International Journal of Otorhinolaryngology and Head and Neck Surgery
Vare R et al. Int J Otorhinolaryngol Head Neck Surg. 2023 Apr;9(4):314-317
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20230763

Case Series

Uncommon congenital swellings around external ear in adults

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Received: 24 January 2023
Revised: 16 March 2023
Accepted: 17 March 2023

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ABSTRACT

Congenital cyst around external ear appearing in adulthood is rare condition. Such lesions arising around the external auditory canal that requires surgical excision are uncommon and their clinical and surgical management is often difficult because these anomalies have variable relationship with facial nerve. In this study, we investigated characteristics of congenital cysts of the external ear by analyzing their clinical and histo-pathological features. Here we described 3 unique cases with localization of an epithelial cyst, type 1 branchial cleft cyst and epidermal inclusion cyst in adults which are uncommon, congenital and rare.

Keywords: External ear swelling, Congenital ear cyst, Branchial cleft cyst, Epidermal cyst

INTRODUCTION

The external ear consists of the auricle and the external auditory canal. The structural foundation of the auricle is the auricular cartilage, which is continuous with the cartilage of the external auditory canal. Congenital malformation of external ear can manifest as periauricular swellings. However this swellings are more commonly seen in pediatric population, its appearance in adult age is rare.[1] These congenital cysts can be due to branchial cleft anomalies or can be due to congenital epidermoid cyst.

Diagnosis of these cysts are based on clinical signs whereas highly suggestive diagnosis is based on lesion site. Generally both types of anomalies can present as progressive enlarging mass or recurrent swellings around external ear. The differential diagnosis of all these lesions can be made by histopathological examination. The preferred treatment for these cysts is complete removal of the mass. It is important because, in the literature, rare cases of malignant transformation of the congenital cyst have been reported. In this study we would like to describe three different unusual adult cases which came to our OPD with complaints of swelling around external auditory canal which was of congenital origin. Surgical excision was performed for management. The definitive diagnosis of these swellings, which were suggestive of epidermoid cyst for 1st case, first branch branchial cleft for 2nd case and epidermal inclusion cyst for 3rd case.

CASE SERIES

A 15 year old female came to our OPD with complaint of swelling on the right earlobe since last 4 months along with fullness of the right ear. There was no pain or history of trauma in the right ear, no history of right ear discharge or hearing loss or any other complaints. On physical examination, the right ear pinna showed a mass which was approximately 3×2 cm lobular, cystic, and painless on touch, smooth surface with normal skin (Figure 1). It was completely occluding the canal and tympanic membrane could not be seen. There was no discharging sinus or pointing abscess. The examination of the left ear was all within normal limits. The hearing test showed normal audiogram for both ears. After proper pre-operative evaluation, the patient was taken under general anaesthesia for removal of swelling, a post-auricular incision was taken, the skin was elevated from a lateral to medial

direction and the most posterior part of swelling was removed first (Figure 2). Care was taken to preserve the tympanic membrane. Swelling was removed in-total and sent for histopathology. Closure was done in layered suturing. Procedure went uneventful and patient was discharged after 2nd post-operative day.

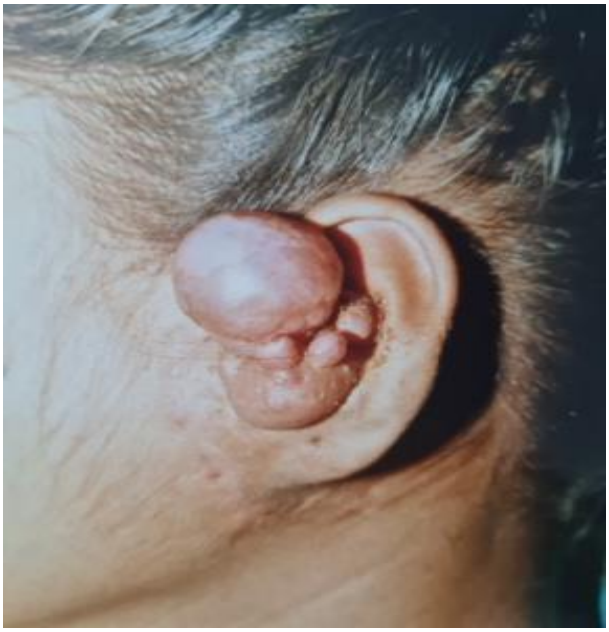

Figure 1: Right sided ear swelling showing involvement of external auditory meatus.

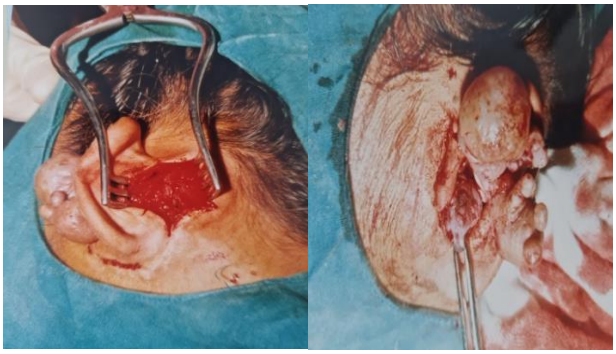

Figure 2: Intra-op pictures.

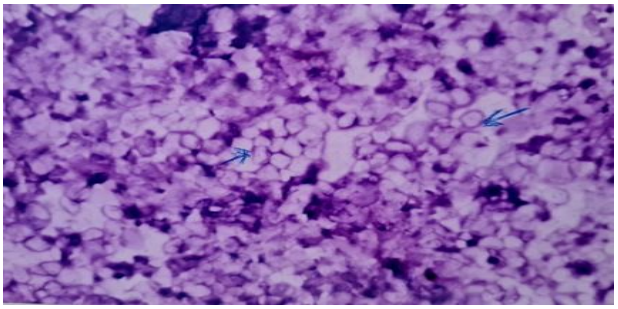

Figure 3: Microscopic picture of epithelial cyst.

The macroscopic specimen consists of cystic mass with cheesy material. Histologically, this specimen consisted of features of the epidermoid cyst-like presence of cystic lumen lined by stratified squamous epithelium with keratin. There was absence of cartilaginous tissue underneath squamous epithelium with presence of abundant cholesterol crystals in tissue as well as serous fluid (Figure 3). The preferred treatment for epidermoid cysts is the complete removal of the mass as we did in this patients. The patient was followed up during the next 6 months without recurrence of the lesion.

A 28 years old female came to our OPD with asymptomatic non-inflammatory cystic enlargement in the right ear pinna behind tragus and extending to postauricular region in vicinity to facial nerve (Figure 4). There was no history of trauma to right ear. No history of any surgical intervention. There was no complaint related to hearing or any ear discharge. On physical examination swelling was soft to firm on touch, without pain with smooth surface and normal overlying skin. The right ear tympanic membrane was intact. The left ear examination was normal. Both ear hearing was within normal limit.

Figure 4: Right sided ear swelling with its anterior and posterior extension.

Figure 5: Intra-op photo with excisied specimen.

After explaining consent and pre-operative evaluation the patient was taken under local anaesthesia. Local infiltration was given and postauricular incision taken. After dissection swelling was removed in total and sent for histopathology (Figure 5). Suturing was done. Procedure was uneventful and the patient was discharged on next day. Histo-pathological studies revealed presence of squamous epithelium on the cyst wall along with presence of cartilaginous tissue under squamous epithelium. There

was absence of cholesterol crystals and keratin material. This finding was suggestive of branchial cyst of ear (Figure 6).

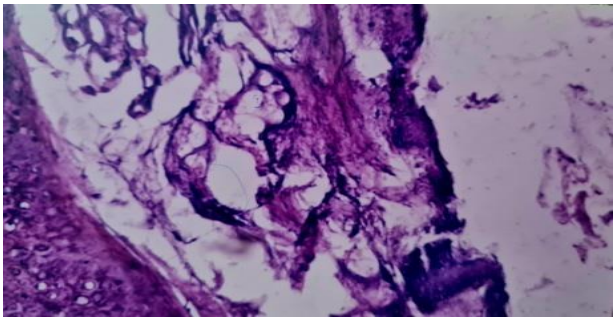

Figure 6: Microscopic picture.

Figure 7: Left eac swelling.

A 19 years old male presented to OPD with complaint of swelling of 0.5×0.5 cm occluding left sided external ear canal since last two years (Figure 7). Tympanic membrane could not be seen. Swelling was gradually increasing in size without any complaints of pain or hearing loss. No complaints of pre or post-auricular swelling or sinus or fistula. The swelling was in vicinity to facial nerve anterior to tragus. Opposite ear findings are within noraml limits. Audiometry revealed no hearing loss. High frequqence USG of local part was suggestive of 8×11×19 mm lesion in deep subcutaneous plane of left preauricular region and tragus of pinna likely to be 1st branchial cleft cyst. Contrast enhanced comupted tomography was also performed which was suggestive it to be of 16×11 mm hypodense lesion in left preauriclar region likely to be of vascular malformation (Figure 8).

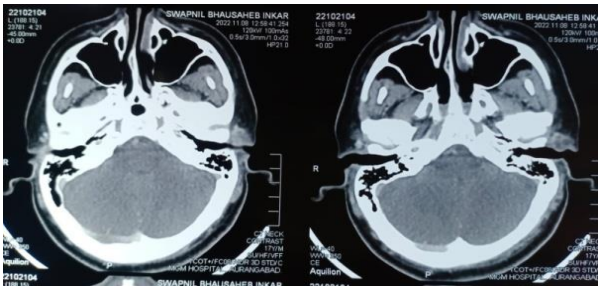

Figure 8: CECT s/o hypodense preauricular lesion.

After explaining the consent, patient was taken in general anaesthesia, endaural incision taken on left ear extending from incisura terminalis to posterior end of tragus (Figure 9). Dissection done, cyst indentified behind tragal cartilage (Figure 10). Anterior boundry to cyst dettached from periphery and cyst removed in-totol with its capsule. Excided specimen sent for histopathology which was suggestive to be of epidermal inclusion cyst.

Figure 9: Incision in incisura terminalis.

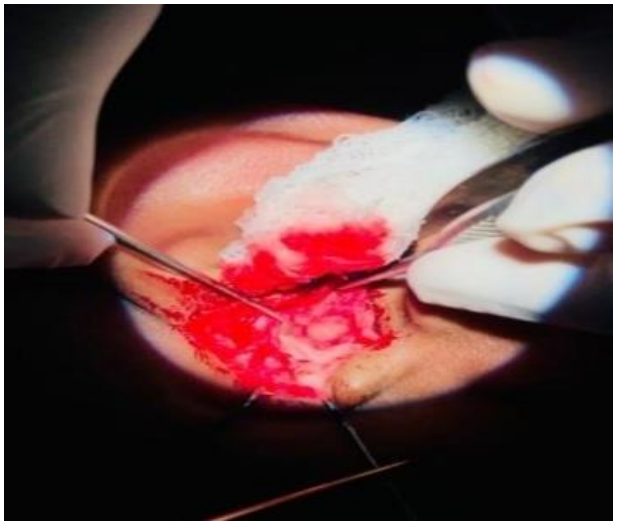

Figure 10: Cyst seen posterior to tragus.

DISCUSSION

Anatomically, the ear has three distinct parts: the external, middle, and internal ear. The outer ear consists of the

visible part called the auricle, or pinna. The auricle is formed from the mesenchyme in the region of the first branchial cleft. It consists of a thin sheet of yellow elastic cartilage covered by closely adherent skin and the tragus is derived from the first arch and the rest from the second arch. Congenital malformation of the external ear can influence the orientation, position, size and configuration pattern of the pinna. Some anomalies may occur at birth which can be ear tags, ear sinus, ear pits and preauricular or postauricular cystic swellings and some can arise from arrested or aberrant embryological development. In the later stages, symptoms may appear suddenly in adulthood without symptoms in childhood.

As we saw in the first case, epidermoid cysts can be congenital or acquired; congenital epidermoid cysts can be caused by entrapment of ectodermal elements intradermally or subcutaneously during fusion of the first and second branchial arches.[2] Acquired cysts result from traumatic or iatrogenic implementation of epidermal cell proliferation into the dermis due to penetrating injury, or occlusion of a sebaceous gland duct in the hair follicle.[3] Tympanomastoid surgery can cause a traumatic epidermoid cyst as a complication.[4,5] However, their appearance in the EAC is extermely rare. The differential diagnosis of an epidermoid cyst in the EAC includes EAC cholesteatoma, dermoid cyst, inflammatory polyp, and soft tissue tumors such as hemangioma, lipoma, meningioma, and acoustic neuroma.[6] Significant time may be required before the final diagnosis of these lesions. Our patient presented with progressive painless ear fullness lasting for about 3-6 months. In our case, the epidermoid cyst of the external ear canal is thought to be an idiopathic type due to the lack of trauma history and the adult age of the patient.

A dermoid cyst with an epidermoid cyst can be confused with each other due to macroscopic features. Epidermoid cyst consists of keratinous cyst covered by stratified squamous epithelium without adnexal structures but the dermoid cyst contains keratin, sebaceous glands, hair follicles, and adnexal structures within the wall of stratified squamous epithelium.[7]

Where as in 2nd case: FBCA accompanied by external ear diseases are uncommon, and to date, only a few FBCA cases along with congenital EAC stenosis or atresia, and cholesteatoma have been reported. The unique presentations and duplicated anomalies of FBCA are challenge for surgeons. As the lesions are usually very deep and often involved with the FN and parotid gland; surgical expertise must be available to remove the lesion completely, preserve FN function, and perform otologic reconstruction if necessary.[8] Despite the rarity of coincident malignancy in an epidermoid cyst, rapidly changing in size, inflamed or painful should be examined histopathologically to exclude malignancy. Malignant transformation takes time and rate of an epidermal cyst into cutaneous squamous cell carcinoma range from 0.011 to 0.045%.[9]

CONCLUSION

Any cyst around external ear should be considered for appropriate management as it is important to remove it completely because of the low possibility of malignant transformation. Their clinical management with successive surgical treatment can be achieved by taking into account the clinical settings and intraoperative finding. Their confirmatory diagnosis can be achieved by histo-pathological findings of excised specimen.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Vare R, Vare A, Desai N. Uncommon congenital swellings around external ear in adults. Int J Otorhinolaryngol Head Neck Surg 2023;9:314-7.