Nonsyndromic Complete Second Branchial Cleft Fistulas: A Clinicosurgical Experience

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ABSTRACT

Branchial cleft anomalies are congenital malformations resulting from the abnormal persistence of branchial clefts during embryogenesis, often presenting as cysts, sinuses, or fistulae. These anomalies account for approximately 20% of pediatric head and neck masses, with bilateral cases being particularly rare. This report discusses a case of an 11-year-old boy with complete bilateral second branchial cleft fistulae, characterized by intermittent mucopurulent discharge and recurrent inflammation, primarily following upper respiratory infections. Diagnostic imaging, including CT scans, confirmed the presence of bilateral fistulous tracts. Surgical management involved a combined transoral and transcervical approach, including bilateral tonsillectomy and complete excision of the fistulous tracts. The procedure resulted in successful removal of the anomalies without postoperative complications. This case emphasizes the importance of thorough diagnostic evaluation and the effectiveness of complete surgical excision in managing bilateral branchial cleft anomalies to prevent recurrence and improve patient outcomes. Further research is warranted to optimize surgical techniques and postoperative results.

KEY WORDS

Congenital neck mass, Excision, Second branchial cleft fistula

INTRODUCTION

Branchial cleft anomalies are rare, congenital malformations that result from the abnormal persistence of branchial clefts during embryogenesis and manifest clinically as cysts, sinuses, or fistulae.[1-3] These anomalies are the second most common pediatric congenital head and neck masses,accounting for approximately 20% of cases.[4] Among these,second-cleft lesions comprise 90-95% of all branchial cleft anomalies.[5] Bilateral anomalies account for approximately 1% of all cases and bilateral second branchial cleft fistulae are much rarer. There have been less than ten cases of non-syndromic, bilateral second branchial cleft fistulae reported in the literature.[6]

Patients commonly present within first two decade of life with slight female preponderance.[7] Patient present with persistent or intermittent mucoid or mucopurulent discharge from an opening in the lateral aspect of the neck following mostly after an upper respiratory tract infection.[7] Branchial fistulas are lined with epithelial tract and a complete second branchial fistula has an internal opening at the tonsillar fossa and an external opening at the skin in the line of the anterior border of the lower third of the sternocleidomastoid(SCM).[8] Diagnosis is mostly clinically along with contrast fistulogram, Computed tomography (CT) scan with intravenous contrast and magnetic resonance imaging (MRI) of neck providing radiographical benefit.[7] Treatment of choice is complete surgical excision of the fistulous tract by using combined transoral and transcervical approach.[8] Here we present a case with complete bilateral second branchial cleft fistulae with intermittent pain,swelling and discharge on anterior neck with imaging characteristics following complete excision bilaterally.

CASE REPORT

A 11 year old boy with an unremarkable perinatal and medical history presented to our Out-patient Department with the complaint of intermittent mucopurulent discharge

from the anterior aspect of the both sides of neck and recurrent attacks of inflammation following an attack of upper respiratory tract infection. There were episodes of acute infection of the fistulas which resolved upon the complete course of antibiotics and with no significant family history.

On examination, a small punctum was seen at mid third of the neck at the medial border of sternocleidomastoid muscle in right side and about the junction of the upper 2/3 and lower 1/3 of the anterior border of the sternocleidomastoid muscle on the left side (Fig. 1). There was a scanty mucoid like discharge from both cervical fistulous openings with no signs of inflammation around it. Oropharyngeal examination revealed bilaterally enlarged grade 2 tonsils according to Friedman grading. Fibreoptic nasopharyngolaryngosopic examination was unsignificant. In the view of possible association with Branchio-oto-renal syndrome, there was normal renal ultrasound findings ruling out hypertrophic or atrophic kidneys. External ear and its surrounding area was grossly normal, and audiometric testing ruled out any kind of hearing loss in the view of Branchio-oto-renal syndrome. There was no any craniofacial deformities as well.

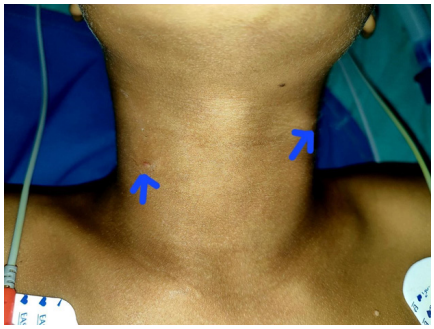

Figure 1. Pre-operative image showing external opening in bilateral anterior neck in medial aspect of sternocleidomastoid.

A computed tomography (CT) sonogram was performed which showed contrast filled small, linear, 4-5 mm wide soft tissue tract beginning in the region surrounding the left tonsillar fossa and extending along the anterior carotid sheath and anterior border of the SCM down to the skin surface near the level of thyroid and similar findings on right side (Fig. 2). An excision was then performed under general anaesthesia with bilateral tonsillectomy done initially and transverse elliptical incision made around cervical fistulous opening after instilling methylene blue into the opening, dissected deep to subplatysmal plane and continuing superiorly, tunneling under subplatysmal layer following the tract with second transverse incision was made just above the level of hyoid in stepladder manner to facilitate further superior dissection (Fig. 3). The tract was followed over hypoglossal nerve, between the carotid and deep to digastric muscles till within the internal opening at posterior pillar of ipsilateral tonsillar fossa and tract was completely excised and delivered out with similar

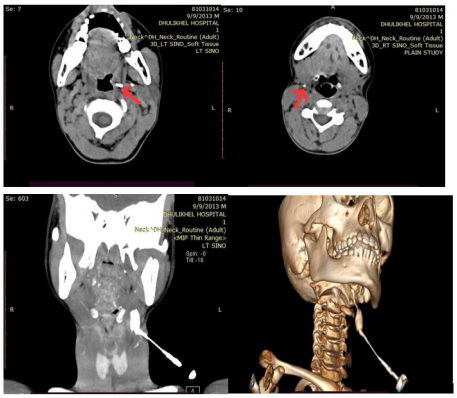

Figure 2. Upper left and upper right-CT scan images in which red arrow pointing towards the fistulous tract inserting into the tonsillar fossa.
Lower left- Fistulous tract Lower right-CT scan 3D reformation

Figure 3. Neck stepladder incision and removed specimen of left and right branchial fistula

procedure done on opposite side (Fig. 4). The patient was discharged with no post-operative issues and no suspicious findings on histopathological examinations.

Figure 4. Upper- Tract showing blue dye opened in the tonsillar fossa post tonsillectomy.
Lower- A complete fistula tract starting from tonsillar fossa to skin overlying anterior neck.

DISCUSSION

Brachial apparatus was first described by Von Baer and its anomalies were first described by Von Ascheron.[9] The branchial apparatus consists of six pairs of mesodermal-derived arches, which are lined internally by endodermal-derived pouches and externally by ectodermal-derived clefts.[2] During embryonic development, as the second arch grows caudally, it fuses with the 3rd and 4th arches to form a deep groove before joining the skin as an external opening, thereby being termed cervical sinus. Persistence of this fistula is often due to the breakdown of endoderm during embryogenesis, and this forms a tract to the skin at the junction of lower 1/3rd and upper 2/3rd of the anterior border of the sternocleidomastoid of the affected neck.[10]

In greater than 95% of cases, branchial cleft anomalies originate from remnants of the second branchial cleft.[11] Anget et al. in his literature review of branchial cleft fistulae found that complete second branchial cleft fistulae are rarely encountered.[12]

In a large series conducted by Chionh et al. about 15% of cases presented in children aged < 10 years, while the majority of cases presented at the age range of 10-40 years.[13] Symptoms include intermittent mucopurulent discharge from the anterior aspect of neck and recurrent attacks of inflammation following an attack of upper respiratory tract infection which was similar to our case. Incision and drainage may be needed for cellulitis or abscess formation on rare occasions.[7] Occasionally, bilateral branchial cleft fistulae may be associated with branchiootorenal syndrome, for example, is characterized by branchial anomalies, auricular malformations), hearing impairment and renal malformations) with our paient not associated with it.[14]

The diagnosis of a branchial cleft fistula can be established by a detailed history and physical exam based on clinical signs that have been described in-depth previously. However, various imaging methodologies such as fistulography, CT, or MRI can assist with confirming the diagnosis as well as defining the extent of the lesion.[2,15] Therefore, the extent of a fistulous tract is best made intra-operatively when muscle relaxants are given, and the tract can be traced using a dye. Special considerations should be also given to clinical differentials, such as malignancies and tuberculous fistulas, which often present in the neck. Complete surgical excision of the branchial fistula remains the cornerstone of treatment.[16] In this case, a bilateral tonsillectomy with bilateral stepladder approach was employed in the management of this case for the purpose of better surgical access. This method was associated with good surgical outcome. The management of bilateral branchial fistulas follows the same principals of unilateral fistula or sinus tract excision wherein complete tract excision is mandatory to prevent recurrent symptoms.[17] Surgical excision through trans cervical approach either by step ladder or longitudinal incision and combined pull through technique is the treatment of choice. The external approach alone may lead to recurrence of the tract due to incomplete excision compared to the combined approach.[18]

Further study needs to be done on the excision technique of a complete bilateral branchial fistulae with or without tonsillectomy and also for improvement of post operative cosmetic outcome.

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