showed extensive residual soft tissue in the right external ear, middle ear and mastoid; features suggestive of recurrent cholesteatoma.

Management: The patient was listed for urgent mastoid exploration; dissection and removal of the recurring cholesteatoma was carried out. Intra-operative findings included a sinus tracking from the cholesteatoma site in middle ear to the pinna.

Discussion: External ear cholesteatomas are a rare entity and often manifest with minimal symptoms. Referring clinicians should be aware of atypically-presenting cholesteatomas, which can lead to a delay in diagnosis, worse prognosis and increased incidence of complications. Present with atypical or even an absence of symptoms of a recurrent cholesteatoma. Often, the referring clinician is unaware of nonconforming presentations of the disease which can invariably lead to diagnostic delay, a poorer prognosis and an increased incidence of adverse complications.

Keywords: Otolaryngology; Cholesteatoma; Mastoidectomy; Education

Introduction

A cholesteatoma is formed from the proliferation of keratinized stratified squamous epithelium, most often into the middle ear and mastoid cavity. A number of speculations regarding the pathogenesis of cholesteatomas have been postulated; however the mechanism of disease largely remains unclear. The most widely recognized pathophysiological basis of acquired cholesteatoma comprises that of defective wound healing processes. This theory holds that negative pressure due to dysfunction of the Eustachian tube leads to the formation of a retraction pocket, the obstruction of which can be difficult to clear due to trapped desquamated keratin and thus, a cholesteatoma results. Superimposed infection causes disturbance to self-cleaning mechanisms and further debris accumulates in the area of retraction. Immune complex deposition as a result of inflammation causes the release of inflammatory mediators and cytokines which ultimately lead to erosion of surrounding structures as the disease progresses [1]. Some scholars postulate that a preneoplastic or neoplastic process predominates in the formation of a cholesteatoma [2]. Defective host inflammatory responses and bacterial infection have also been implicated in the development of cholesteatomas [3,4]. Cholesteatomas that present extradurally most often involve the middle ear, yet can manifest in any part of the petrous temporal bone including the mastoid, petrous apex and external auditory canal. The expansion and growth of this destructive lesion has the potential to degrade into important neighboring anatomical structures. As cholesteatomas commonly form within the middle ear cleft, they occur in very close proximity to vital middle ear structures such as the ossicular chain as well as the facial and vestibulocochlear nerves. Should the lesion extend beyond the bony matrix of the temporal bone and breach intracranial structures, fatal complications such a meningitis and brain abscesses can result. Violation of the components of the internal auditory meatus can cause symptoms of hearing loss, vestibular dysfunction and paralysis of facial muscles. The most common presentation of acquired cholesteatoma is that of a protracted history of malodorous otorrhea, otalgia and hearing loss. The only effective management for a diagnosed cholesteatoma is surgical intervention. It is therefore imperative that practitioners are able to recognize the symptoms of cholesteatoma and importance of prompt referral to an ENT surgeon to allow timely organization for definitive surgical treatment, and thus avoidance of the

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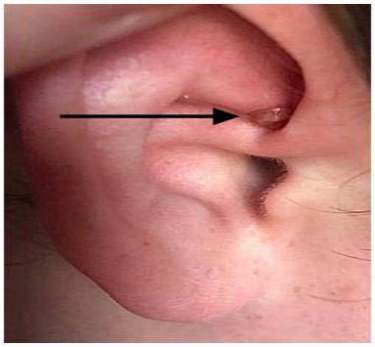

Figure 1: Image of presumed pinna "abscess" on initial presentation.

serious complications. Patients with previous mastoid surgery and by definition, altered anatomy, are more likely to present with atypical or even an absence of symptoms of a recurrent cholesteatoma. Often, the referring clinician is unaware of nonconforming presentations of the disease which can invariably lead to diagnostic delay, a poorer prognosis and an increased incidence of adverse complications.

Case Presentation

A 23 year old female attended the Emergency department on advice from her general practitioner with a right-sided pinna swelling and otalgia. The lesion over the right pinna had been observed to discharge yellow pus. The patient denied complaints of hearing loss, tinnitus, vertigo, malodorous otorrhea or a sensation of "fullness" in the affected ear. Otherwise, the patient reported to be systemically well, with no fevers, coughs or respiratory symptoms. Past medical history consisted of polycystic ovarian syndrome, gastroesophageal reflux disease and mixed anxiety and depressive disorder. Past surgical history comprised a previous right mastoid exploration and cartilage tympanoplasty 4 years previously for cholesteatoma. Following a disease-free Magnetic Resonance Imaging (MRI) scan 3 years after the surgery, the patient was discharged from ENT services. Medications included omeprazole and antidepressant tablets. The patient reported an intolerance to both erythromycin and amoxicillin which caused a rash and difficulty breathing. The patient lived at home with her mother and was independent of activities of daily living. Examination revealed a lesion over the right pinna with discharging pus (Figure 1). Ear examination was unremarkable, with normal appearances of the external ear canal and tympanic membranes bilaterally. Facial nerve function was preserved and no cranial nerve palsies were identified. There was no lymphadenopathy identified and neck movements were normal; examination of the oropharynx was unremarkable.

Attempts to drain the assumed pinna abscess under local anesthetic were unsuccessful with minimal pus aspirated and the wound was subsequently dressed. The patient was commenced on a 7 day course of oral antibiotics and a Computed Tomography (CT) scan of the temporal bones was arranged to exclude a possible cholesteatoma recurrence.

Investigations/differential diagnosis

Blood tests revealed urea, electrolytes and creatinine all within normal ranges. There was a slight increase in inflammatory markers with a C-reactive protein of 24.9 mg/L, white cell count of 11.2 × 10^9/L and platelets of 456 × 10^9/L. Hemoglobin was measured at 129 g/L. A CT scan of the temporal bones showed extensive residual soft tissue in the right external ear, middle ear and mastoid. This was

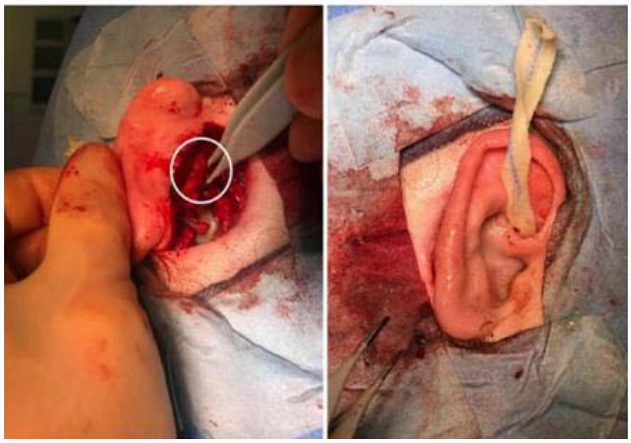

Figure 2: Intra-operative images of external ear site of fistulating tract from middle ear to pinna.

suggestive of recurrent cholesteatoma. It is important to consider a localized infection in a young patient presenting with unilateral ear pain and features of inflammation of the external ear. An important differential diagnosis considered in this particular case included a pinna abscess due to the characteristics of the lesion and overlying skin. The diagnosis of pinna hematoma was reliably excluded due to a lack of trauma and the location of the ear swelling. It was prudent to further investigate the patient's symptoms with imaging as recurrence of cholesteatoma in a patient that had undergone previous mastoid exploration ought to be considered in such cases. Altered anatomy due to previous surgery further lowers thresholds for recurring disease to present in an atypical fashion [5].

Management/follow-up

Following review of the imaging results, the patient was listed for an urgent canal wall down mastoid exploration. The surgical procedure was carried out 4 weeks following initial presentation to clinic. Intra-operative findings included a sinus tracking from the cholesteatoma site in middle ear to the pinna (Figure 2). The cholesteatoma was successfully dissected from the attic of the middle ear, a bone graft carried out and temporalis fascia used to reconstruct the tympanic membrane which was positioned over the posterior bony pocket. The patient was closely monitored on the surgical high dependency unit following the operation and was discharged home the same day. The patient was followed up 4 weeks following surgery in an outpatient setting. On examination the post-auricular wound had healed well with no overt signs of erythema, edema or tenderness on palpation. The mastoid cavity was free of inflammation and there was no significant debris observed within. The patient still continues to be followed up by the ENT specialists for monitoring and micro-suction of the mastoid cavity.

Discussion

Cholesteatoma is a recognized destructive benign lesion that has the potential to cause fatal complications with diagnostic delay. It is increasingly important for treating practitioners to be able to appreciate unusual expressions of cholesteatoma, as there have been a number of atypical presentations reported in the literature. External ear cholesteatomas are a rare entity with an estimated incidence of 1.2 per 1,000 [6]. Symptoms of otalgia and otorrhea have been reported, however there it is surprising that many patients display minimal or no symptoms [7,8]. External ear cholesteatomas have further been

associated with recurrence as well as bilateral disease [9]. Previously published case reports highlight the detriments of delayed detection of the disease as well as the paradoxical findings of extensive disease in a largely asymptomatic patient [10]. These accounts highlight the insidious nature of atypically-occurring cholesteatomas, and the extent to which the resulting damage incurred by the disease can be obscured by little or no symptoms. These factors highlight the importance of an increased degree of vigilance when examining patients with external ear lesions and few signs and symptoms consistent with that expected of a typical cholesteatoma. Further unusual presentations of this phenomenon have been documented, with reports of cholesteatomas presenting as a neck swellings [11], as well as extensive disease with intracranial involvement presenting as a head lump in an asymptomatic patient [12]. These cases highlight the potential sporadic element of such lesions. This becomes more relevant in patients that have undergone previous mastoid surgery and thus increased recurrence rates for cholesteatomas [13]. One such study reported a recurrence rate of 5% following mastoid surgery [14]. Undoubtedly and predictably, these altered presentations have lead to delays in definitive treatment as a result of deferred identification, owing to an uncharacteristic presentation of the disease. With a profile of potentially fatal complications, it is in patients' best interests that primary care doctors and referring clinicians are aware of unconventional expressions of this phenomenon.

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Bullet Point Summary

- Atypically-occurring cholesteatomas may occur as an insidious entity, concealing extensive damage caused by the disease due to a lack of physical symptoms.
- Altered presentations of cholesteatoma have lead to delays in diagnoses and therefore definitive treatment, causing increased complications rates as a result.
- It is imperative that referring practitioners are able to recognize typical and atypical symptoms of cholesteatoma and importance of prompt referral to an ENT surgeon to allow timely organization for definitive surgical treatment.
- Patients with previous mastoid surgery have been found to have 5% recurrence rate of cholesteatoma; a proportion of which have displayed atypical features of the disease.

References

1. Maniu A, Harabagiu O, Perde Schrepler M, Cătană A, Fănuță B, Mogoantă CA. Molecular biology of cholesteatoma. Rom J Morphol Embryol. 2014;55:7-13.
2. Albino AP, Reed JA, Bogdany JK, Sassoon J, Desloge RB, Parisier SC. Expression of p53 protein in human middle ear cholesteatomas: Pathogenetic implications. Am J Otol. 1998;19(1):30-6.
3. Wendt H. Desquamative Entzündung des Mittelohres ("Cholesteatom des Felsenbeins"). Arch Ohr Nasen Kehlkopfheilk. 1873;14:428-46.
4. Sadé J. Cellular differentiation of the middle ear lining. Ann Otol Rhinol Laryngol. 1971;80(3):376-83.
5. Venkatraman G, Mattox DE. External auditory canal wall cholesteatoma: A complication of ear surgery. Acta Otolaryngol. 1997;117(2):293-7.
6. Anthony PF, Anthony WP. Surgical treatment of external auditory canal cholesteatoma. Laryngoscope. 1982;92(1):70-5.
7. Holt JJ. Ear canal cholesteatoma. Laryngoscope. 1992;102(6):608-13.
8. Vrabec JT, Chaljub G. External canal cholesteatoma. Am J Otol. 2000;21(5):608-14.
9. Garin P, Degols JC, Delos M. External auditory canal cholesteatoma. Arch Otolaryngol Head Neck Surg. 1997;123(1):62-5.
10. Hartley C, Birzgalis AR, Lyons TJ, Hartley RH, Farrington WT. External ear canal cholesteatoma: Case report. Ann Otol Rhinol Laryngol. 1995;104:868-70.
11. Fliss DM, Puterman M, Tovi F. Iatrogenic cholesteatoma of the neck. Head Neck. 1989;11:558-61.
12. Griffiths H, Raza A, Hayes M. Cholesteatoma: An unusual presentation. J Laryngol Otol. 2000;114(12):957-8.
13. Vartiainen E. Factors associated with recurrence of cholesteatoma. J Laryngol Otol. 1995;109:590-2.
14. Sheehy JL, Brackmann DE, Graham MD. Cholesteatoma surgery: Residual and recurrent disease: A review of 1,024 cases. Ann Otol Rhinol Laryngol. 1977;86:451-62.