Unusual presentation of a first branchial arch fistula with maxillofacial infection: a case report

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Abstract

Background: First branchial cleft anomaly (FBCA) is a rare congenital defect that arises due to incomplete closure of the ventral portion of the first and second branchial arches. There are variable complex clinical manifestations for patients with FBCA, which are prone to misdiagnosis and inadequate treatment. FBCAs usually involve the facial nerve with a consequent increased risk of facial nerve damage. Here, we present an unusual case of FBCA presenting with two preauricular pits in association with an abnormal maxillofacial cyst.

Case presentation: A 10-month-old girl presented to our department due to recurrent maxillofacial infections accompanied by swelling or abscess of the left cheek and purulent discharge from the preauricular pit for 4 months. A 3D-computed tomography (CT) fistulogram and magnetic resonance imaging (MRI) revealed two conjunctive tract lesions: one tract arose from the skin surface anteroinferior to the external auditory canal (EAC), through the deep lobe of the left parotid, and anteriorly extended to the left masseter; the other extended from the superficial lobe of the left parotid to the intertragic notch. After the maxillofacial infection was controlled by intravenous antibiotic administration, surgery was performed. Intraoperative tools, such as facial nerve monitors, microscopes, and methylene blue dyes, were used to facilitate the complete dissection and protection of the facial nerve. On follow-up over one year, the patient recovered well without facial palsy or recurrence.

Conclusion: FBCA with maxillofacial cysts is rare and prone to misdiagnosis. Physicians should pay attention to this anatomic variant of FBCA with the fistula track located deep inside the facial nerve and projected medially to the masseter.

Keywords: First branchial cleft anomalies, Facial nerve, Maxillofacial, Case report

Background

First branchial cleft anomalies (FBCAs) are rare congenital defects that account for less than 8–10% of all branchial cleft anomalies and arise due to incomplete closure of the ventral portion of the first and second branchial arches [1, 2]. A number of classification systems have been developed in an attempt to assist preoperative assessment and surgical planning for FBCAs. Work’s classification is the most common system based upon both anatomical and histological features. According to this classification, FBCAs are divided into two distinct types: type I has only ectodermal components and is usually superficial to the facial nerve and lies in close proximity to the ear; type II has ectodermal and mesodermal components, often lying medial to the facial nerve and communicating with the external auditory canal (EAC) [3]. Olsen et al. [4] also introduced a classification of defects into cysts, sinuses, or fistulas based on the number of surface openings present. To determine the relationship between FBCAs and the facial nerve prior to surgery, Liu et al. introduced a new subclassification for type II FBCAs into three subtypes based on...
MRI findings [2]. A common location of FBCAs is the area from the EAC to the level of the hyoid bone. The opening of the fistula of FBCAs is typically located in the periauricular area, but more rare locations have also been reported, such as the EAC, middle ear cleft, postauricular region, and even within the neck over the angle of the mandible [5]. Usually, FBCAs have a close anatomical relationship to the facial nerve owing to their embryologic origin. D’Souza et al. [6] performed a comprehensive review of the literature and found that FBCAs had diversiform patterns with the facial nerve; they can be lateral to, medial to, or between branches of the facial nerve. Patients presenting at a younger age were more likely to have a deep tract with a consequent increased risk of facial nerve damage. Thus, treatment for FBCAs is complicated by both variable tract lesions and the complex anatomical relationship to the facial nerve. In this article, we describe a paediatric case of an exceptional type II FBCA presenting with two preauricular pits in association with an abnormal maxillofacial cyst and a tract that passes into the deep lobe of the parotid gland and extends anteriorly to the masseter.

Case presentation

We obtained written informed consent from the patient’s parents. The case is compliant to the SCARE guidelines [7]. A 10-month-old girl was taken to our hospital by her parents due to recurrent maxillofacial infections accompanied by swelling or abscess of the left cheek and purulent discharge from the preauricular pit for 4 months. On physical examination, a swollen erythematous maxillofacial lesion was observed in the left cheek (Fig. 1). A pit with white purulent secretions in the intertragic notch and another small cutaneous dimple in the left parotid gland region were observed (Fig. 1A). EAC contained no fistula track. All other head and neck examinations were unremarkable. Ultrasound performed for the maxillofacial area demonstrated a lesion with inflammatory changes. The other auxiliary examinations, including oto-scopy, pure tone audiometry and renal ultrasound, were without abnormalities. There was no history of previous incision or drainage procedures. No particular family history was recorded. A 3D-computed tomography (CT) fistulogram and magnetic resonance imaging (MRI) were performed to delineate the course of the tract and the extent of the lesion. The results revealed two conjunctive tracts: one tract arose from the skin surface anterior to the EAC, beneath the facial nerve, and passed into the deep lobe of the parotid gland and projected to the masseter; the other tract extended from the superficial lobe of the left parotid to the intertragic notch (Figs. 2 and 3A–C).

Originally, the infection could be controlled by intravenous antibiotic administration, which resulted in maxillofacial infection improvement (Fig. 1B). However, recurrent inflammation had been persisting for one year. Finally, after obtaining parental consent, the patient underwent surgical management under general anaesthesia at 2 years old. The surgery was performed under facial nerve monitoring. First, a methylene blue staining

![Fig. 1](image-url) First branchial cleft anomalies presenting as two preauricular pits associated with an abnormal maxillofacial cyst. One pit with purulent secretions is in the intertragic notch (red arrow), and another small cutaneous dimple (black arrow) is shown in the left cheek region. The maxillofacial infection with abscess resolved with conservative treatment. A The black arrow indicates the cutaneous dimple before treatment, and B the black arrow shows the same region post-treatment.
agent was injected into the cutaneous pit of the intertragic notch. Then, a modified Blair incision encompassing the pit was made. The facial nerve was identified first, which allowed the cyst to be incised easily. During superficial parotidectomy, one cyst lying in the superficial lobe of the parotid and cartilage of the intertragic notch were removed. Blunt dissection was then used to follow the other larger fistula tract, which was located deep inside the facial nerve and projected medially to the posterior aspect of the masseter (Fig. 4A). Using a microscope, the tract was carefully separated from the facial nerve, which formed a blind pouch alongside the masseter (Fig. 4B). Finally, a suction drain was placed and kept until 48 h postoperatively (Fig. 4C). The patient recovered well, and no complications occurred in the following days. The final pathology of the excised tissue was consistent with that of type II FBCA (Fig. 4D). At follow-up over one year postoperatively, the surgical site was well healed, and the patient had no facial palsy or recurrence by coronal and axial MRI imaging (Fig. 3D–F).

Discussion and conclusions
Branchial anomalies arise when the branchial arches and their associated clefts or pouches fail to regress or develop normally [8]. FBCAs are relatively uncommon, and there are various clinical manifestations in patients. As shown in Table 1, FBCAs can appear at any age, and the tract may extend to the osteocartilaginous junction of the ear canal, Eustachian tube, submandibular gland, pharyngeal cavity, and digastric muscle [1, 9–14]. Muranishi et al. [15] also reported a case of FBCA that was clinically typical but occult in images and pathology. In our case, repetitive inflammation around the maxillofacial region was observed. The unusual cord structure beneath the facial nerve, which passed into the deep lobe of the parotid gland, projected to the masseter. This rare case of type II FBCA is prone to misdiagnosis and inadequate...
Fig. 4  Intraoperative findings and surgical strategy. A The facial nerve was identified during surgery. The larger tract (red arrow) projecting medially deep within the temporal trunk of the facial nerve (black arrow) and the smaller tract ending at the cartilage of the intertragic notch (white arrow) are shown. B The black arrow indicates the lesion boundary as a blind pouch lying tightly along the masseter. C A suction drain was placed into the tract tunnel after the first branchial cleft anomaly excision. D A specimen containing cartilage post-removal is shown. The complete fistula was approximately 6 cm in length.

| Author | Date  | Age       | Complaints of presentation                                                                 | Description the course of FBCAs                                                                 |
|--------|-------|-----------|------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------|
| Fastenberg [9] | 2016  | 3-year-old | Pain in the right ear and swelling in the postauricular area                               | A fistula track into the osteocartilaginous junction of the ear canal                            |
| Faruque [10]  | 2019  | 12-year-old | A draining cervical pit                                                                   | Fistulas opening to the Eustachian tube                                                         |
| Watanabe [11]  | 2017  | 14-month-old | Redness and swelling in the left neck area                                                | Fistula extending from a cutaneous opening in the left submandibular area penetrating the submandibular gland, and ending in the pharyngeal cavity |
| Fanous [1]    | 2020  | 6-year-old | Left conductive hearing loss and an ipsilateral painful cervical mass                     | A presumed ear canal cholesteatoma in association with an abnormal bony canal and a pharyngeal cyst |
| Chaouki [12]  | 2021  | 3-year-old | Recurrent right lateral cervical infection                                                | A fistulous path underneath the facial nerve and ends under the digastric muscle                 |
| Roche [13]    | 2016  | 4-year-old | Recurrent left neck abscesses and palpable persistent submandibular swelling             | Duplication of the external ear canal running medial to the facial nerve                          |
| Zhang [14]    | 2020  | 19-year-old | A mass behind the right earlobe and recurrent post-auricular swelling and pain            | The mass originated from the stylomastoid foramen and adhered to the posterior surface of the parotid gland, invading the temporal bone |
| Muranishi [15] | 2020  | 8-year-old | An infectious epidermal cyst                                                             | A cord structure attached to subcutaneous tissue at the intertragal notch, no opening to the external auricular canal |
typical manifestations of BORS are hearing loss, otologic complaints, and may require ear-specific surgery such as tympanoplasty or canaloplasty [27]. Additionally, methylene blue dye injection during surgery may help verify the suspected path of a fistula, and intraoperative microscopy and facial nerve monitoring are indispensable for protecting the facial nerve. These methods have the potential to reduce facial nerve injury and recurrence rates.

In summary, we described an unusual presentation of type II FBCA with maxillofacial infection, and the main fistulous tract travelled medial to the facial nerves into the deep lobe of the parotid gland and extended anteriorly to the masseter. Surgical excision should be performed after the resolution of any infection as it is important for identification of the facial nerve in almost all cases, especially in young patients. Intraoperative tools, such as facial nerve monitors, microscopes, and methylene blue dyes, facilitate the complete dissection and protection of the facial nerve.

Abbreviations
FBCA: First branchial cleft anomaly; FBCAs: First branchial cleft anomalies; EAC: External auditory canal; MRIs: Magnetic resonance images.

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Authors’ contributions
YH was responsible for writing the paper, RY collected and analyzed the clinical data, LH revised and edited the manuscript, CZ took part in following up the patient and DZ took responsibility for the integrity of the content of the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials
Not applicable.

Declarations
Ethics approval and consent to participate
Ethical approval Ethical approval was given by the medical ethics committee of the First Affiliated Hospital of the Air Force Medical University under number KY20212022-C-1.

Consent for publication
Written informed consent was obtained from the patient’s parent.

Competing interests
The authors declare no competing interests.

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