Type I First Branchial Cleft Abnormality with Aural Atresia: Undetected External Auditory Canal

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INTRODUCTION

Type I first branchial cleft abnormalities (FBCA) are a rare and challenging condition to diagnose, with a high incidence of complications. These abnormalities are caused during embryologic development and account for only 10% of all branchial cleft abnormalities. They present with a fistula, sinus, or cyst on the floor of the external auditory canal, near the parotid region or hyoid bone, along with an abnormal or absent external auditory canal.

A clinical symptom of this abnormality includes recurrent infections in the external auditory canal (EAC) or, as in this case, an abscess in the postauricular or parotid area. We present a case of Type I FBCA associated with microtia and aural atresia that exhibited no canal presence on MRI, but with the subsequent discovery of an external auditory canal on endoscopy.

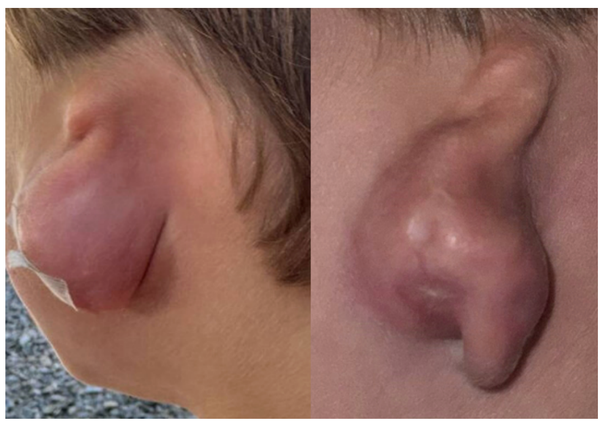

Figure 1: Abscessed microtia following initial branchial cleft excision

BACKGROUND

During development, the first branchial arch develops into the external auditory canal (EAC) and tympanic membrane. Abnormal fusion between the first and second branchial arches can lead to first branchial cleft anomalies (FBCA), which can exist without disrupting the external auditory canal anatomy. The classic presentation of FBCA is drainage from the external auditory canal or swelling in the parotid area.[1] These anomalies fall into two categories: Type I FBCA or Type II FBCA. Type I is characterized by a cystic mass in the postauricular area that extends into the posterior wall of the EAC. Type II FBCA occurs along the earlobe to mandible angle and may present as a fistula or cyst.[1] A fistula is defined as a tract with two openings, while a cyst is a tract with no opening.[1] Both types can form along a spectrum, ranging from no extra cartilaginous formation to variations in EAC development.[1] FBCA accompanied by microtia and aural atresia, as in this case, has only been reported in a few instances.[3]

CASE PRESENTATION

We present a 5-year old male with a history of left sided craniofacial microsomia, grade 3 microtia and aural atresia who developed persistent drainage from a preauricular pit. CT scan showed concern for FBCA along with dysmorphic but intact ossicles. Both CT and MRI showed no evidence of a cartilaginous external auditory canal.

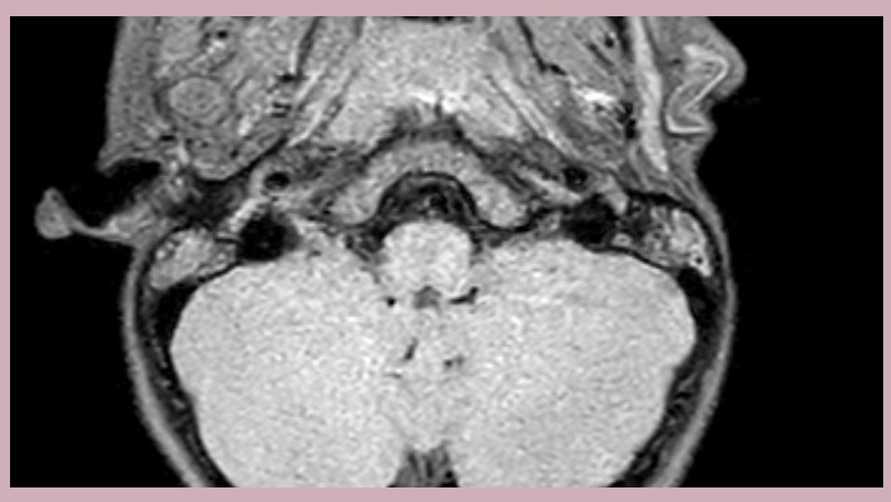

Figure 2: Axial image showing no external auditory canal on left side

A first branchial cleft abnormality excision was performed where a sinus tract that was identified, excised and tied off deep. The patient had recurrent abbesses drained from the surgery site prompting an exploratory repeat branchial cleft remnant removal surgery. During the procedure, nonviable tissues were removed uncovering normal cutaneous skin along the cartilage internally. An endoscope revealed a circumferential cartilaginous canal along with a rudimentary tympanic membrane.

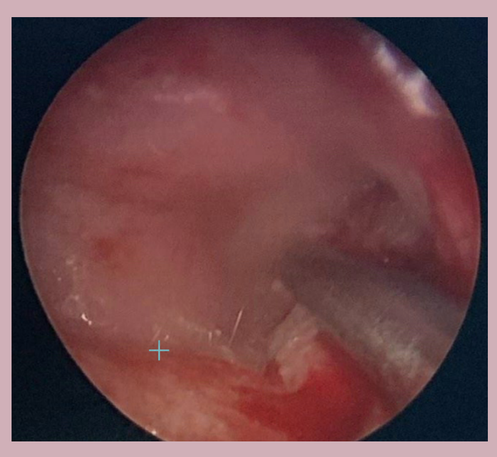

Figure 3: endoscopic view of complete circumferential cartilaginous canal, and a rudimentary tympanic membrane

A canaloplasty was preformed and a small rim of cartilage was removed around the canal to expose the external auditory canal skin. The skin of the microtia was then sutured to the newly uncovered external canal skin using 4-0 Monocryl suture to recreate a normal canal. Local rearrangement of surrounding soft tissue was used to embellish the look of the external ear once the canal was created.

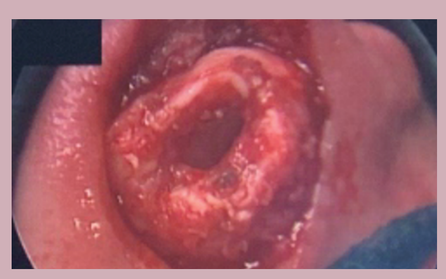

Figure 4: External auditory canal uncovered

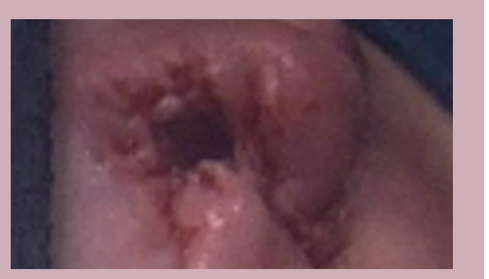

Figure 5: New external ear canal following canaloplasty

Creating this functional canal improved sound recognition on the affected side but due to hypoplastic ossicles and tympanic membrane, complete resolution of conductive hearing loss is not possible. Postoperative audiometric testing showed a slight improvement in the conductive hearing loss along with a type B tympanogram in left ear. The patient's conductive hearing loss is being successfully managed with a bone-conduction hearing aid attached to a flexible headband.

CONCLUSION

In this case, an exploratory incision and drainage (I&D) procedure led to the discovery of an external auditory canal (EAC), prompting a canaloplasty to improve hearing. This finding contradicts existing literature that individuals with FBCA, craniofacial microsomia, and grade 3 microtia typically have aural atresia. The surgeon's successful identification of the tympanic membrane, combined with prior knowledge of a possible EAC in FBCA, significantly improved this patient's quality of life. This case contributes to the understanding of potential FBCA presentations and successful treatments in improving both hearing and cosmetic outcomes.

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