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CASE REPORT

A Rare Extension of First Branchial Cleft Fistula to the External Auditory Canal

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ABSTRACT

Anomalies of the first branchial cleft are rare congenital malformations of the head and neck, making up 8% of all branchial cleft anomalies. Presented here is a case of a 41-year-old female patient with a 30-year history of mild swelling inferior to the left lobulus auriculae, who complained of discharge from the left ear which responded to external otitis media treatment. She presented with discharge from a fistula opening on the posteroinferior aspect of the left mandibular angle of one-year duration. Following extensive workup and surgical exploration, it was discovered that the fistula tract traversed across the midline of the neck, deep to the main trunk of the facial nerve with an opening at a point anteroinferior to the left external acoustic meatus. The main trunk of the facial nerve and it branches were first isolated, followed by complete excision of the fistula tract along with surrounding skin and cartilage which formed part of the external acoustic meatus. The patient did not show any signs of postoperative facial weakness. In spite of their rarity and the difficulties associated with making a correct diagnosis, anomalies of the first branchial cleft should always be considered in the differential diagnosis, particularly in patients with ear discharge resistant to treatment despite the presence of an intact ear drum.

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Introduction

Anomalies of the first branchial cleft are rare congenital malformations of the head and neck, accounting for less than 8% of all branchial cleft anomalies[1,2]. There are more than 200 reported cases in the literature[3]. Although such anomalies are congenital in origin, patients usually present at a later age (mean age 18.9 years) with a variety of complaints, most commonly periauricular swelling[1,4,5]. Patients are often misdiagnosed, while iatrogenic nerve injury and recurrence after treatment are not uncommon. Definitive treatment requires complete surgical removal of the tract[6].

Anatomically, an anomaly of the first branchial cleft may be classified as a fistula (tract with two openings), a sinus (tract with one opening) or a cyst (tract without an opening) [7]. These are most commonly present as fistulas (52%) followed closely by sinuses (48%). Presentation as a cyst is rare[1,6].

Lower fistula tract openings usually occur in an area bordered by the sternocleidomastoid muscle posteriorly, the mandibular angle anteriorly, and the hyoid bone inferiorly. Upper fistula tract openings, on the other hand, generally occur inferior to the external acoustic meatus. Sinuses also drain to the same areas[6]. Furthermore, fistula tracts generally extend deep to the facial nerve compared to sinus tracts which traverse lateral to the nerve[1,8]. Due to this close association with the facial nerve, accurate diagnosis and treatment requires wide surgical exploration and/or facial nerve monitorization[2,8,9].

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Case Summary

A 41-year-old female patient presented to the ENT outpatient clinic with discharge from a small depression on the neck, posteroinferior to the left mandibular angle which she claimed to have had since birth. At the age of 9 years, she received treatment for external otitis after presenting with discharge from the left ear which ensued soon after a mild swelling developed on her neck. Although her symptoms resolved at the time, she presented to our clinic with discharge from her neck of one-year duration. Physical examination revealed the presence of a fistula opening posteroinferior to the left mandibular angle. Methylene blue injection by inserting a catheter through this opening helped to localize the other opening at a point lateral to the junction between the anteroinferior aspect of the left external acoustic meatus and adjacent cartilage. The dimensions of the fistula tract were determined using a fistulogram (Figure 1).

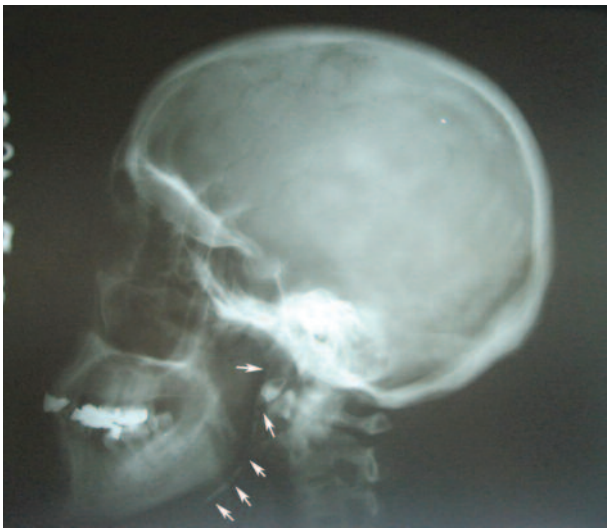

Figure 1. A fistulogram showing a fistula extending towards the left external acoustic meatus from a point on the neck, posteroinferior to the left mandibular angle (white arrows: fistula tract).

Surgery was scheduled for excision of the fistula tract. Citing a possible close association between the fistula tract and the facial nerve, a modified Blair incision was preferred to allow for wide surgical exploration which confirmed the presence of a 4-mm wide fistula tract starting from a point posteroinferior to the left mandibular angle, extending deeper than and in close proximity to the main trunk of the facial nerve, with its other opening at a point lateral to the junction between the anteroinferior aspect of the external acoustic meatus and adjacent cartilage. The main trunk of the facial nerve and its branches were first isolated, followed by complete excision of the fistula tract along with surrounding skin and cartilage which formed part of the external acoustic meatus (Figure 2). The patient did not show any signs of postoperative facial weakness.

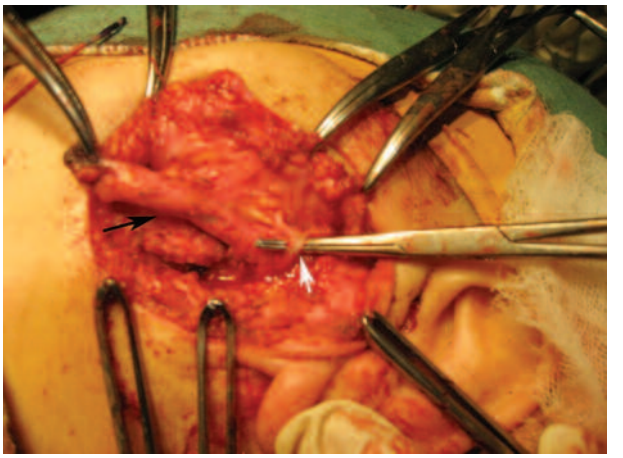

Figure 2. During surgery, the fistula tract is seen to run below the main trunk of the facial nerve, extending towards the external acoustic meatus. (White arrow: main trunk of the facial nerve; black arrow: fistula tract).

Histopathological examination of resected material revealed the fistula wall to be lined by stratified squamous epithelium with underlying skin appendages and an isolated cartilage islet (Figure 3).

Discussion

By the fourth week of human embryogenesis, 6 pairs of branchial archs develop which later form the lower part of the face and neck. Archs are made up of 5 external clefts (ectoderm) and 5 internal pharyngeal pouches (endoderm). By week 7 of development, the archs disappear. The structures which are derived from the first branchial cleft are cavum conchae, external acoustic meatus and external layer of the tympanic membrane. Anomalies of the first branchial cleft occur as a result of incomplete closure of the ventral parts of the first and second branchial archs[2].

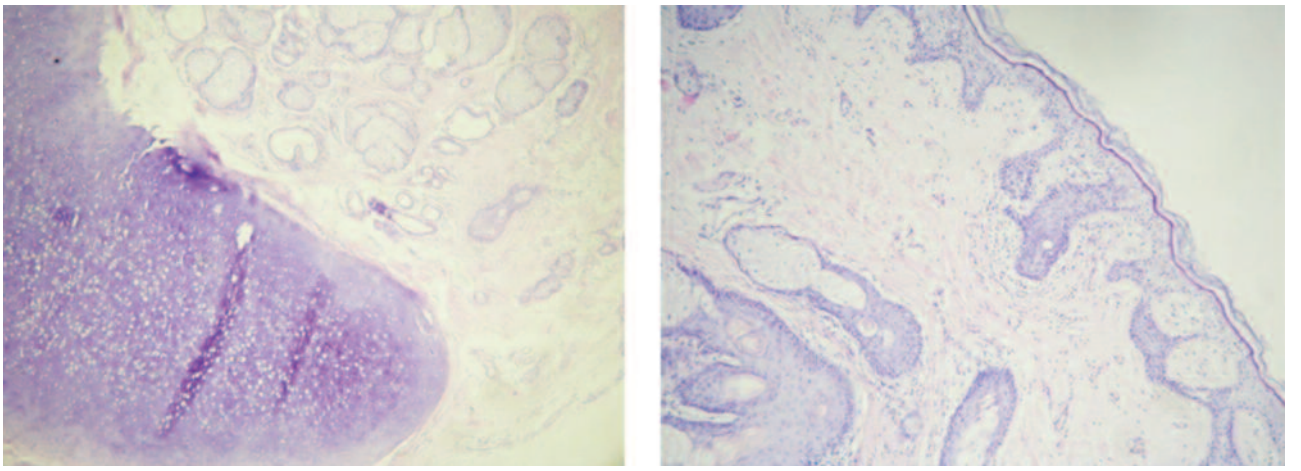

Figure 3. Microscopic image of excised tissue showing the wall of the fistula lined by stratified squamous epithelium. Skin appendages and isolated cartilage islet can be seen underneath the epithelium (Left slide: Hematoxylin and Eosine staining, magnification x40; right slide: Hematoxylin and Eosine staining, magnification x100).

In 1971, Arnot described an anatomic classification for these anomalies[10]. Defects of the parotid area which manifest in early adulthood were classified as type 1, whereas defects of the childhood era which result in the development of a tract between the anterior cervical triangle and the external acoustic meatus were considered type 210. A histological classification was introduced by Work in 1972[11]. Based on this classification, type 1 defects are of ectodermal origin, which occur as a result of duplication of the membranous external acoustic meatus. This type generally presents in early adulthood as a cyst with a squamous epithelial lining and without external connections. Type 2 lesions, on the other hand, are ectodermal and mesodermal in origin containing adnexial tissue like skin and cartilage. These generally occur in early childhood as a fistula or sinus that extends from the submental triangle upwards towards the parotid area, opening into either the cartilage or the bone-cartilage junction of the external acoustic meatus[11].

Facial malformations rarely accompany anomalies of the first branchial cleft, which makes the diagnosis more challenging[1]. Such anomalies may occur with other otologic conditions. Yalcin et al. reported a case with concomitant cholesteatoma and aural atresia[4].

Triglia et al. reported on a mean period of 3.5 years between manifestation of symptoms and access to appropriate treatment, with 50% of patients having prior history of unsuccessful treatment[6]. Treatment involves complete surgical excision of the tract. Due to a close proximity to the facial nerve, wide surgical exploration and/or facial nerve monitorization is sometimes required[2,8,9]. Surgical intervention based on a wrong diagnosis increases the risk of iatrogenic facial nerve injury. A detailed history and a careful physical examination (cervical, parotid, auricular) are essential for timely and accurate diagnosis. Examination of the external acoustic meatus may reveal the presence of a fistula opening or a web on the eardrum6. For many years our patient presented to numerous centers with ear discharge, repeatedly receiving medical treatment for otitis externa.

Fistula tracts generally tend to extend at a level deeper than the facial nerve, while sinus tracts traverse lateral to it[1,8]. The fistula tract in our patient, which was type 2 both histologically and anatomically, was 4-mm wide and ran deep to the main trunk of the facial nerve.

Radiological modalities such as computed tomography (CT) or fistulography may be utilized to confirm a diagnosis. While a fistulogram helps to differentiate between fistulas and sinuses, CT may assist in

detecting a tract surrounded by cartilage adjacent to the external acoustic meatus[2,12].

In spite of their rarity and the difficulties associated with making a correct diagnosis, anomalies of the first branchial cleft should always be considered in the differential diagnosis when evaluating lesions of the cervical, parotid and auricular areas. Particular care should be taken in patients with ear discharge resistant to treatment despite the presence of an intact ear drum. Although this disorder is generally encountered in children, some patients are diagnosed in adulthood. Early diagnosis and treatment are essential for the prevention of recurrent infections and/or the development of a secondary fistula tract. Complete excision of the tract, particularly one that traverses the external acoustic meatus, may require removal of surrounding skin and cartilage islets. A wide surgical field should be established to help preserve of the facial nerve.

Conflict of interest/Funding

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References

1. D'Souza AR, Uppal HS, De R, et al. Updating concepts of first branchial cleft defects: a literature review. Int J Pediatr Otorhinolaryngol 2002; 62:103–09.

2. Stokroos RJ, Manni JJ. The double auditory meatus. A rare first branchial cleft anomaly: clinical presentation and treatment. Am J Otol 2000; 21:837-41.

3. Wittekindt C, Schöndorf J, Stennert E, Jungehülsing M. Duplication of the external auditory canal: a report of three cases. Int J Pediatr Otorhinolaryngol 2000; 58:179-84.

4. Yalcin S, Karlidag T, Kaygusuz I et al. First branchial cleft sinus presenting with cholesteatoma and external auditory canal atresia. Int J Pediatr Otorhinolaryngol 2003; 67:811–814.

5. Elsurer C, Yilmaz T. First branchial cleft fistula presenting with external opening on earlobe. Eur Arch Otorhinolaryngol 2006; 263: 685–687.

6. Triglia JM, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN. First branchial cleft anomalies: a study of 39 cases and a review of the literature. Arch Otolaryngol Head Neck Surg 1998; 124:291-5.

7. Olsen KD, Maragos NE, Weiland LH. First branchial cleft anomalies. Laryngoscope 1980; 90:423-36.

8. Solares CA, Chan J, Koltai PJ. Anatomical variations of the facial nerve in first branchial cleft anomalies. Arch Otolaryngol Head Neck Surg 2003; 129:351-5.

9. Isaacson G, Martin WH. First branchial cleft cyst excision with electrophysiological facial nerve localization. Arch Otolaryngol Head Neck Surg 2000; 126:513-6.

10. Arnot RS. Defects of the first branchial cleft. S Afr J Surg 1971; 9:93-8.

11. Work WP.Newer concepts of first branchial cleft defects. Laryngoscope 1972; 82:1581-93.

12. Sichel JY, Halperin D, Dano I, Dangoor E. Clinical update on type II first branchial cleft cysts. Laryngoscope 1998; 108:1524-7.