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CASE REPORT

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A Case of Type I and Type II First Branchial Cleft Sinus and Fistula

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Abstract

The diagnosis of head and neck masses and fistulas can be challenging. Differential diagnoses include tumors, infections, or congenital lesions. Knowledge of embryology and anatomy of both common and rare anomalies, as well as indications for imaging, is required for timely diagnosis. We describe a branchial anomaly with atypical presentation as an uninfected "pit" in the earlobe that required multiple procedures to achieve complete resection. Reviewing the challenges experienced in the treatment of this case and the literature, we discuss the role of imaging in surgical planning to avoid incomplete resection, prevent recurrence, and minimize need for multiple procedures.

Keywords: First branchial cleft cyst; Imaging; Fistula; Misdiagnosis; Recurrence

Abbreviations: FBCC=First Branchial Cleft Cyst; MRI=Magnet Resonance Imaging; CT=Computed Tomography

Case Report

A healthy 3-year-old girl presented to the Otolaryngology clinic with a history of rare moisture draining from a pit in her left earlobe. Though the pit was present since her birth, she passed her newborn hearing test. One prior infection of the earlobe was reported which responded to oral antibiotics. There was a second miniscule pale spot (no opening) under the mandible which has never drained. On physical examination, the left auricle and external auditory meatus were normal without evidence of duplication of the external auditory canal. On the anterior surface of the left earlobe was a 1-mm punctum with no obvious cyst or mass. Scant moisture was expressed when massaged. In the level II neck below the border of the mandible, a 2-3-mm flat, pale spot was noted. On palpation, the left parotid and face were normal without masses or facial nerve paresis. A clinical diagnosis of congenital left earlobe pit/fistula with left neck branchial remnant was made. Surgical excisions of both lesions were offered, and preoperative imaging was not ordered.

When elliptical surgical excision for the neck spot was performed, no tract was identified deep to the skin. A separate elliptical incision was made around the ear lobe pit, and a lacrimal duct probe was used to identify potential fistula or tract. The probe only passed about 2 mm and did not lead to any tract, and the wound was closed primarily. The patient presented 3 weeks postoperative with a swollen and erythematous ear lobe with mucopurulent, sticky drainage (Figure 1). Cephalexin did not lead to improvement. The patient was taken to the operating room for incision and drainage of presumed localized wound abscess. Intraoperative exploration within the left earlobe soft tissue showed no identifiable tract or meatus. The wound was loosely closed to allow healing by secondary intention, and the patient was treated postoperatively with a course of antibiotics and topical 2% mupirocin ointment. Cultures for aerobic and anaerobic organisms were negative.

The patient developed recurrent symptoms and another excision was performed with extension of dissection to the tragus. After removing all granulation, the surgeon identified a fistula completely lined by skin from the earlobe towards the auricle, appearing to end at the inferior aspect of the tragus. This tract was followed, and no cysts were identified. The tract was ligated and removed, and the wound was irrigated and closed. A histology report identified a fistula lined by stratified squamous keratinized epithelium and chronic inflammation. Once again, postoperative antibiotics and ointment were prescribed.

Eighteen days later, the child had recurrent symptoms of erythema and drainage from the left earlobe. At this time, the surgeon recommended an MRI with contrast to identify the exact extent of the fistula. Given that general anesthesia was required for this study, the family declined the MRI, concerned their child had already undergone several anesthetics. Instead, they opted to proceed with

another attempt at excision. The parents met with a second surgeon in the group, who ordered a repeat attempt at complete excision. The second surgeon excised all granulation tissue from the left ear lobule to the tragal cartilage again, as well as the cartilage of the anterior and inferior external auditory canal. After careful exploration, no further fistula was identifiable, and the wound was closed primarily.

The child did well for one month after the excision of the granulation tissue, but symptoms recurred. She was admitted for IV antibiotics and an MRI of the face/neck with contrast.

Gadoterate meglumine (Dotarem; Guerbet, Bloomington, IN, USA) was given at the standard dose of 0.1mmol/kg. MRI revealed a large-caliber fistula starting from the left ear lobe, traversing medially through the parotid gland, deep and parallel to the facial nerve, then diving deep inferiorly, exiting through a pinpoint where the prior neck dimple was excised (Figures 2,3). The fistula tract was anterior to the expected location of the facial nerve. Complete excision was performed the next day through a parotidectomy approach with facial nerve monitoring (Figure 4). The histopathological findings of the specimen showed a fistula tract lined by squamous keratinizing epithelium and adjacent parotid gland tissue (Figure 5). The skin and cartilaginous part of the inferior aspect of the external ear was also resected to ensure a complete excision of the fistula. The child recovered well without any complications and was asymptomatic at the 7 years follow-up.

Discussion

First branchial cleft cysts (FBCCs) are rare anomalies caused by embryonic development error between the fourth and seventh weeks of development because of incomplete obliteration of branchial

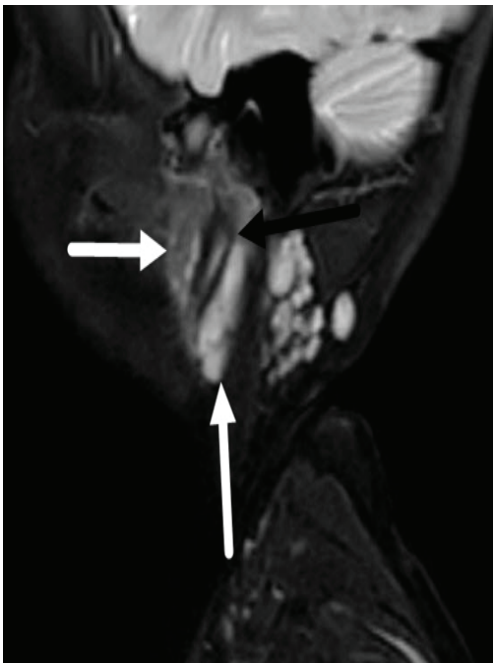

Figure 2: Coronal T2 weighted fat saturated image through the mid parotid gland. The dark arrow shows the double dark lines representing the walls of the fistula (tubular structure) starting from the lower ear lobe extending medially in a horizontal fashion, diving down and dividing the parotid gland (white arrows) into a medial and lateral part.

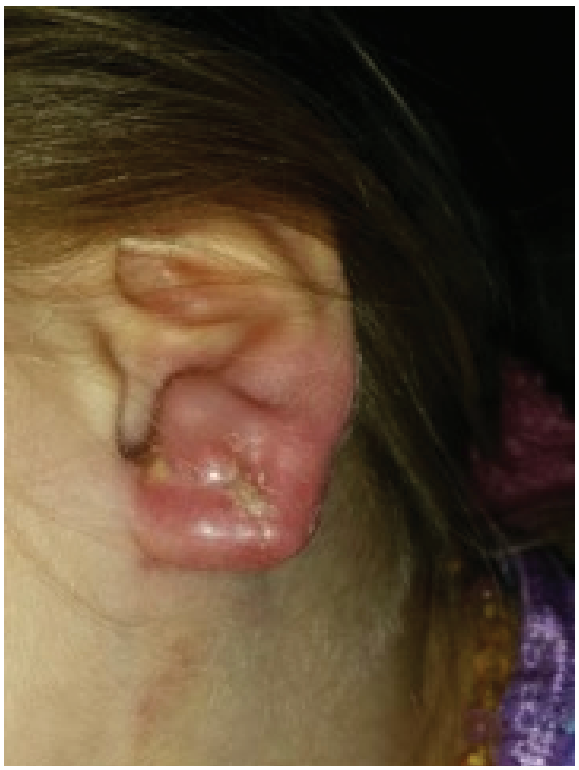

Figure 1: Gross specimen showing left earlobe abscess with erythema, edema, and purulent drainage.

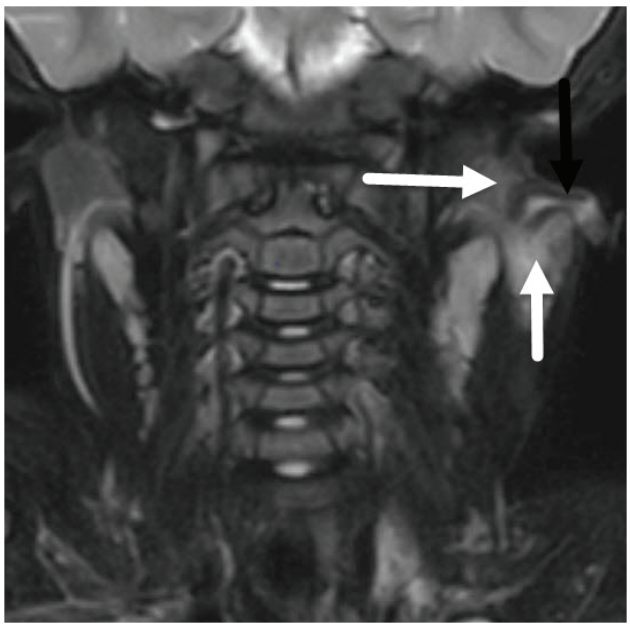

Figure 3: Sagittal T2 weighted fat saturated image showing similar finding in a different plane. Black arrow shows the double black tube (fistula) dives anteriorly and inferiorly from the horizontal component of the fistula after making almost a 90-degree angle. The distal end of the fistula ends at the pit of the left mandibular angle. The fistula divides the parotid gland into an anterior and posterior part on this plane.

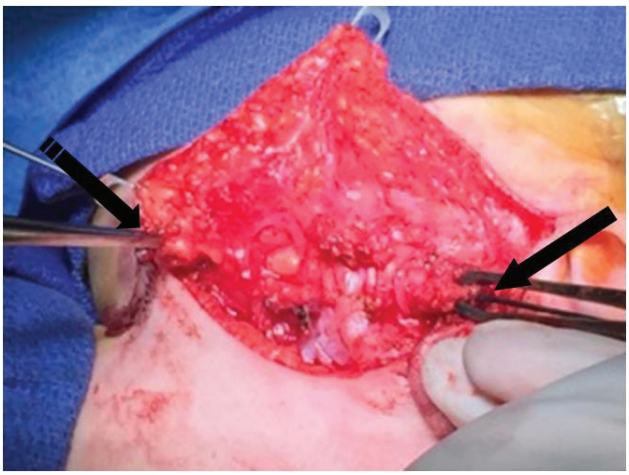

Figure 4: Intraoperative gross specimen: Parotidectomy approach with facial nerve dissected and identified. The entire tubular fistula is found to be deep to the facial nerve. A lacrimal duct probe has been inserted through the fistula (dark arrow) proximally with an Ellis clamp. The fistula ends distally near the mandible.

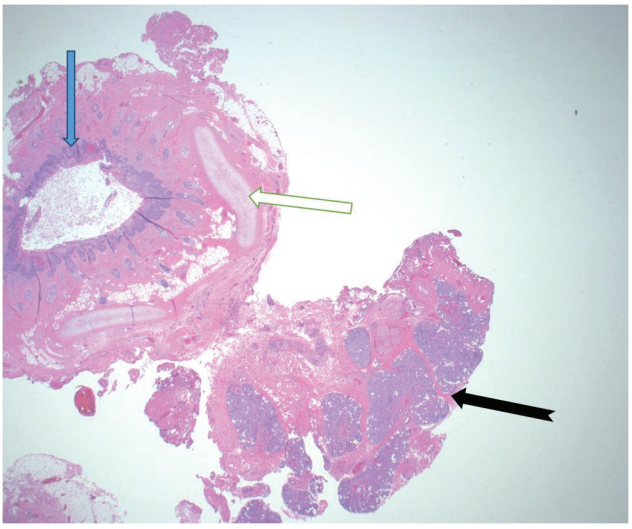

Figure 5: Image with H&E staining at 12.5× magnification. Fistula tract lined by stratified squamous keratinizing epithelium (single-headed arrows) and adjacent parotid gland tissue (double-headed arrow).

arch structures [1]. These anomalies are defined by their anatomical relationship to branchial arch structures and account for 1–10% of all branchial cleft anomalies [2-5].

The first classification of FBCC was described in 1971 by Arnot. Type I describes a cyst or sinus inside of the parotid gland coated by squamous cells, which are present in adult life. Type II describes a cyst or sinus developed during childhood, communicating with the auditory external canal and the neck [3]. In 1972 [6], Work suggested another classification: type I is defined by the duplication of the external auditory canal, parallel to normal external canal, covered by squamous cells, and ended in a cul-de-sac; type II describes the duplication of the external auditory canal and the pinna, lined by skin and cartilage, communicating the auditory external canal with the mandible or neck. In 1980, Olsen et al proposed that defects of the first branchial groove can occur as cysts, sinuses, or fistulas [1]. Cysts are caused by hidden cells inside the groove that are not obliterated. The sinus has the same component as the cyst but is accompanied by a sinus tract that opens at the surface of the ectoderm or auditory external canal. Fistulas are an incomplete closure of the first groove, creating a fistulous tract going through the middle or external auditory canal to the neck or mandible.

We describe a challenging case of both type I and type II anomaly with the presentation of only a "pit" in the earlobe. An extremely large fistula was ultimately discovered on imaging after recurrent incomplete excisions for persistent earlobe infection. Complete resection of the fistula was achieved after imaging.

Work's is the most often used classification to describe FBCC anomalies. The parotid gland appears at the sixth week of embryonic development close to the angle of the mouth and migrates to form a tube that achieves contiguity with the ear where it is inserted. The facial nerve and its muscles migrate upward at the sixth to eighth week to the same region [1]. Type I FBCC is a differential diagnosis of parotid tumor, which is common between ages 40 and 70 years (male to female ratio, 2:1) [7]. Often, FBCC may present as recurring parotid swelling or mass in children and may be mistakenly treated with incision and drainage of a parotid abscess and antibiotic therapy, which is more compatible with FBCC type II [6,8]. In our case, MRI allowed identification of the entire extent of this anomaly, enabling definitive excision of the entire fistula tract. Our case is a rarity of both type I and type II, given the extent of the anomaly from earlobe to the neck [1,3,5,6,8].

The histopathological finding at the first surgical resection, in March 2015, was compatible with the findings of Work classification FBCC type I [6]. The last surgery, in August 2015, with complete resection of the anomaly, was compatible with FBCC type II [6]. In hindsight, we realize preoperative imaging would have identified the true extent of this complex anomaly. Preoperative imaging is not routine for surgical planning of excision of preauricular pit/fistulas. As this patient presented initially with minimal drainage from the pit in the lateral earlobe, it did not suggest duplication of the external auditory canal. The surgeon made a diagnostic error in not recognizing the type II FBCC at the outset given the spot under the mandible. Seeing both should have increased suspicion and prompted use of MRI with contrast to rule out fistula before surgical excision was attempted [4,5].

This case highlights the importance of imaging, especially when patients present with minimal symptoms [2,4,7- 11]. Given the current focus on the Image Gently campaign and the goals of minimizing radiation to children, options for imaging modality include CT scans, MRI, and ultrasonography, with MRI being the best option for avoidance of radiation. As Coppens et al articulates, imaging is a valuable tool in these cases: "A definitive CT diagnosis of FBCC type II was possible in 83% of the cases. Exclusion of diagnosis was possible in 94% of FBCC mimics. The total accuracy was accuracy of 90%" [4,8]. According to Sichel et al, "A ring of cartilage around the fistulous tract in CT imaging may also be another significant diagnostic finding" [12]. We did not identify such a ring.

Conclusion

Preoperative imaging for unimpressive ear pits or fistula may be necessary-even critical-to identify the full extent of branchial anomaly and to prevent multiple unnecessary procedures.

Teaching Point

The diagnosis of masses and fistulas of the head and neck can be challenging. Imaging is crucial in diagnosis and surgical planning to avoid incomplete resection, recurrence, and unnecessary repeated procedures.

Disclosures

None.

References

1. Olsen KD, Maragos NE, Weiland LH (1980) First branchial cleft anomalies. Laryngoscope 90: 423-436.
2. Triglia JM, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN (1998) Frist branchial cleft anomalies: a study of 39 cases and review of the literature. Arch Otolaryngol Head Neck Surg 124: 291-295.
3. Arnot RS (1971) Defects of the first branchial cleft cyst. S Afr J Surg 9: 93-98.
4. Alshihmani SHA (2023) A second branchial cleft cyst, a case report. Int J Surg Case Rep.
5. Han Y, Yang RQ, Hong L, Zhong CP, Zha DJ (2021) Unusual presentation of a first branchial arch fistula with maxillofacial infection: a case report. BMC Surg 21: 306.
6. Work WP (1972) Newer concepts of first branchial cleft defects. Laryngoscope 82: 1581-1593.
7. Saarinen RT, Kolho KL, Pitkäranta A (2007) Cases Presenting as parotid abscesses in children. Int J Pediatr Otorhinolaryngol 71: 897-901.
8. Coppens F, Peene P, Lemahieu SF (1990) Diagnosis and differential diagnosis of branchial cleft cyst by CT scan. J Belge Radiol 73:189-196.
9. Chen MF, Ueng SH, Jung SH, Chen YL, Chang KP (2006) A type II first branchial cleft cyst masquerading as an infected parotid Warthin's tumor. Chang Gung Med J 29: 435-439.
10. Ho S, Lee D, Bluestone C (2000) Imaging quiz case 2. First branchial cleft cyst (FBCC). Arch Otolaryngol Head Neck Surg 126: 796-797.
11. Noel F, Leung A (1987) Frist branchial cleft fistula: case reports and literature review. J Otolaryngol 16: 247-251.
12. Sichel JY, Halperin D, Dano I, Dangoor E (1998) Clinical update on type II first branchial cleft cysts. Laryngoscope 108: 1524-1527.