Work type II first branchial cleft cyst: a rare anomaly with a classical presentation

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First branchial cleft cysts are rare and can present as a diagnostic challenge to the physician. There can be frequent misdiagnoses, leading to a delay in treatment. This may result in mismanagement, causing an increased rate of recurrence. Moreover, their close relationship to the facial nerve would necessitate the exposure and preservation of the facial nerve. We report a case of a patient with the classical presentation of a Work type II branchial cleft cyst. Imaging showed a lesion just adjacent to the external auditory canal. Intraoperatively, a cartilage-lined blind-ending sac with hair-bearing contents duplicating the external auditory canal was found. The case highlights the need to consider the diagnosis of first branchial cleft anomaly especially in the presence of cysts and sinuses within the region of the parotid and the upper neck. Complete surgical excision would be the mainstay of treatment to prevent future recurrence. ***Ann Pediatr Surg*** **10:81–82 © 2014 Annals of Pediatric Surgery.**

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Introduction

First branchial cleft anomalies are considered to be rare and make up about 10% of the branchial cleft anomalies encountered [1,2]. They can present in variable locations, from the retroauricular and parotid region to below the mandible and above the hyoid bone [3]. In view of their rarity, the physician needs to have a high index of clinical suspicion when dealing with cysts around that region, especially in the pediatric population.

Understanding the embryology of first branchial cleft anomalies is crucial in knowing how to adequately manage such cases. The branchial apparatus starts to appear between the fourth and the fifth weeks of fetal development. The apparatus consists of five arches consisting of the mesoderm, four clefts that consist of the ectoderm and four pouches that consist of the endoderm. The first branchial pouch forms the Eustachian tube and the tympanic cavity, whereas the first cleft forms the external auditory meatus and part of the concha [4].

Work [5] described a classification based on histology and embryology. Type I is of ectodermal origin and is a duplication of the membranous auditory canal. Type II consists of duplication anomalies of the membranous external auditory canal and pinna, and they contain both the ectoderm and the mesoderm.

In this case report, we have intraoperative images depicting a Work type II case of duplication of the external auditory canal that is cartilage lined, containing hair-bearing skin.

Case report

A 6-year-old boy was referred for recurrent left infra-auricular swelling associated with discharge since 2011. He presented with recurrent episodes of facial swelling associated with an infected cyst and had undergone two previous incision and drainage procedures. He was otherwise healthy.

On physical examination, there was a cyst located in the infra-auricular region with a scar from previous incision and drainage. His tympanic membrane was normal and there was no discharge in his external auditory canal.

The patient's subsequent computed tomography showed a well-circumscribed lesion about 0.9 by 0.7 cm in dimension, located within the superior left parotid parenchyma adjacent to the inferior border of the external auditory canal, consistent with a first branchial cleft cyst (Fig. 1).

During the surgery, the branchial cleft cyst was identified to be superior and deep to the facial nerve trunk. Because of the repeated incision and drainage procedures, there was a lot of granulation tissue seen. The facial nerve monitor assisted in differentiating the scar tissue on the nerve and the nerve itself. The main trunk of the facial nerve was identified and preserved. Duplication of the external auditory canal was found with a hair-bearing and cartilage-lined blind-ending sac. There was an epidermal layer without dermis that separated the duplicated track and the real external auditory meatus. The cyst was dissected free and the sac was removed with no injury to the facial nerve (Fig. 2). After dissection of the cyst, a portion of the external auditory canal was left deficient of cartilage where it was originally attached to the duplicated track.

During the postoperative recovery, the cartilage-deficient area of the external auditory meatus broke down, and seroma was drained from that area for 1 week. The external auditory meatus was stented with pope wicks for 5 days at the site where seroma formed. Thereafter, it was stented for another week to prevent narrowing of the external auditory canal. The patient subsequently recovered well with no recurrences.

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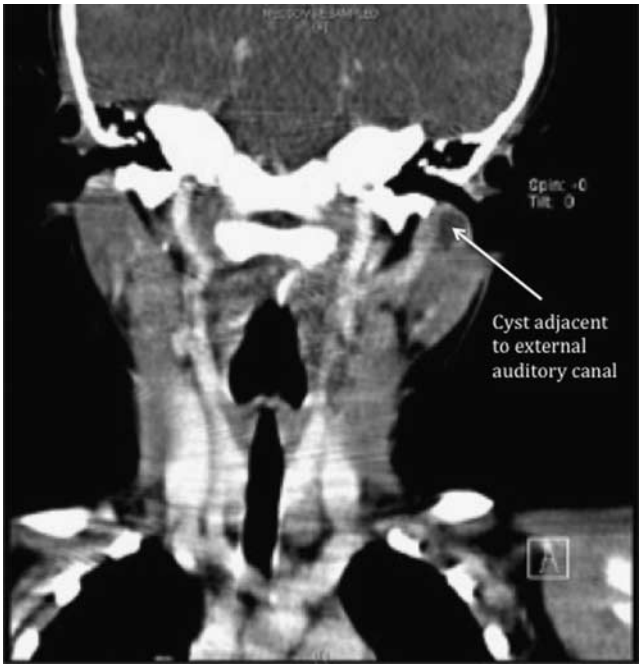
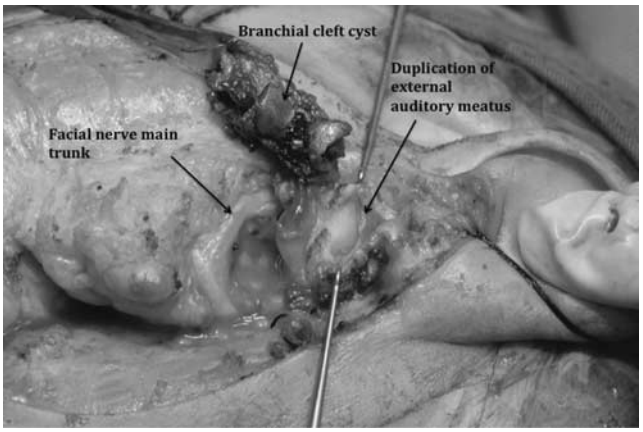
Fig. 1



Computed tomographic findings of the patient showing a cyst just adjacent to the external auditory canal. There was no definite track seen.

Fig. 2



Duplication of the external auditory canal opened up revealing a blind sac lined with cartilage containing hair-bearing epithelium.

Discussion

As first branchial cleft anomalies are rare, they are easy to misdiagnose and mismanage. In the series by Magdy and Ashram [6], 50% of the patients received unsuccessful treatment and one patient even had nine incision and drainage procedures for his infected branchial cyst. This emphasizes the fact that there needs to be high index of suspicion when it comes to managing cysts around the region of the parotid in the pediatric population.

It is described that cutaneous defects tend to occur within a triangle with the apex at the external auditory meatus, its base a line between the tip of the chin and the middle of the hyoid bone, and its two remaining sides curving from external auditory meatus to the tip of the chin along the body of the mandible and from the external auditory meatus to the greater cornu of the hyoid bone [7].This area is known as Poncet's triangle [8]. Thus, physicians managing cysts and sinuses in that area have to be aware of the possible diagnosis of first branchial cleft anomaly.

The treatment of first branchial cleft anomalies would be surgical excision. Most surgeons have reported surgical excision using a superficial parotidectomy approach [2,3,6]. Complete excision is of importance and limited resection would lead to recurrence. It is vital to identify and preserve the facial nerve during the surgery. Attempts at limited excision through small skin incisions have caused injury to the branches of the facial nerve [7]. The relationship of the facial nerve is variable and no definite conclusion can be drawn. Triglia *et al*. [7] demonstrated that 28% passed deep to the nerve, 8% split around the nerve, and 64% were superficial. Magdy and Ashram [6] reported that the facial nerve was superficial in 72% of the cases and deep or partially deep in 28% of the cases. Given this variability, the surgeon involved has to be familiar with parotid surgery in children and have a readiness to expose the nerve when required.

This case illustrates a patient with a first branchial cyst, from diagnosis to successful surgery. The intraoperative images show the duplication of the external auditory canal that contains both ectoderm and mesoderm elements. With the facial nerve in such close proximity, the surgeon has to take considerable care to avoid injury to the main trunk and its branches.

Conclusion

First branchial anomalies are rare clinical entities and diagnosis can be challenging at times. Physicians have to be mindful when dealing with cysts and sinuses in the region of the parotid and the upper neck. Surgical excision remains the mainstay of treatment, and the surgeon has to be willing to expose and delineate the facial nerve when necessary to avoid injury.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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