A case of salivary gland choristoma presenting with ear discharge in a child having external auditory canal stenosis

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SUMMARY

Salivary gland choristoma in the middle ear or external ear canal is rare. Generally, it is difficult to differentiate salivary gland choristoma from congenital cholesteatoma or cholesterol granuloma on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). It is challenging to diagnose preoperatively without specific clinical or radiological symptoms or signs. Thus, histopathological examination is required for a definitive diagnosis, and a histopathological sample of the lesion is acquired. We discuss the imaging and histology of salivary gland choristoma with the best recommendation on the management.

INTRODUCTION

Salivary gland choristoma is a type of heterotopia, a phenomenon where normal salivary gland tissues are found in an abnormal site.[1] The common locations are the posterior lobe of the pituitary gland, the periparotid lymph nodes, the middle ear, and the lower neck.[2] A unilateral conductive hearing deficit is typically the first manifestation of choristoma, with otorrhoea and occasionally a mass in the middle ear. This lesion may be associated with deformed or absent ossicles as well as branchial arch and facial nerve abnormalities.[1] Most of the reported cases of salivary gland choristoma were in children (10 cases) and adults (38 cases). Thus, we present a unique case of salivary gland choristoma presented in an infant, its clinical presentation, and radiological findings with histopathological reports.

CASE REPORT

A one-year-old baby girl was seen in the otorhinolaryngology (ORL) clinic with the complaint of episodic right ear discharge. She was born with right microtia and external auditory canal (EAC) stenosis. She also presented with right facial nerve palsy, evidenced by slight deviation at the right angle of mouth and loss of nasolabial fold. Hearing screening with the distortion product otoacoustic emission (DPOAE) machine showed a pass result in the left ear and refer result in the right ear. During her clinic follow-up, the mother claimed that the episodic mucoid discharge from the right ear improved after using topical eardrops (ofloxacin). However, since the age of eight months, there was an off and on fluctuant swelling measuring 2 × 2 cm with erythematous skin at the right infra-auricular area. Initially, it was treated with oral antibiotics, but on the third episode, the swelling did not resolve with oral medication. Hence, aspiration of the swelling was done whereby 3 ml of pus was aspirated using a needle and syringe. On follow-up, there was fibrosis at the aspirated area with the formation of cutaneous fistula with off and on mucoid discharge. Otherwise, the mother claimed that her baby was comfortable and growing well.

Due to persistent ear discharge from the canal and fistula, the baby was referred for a high-resolution computed tomography (HRCT) of the temporal bone and MRI of the ear. The HRCT findings showed a stenotic cartilaginous part and an atretic bony part of the right external auditory canal. The impression was right ear congenital cholesteatoma with absent right stapes, dysplastic long crus of right incus, and collection at the right posteroinferior auricular region. The right and left facial nerves courses were intact; however, the right stylomastoid foramen was widened compared to the left side. The MRI showed a well-defined rounded lesion posterior to the bony part of the EAC measuring 0.76 cm anteroposterior (AP) × 1.2 cm width (W) × 0.9 cm craniocaudal (CC) (Figure 1a). Lateral to this lesion, there is a well-defined collection measuring 0.3 cm in maximum thickness. Axial view of T2 drive showed normal right facial nerve from cisternal part of cerebellopontine angle to first genu. The impression was right middle ear cholesterol granuloma with the adjacent collection and right mastoiditis. The patient underwent right cortical mastoidectomy and removal of the right posterior EAC mass via trans-mastoid approach with the excision of the right infra-auricular cutaneous fistula. Intraoperative findings showed a pearly white sac occupying the right EAC measuring 2.0 × 1.5 cm (Figure 1b), that the EAC was narrowed, and the presence of a tract connecting the fistula area to the canal and noted mucoid discharge coming out from the fistula. No keratin was seen in the mastoid cavity, whereas keratin debris was seen at the EAC. The facial nerve was unable to be identified.

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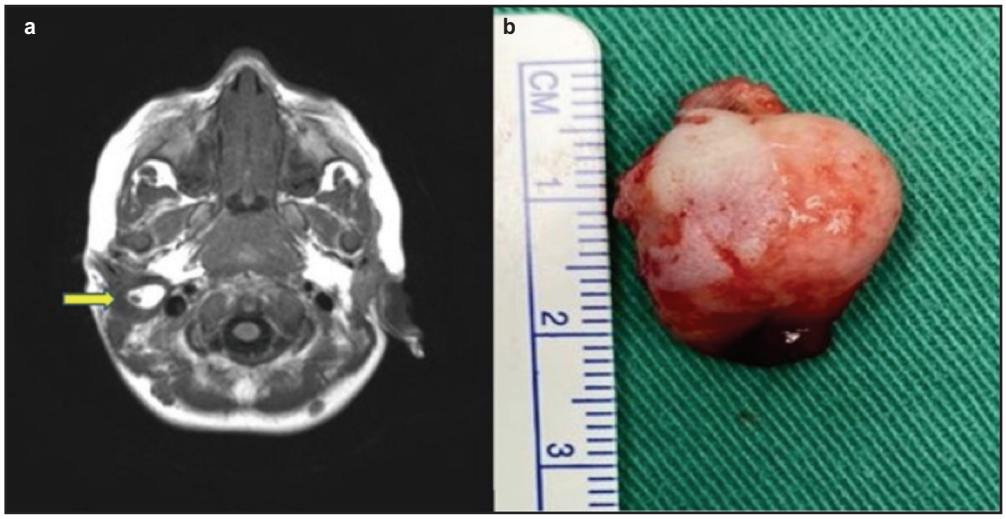

Fig. 1: Images of the choristoma. (a) MRI T1-weighted images in axial view; the arrow pointed to a well-defined rounded lesions seen posterior to the bony part of the right EAC. (b) Gross specimen of the whole sac of right auditory canal mass (pearl white sac size 2.0 × 1.5 cm) that was removed intraoperatively.

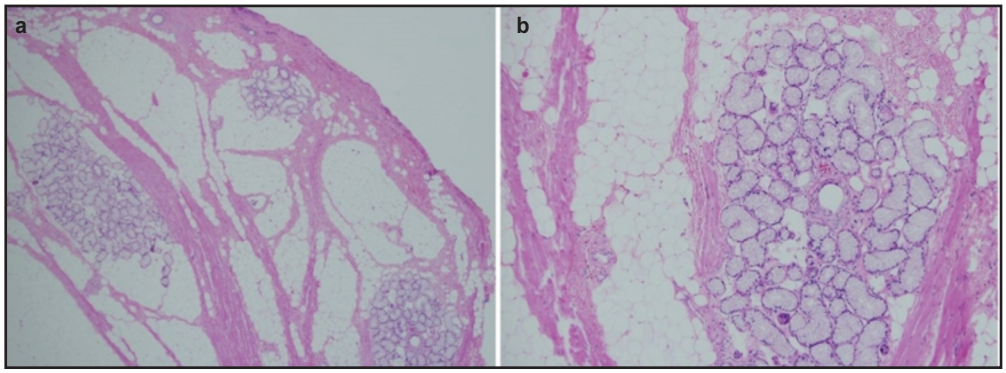

Fig. 2: Histopathological slides of the choristoma. (a) Lobules of mature adipocytes are separated by fibrous septa and scattered seromucous gland units (x10 magnification). (b) Benign seromucous gland units within the fatty tissue lobules as well as skeletal muscle bundles (x40 magnification).

The histological report stated a fairly well-circumscribed mass partly covered by stratified squamous epithelium. It is composed of lobules of mature adipocytes separated by fibrous septa. Scattered lobules of seromucous gland units were noted within the fatty tissue lobules as well as skeletal muscle bundles (Figure 2a and Figure 2b). The features were compatible with salivary gland choristoma.

DISCUSSION

The pathogenesis of the disease of choristoma is unclear. However, the choristoma occurrence is before the fourth month of gestation due to the failure of the development of the first or second branchial arch.[3] It is related to the ectopic expansion's remnant parotid epithelium or pharyngeal endoderm.[4] A few syndrome components include salivary gland choristoma, hearing loss, ossicular chain abnormalities, facial nerve anomalies, second branchial arch anomalies,[5] and other brachial arch anomalies that may also accompany the lesion.[6] The various age ranges were between 9 months and 52 years old,[7] with a ratio of two women to one man.[4]

The clinical disease feature of salivary gland choristoma occurrence in the middle ear is usually a unilateral lesion (96.8%), on which the left side was the most frequently affected (61.3%).[7] The middle ear mass must be differentiated from congenital cholesteatoma, dermoid cyst, teratoma, glomus tympanicum, granuloma, neuroma, or glioma, which are present with unilateral hearing loss without perforation of the tympanic membrane.[8] Less common areas include the upper neck, lingual mandible, external auditory canal, thyroid gland, mediastinum, prostate gland, vulva, and rectum.[2] The choristoma can be completely excised, but if it is poorly defined and intimately associated with the facial nerve, partial excision of the lesion is appropriate to prevent damage to the nerve.[1]

The extend of the excision of salivary gland choristoma is controversial. Some authors thought that salivary gland heterotopia consists of normal tissue and, thus, did not require complete excision when diagnosed histologically.[9] However, when there are signs of infection or neoplasm, it requires complete removal of the lesion.[9] The possibility of malignant transformation is very rare, but, theoretically,

choristomas are immature in nature, increasing the chance of malignancy.[10] In our case, we removed the mass with its sac and closed the fistula as a whole. It has given a better quality of life to the patient and family. There is no recurrence of ear discharge and new swelling at the operation site post-surgery.

CONCLUSION

Salivary gland choristoma of the external auditory canal is a rare benign condition and should be differentiated from other middle ear or external ear canal lesions. Infants with ear mass and recurrent otorrhea should have the salivary gland choristoma as one of the differential diagnoses. While the diagnosis of salivary gland choristoma is confirmed by histology, the best way is to remove all the lesions operatively (diagnostic and therapeutic).

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