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Case Report

Early presentation of complete branchial fistula with review of literature

Shilpi Arora*

Department of Otorhinolaryngology, General Hospital Palwal, Palwal, Haryana, India

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***Correspondence:**
Dr. Shilpi Arora,
E-mail: doctor_shilpi@yahoo.co.in

ABSTRACT

Branchial anomalies are rarely seen in clinical practice. They arise as a result of aberrations of the branchial apparatus during fetal development. Though present since birth, they are usually diagnosed in the first or second decade of life. Second arch anomalies are by far the most common and present as a neck mass or discharging sinus that may be complicated by infection. Clinical examination often reveals the lesion to be related to the junction of the upper two thirds and the lower one third of the sternocleidomastoid muscle. Branchial fistulas often present as a discharging sinus in the neck. The fistulous tract may extend for a variable distance within the neck. However complete branchial fistula of the second cleft with an internal opening in the tonsillar area and an external opening in the skin is rare. Surgical excision using the step ladder approach was used. No recurrence was seen at one year follow up. The incidence of such lesions presenting within first two years of age is extremely rare. Surgical excision is the treatment of choice for branchial anomalies. We present the case of a two-year-old female patient with complete branchial fistula and discuss the clinical presentation and surgical management of such lesions, while reviewing the relevant literature.

Keywords: Branchial fistula, Step ladder incision, Sternomastoid muscle

INTRODUCTION

Brancial apparatus was first described by Von Baer while von Ascheroni described their anomalies.[1] The branchial arches serve as precursors to the structures found in the face, neck and pharynx and develop around the 4-7 weeks of gestation.[2] Branchial arch anomalies form due to failure of obliteration of the branchial clefts.[2]

Of the six pairs of branchial arches, the second arch grows caudally and covers the second, third and fourth branchial clefts. The cervical sinus of His is formed by the fusion of the second arch with the enlarging epipericardial ridge of the fifth arch.[3] The fifth pharyngeal arch is usually rudimentary, or disappears and the sixth arch is often represented as part of the fourth arch due to its small size.[1-5] Depending on the anatomic location, branchial anomalies have been classified into first, second, third and fourth arch anomalies. Of these 95% of the arch anomalies arise from the Second branchial arch, 1-4% cases from the first, with third and fourth branchial arch anomalies being extremely rare.[2-6]They may present as cysts, sinus tracts, fistulae or cartilaginous remnants. Cysts are the entrapped remnants of branchial clefts; sinuses are remnants of clefts or pouches; and fistulae can result from persistence of both pouch and cleft.[2,7,8] Branchial cysts present as 80% of the cases and the remaining 20% emerge as sinuses, fistulas, or cartilage remnants.[9]

Branchial fistulas are uncommon anomalies. True or complete brachial fistulas of the 2nd arch, with an internal opening at the tonsillar fossa and an external opening overlying medial aspect of sternocleidomastoid are rare, as most branchial fistulas present in an incompleteforms.[10-12] There is equal preponderance in both males and females. The 96-97% of these anomalies

are unilateral and present in the first or second decade of life.

Clinically the lesions present as a small punctum in the lower neck, anterior to sternomastoid muscle, with a history of recurrent discharge. Complete excision of the fistulous tract is the gold standard for treatment for these lesions.[13]

CASE REPORT

A two year old female patient was brought with the chief complaints of a small opening in lower part of the right side of her neck since birth.

There was a history of recurrent mucoid to mucopurulent discharge from the opening which was relieved with oral antibiotics. There was no family history of a similar diagnosis.

Examination revealed a healthy child with no craniofacial deformities. A punctum was seen at the junction of middle and lower one third of anterior border of the right sternocleidomastoid muscle (Figure 1). There was a scanty mucoid-like discharge from the cervical opening. Oropharyngeal examination and renal ultrasound abdomen was normal.

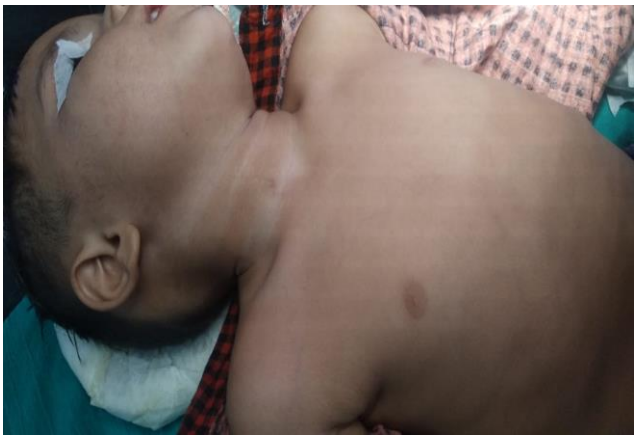

Figure 1: Preoperative picture of patient showing the fistula.

CECT neck showed a fusiform cystic lesion along anterior border of lower part of sternomastoid muscle extending upto and communicating with the tonsillar fossa (Figures 2-4).

On the basis of clinical history and physical examination a diagnosis of second branchial cleft fistula was made. Anesthetic fitness was taken and patient was posted for total excision of fistula under general anesthesia.

The surgical procedure involved a transcervical approach with stepladder technique for complete excision of tract. An elliptical incision was taken over neck around the fistula opening. Soft tissue dissection was done and continued superiorly tunneling under the subplatysmal layer to follow the path of the fistulous tract. The second incision was made at the level of the hyoid bone (stepladder fashion) to facilitate superior dissection. (Figure 5).

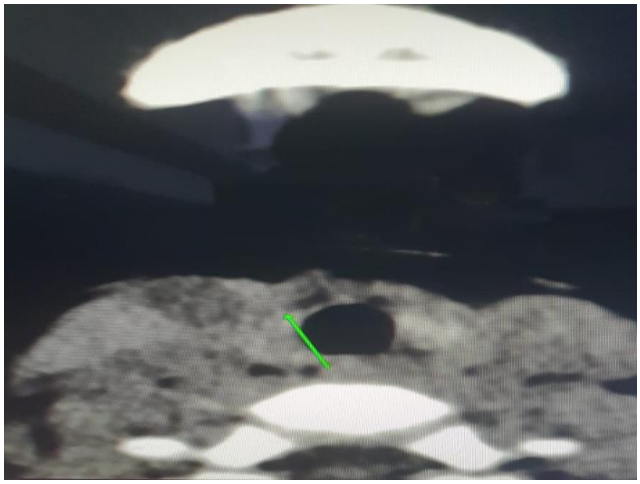

Figure 2: CT scans showing cervical opening of the fistula.

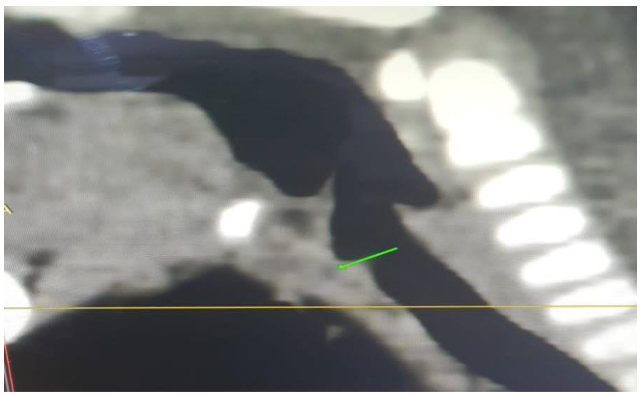

Figure 3: CT scan of pharyngeal opening of the fistula.

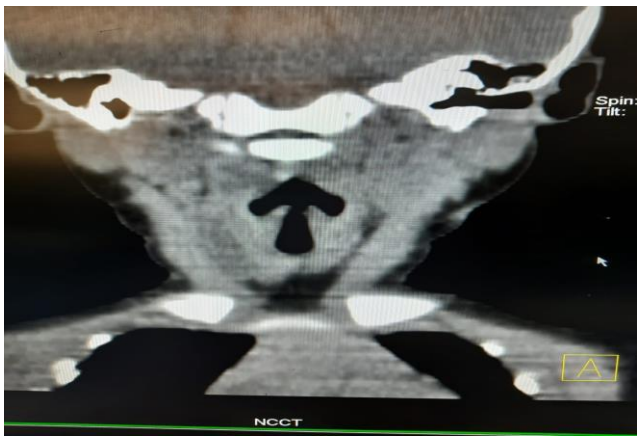

Figure 4: CT scan showing fusiform tract.

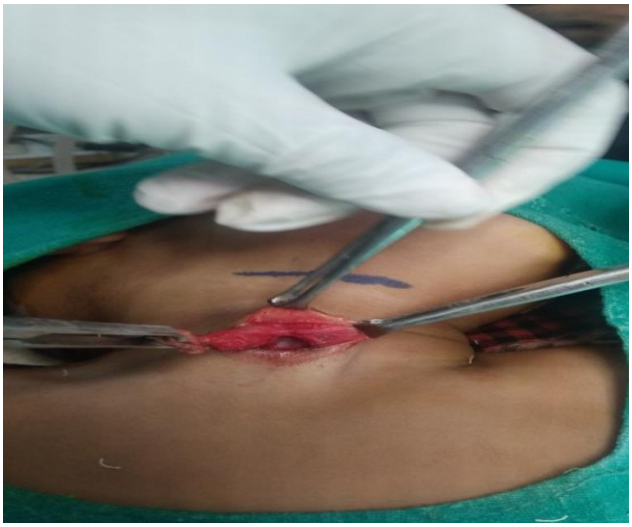

Figure 5: Intra op picture with sinus and stepladder incision.

The tract was followed upto the posterior aspect of the hyoid bone i.e., the level of the tonsillar fossa. Bimanual palpation was done, from the tonsillar fossa orally and from the parapharyngeal space. An indented area was felt in the tonsillar fossa when traction was applied to the tract. The tract was cross clamped, doubly ligated and excised close to the tonsillar fossa. The whole tract was delivered out through the upper cervical incision. The length of the tract was approximately 4.5 cm with a width of 1.2 cm (Figure 6). After achieving hemostasis both neck incisions were closed in layers.

The postoperative period was uneventful and the patient was kept on regular follow up for the one year with no recurrence. Histopathological report confirmed branchial fistula tract lined with tall columnar ciliated epithelium with subepithelium showing dilated congested blood vessels (Figure 7).

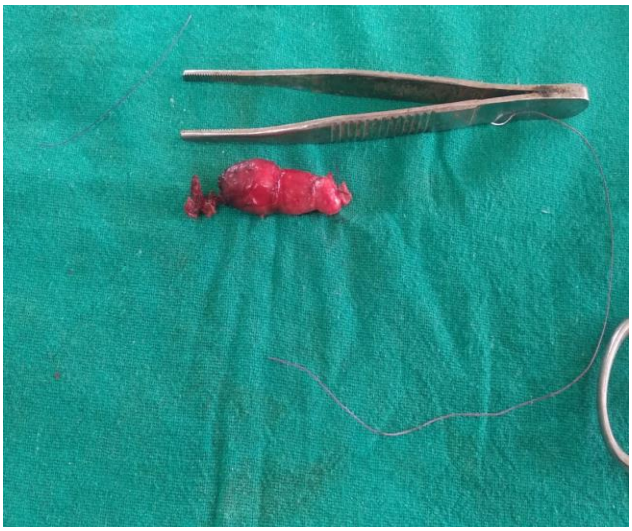

Figure 6: Excised fusiform tact.

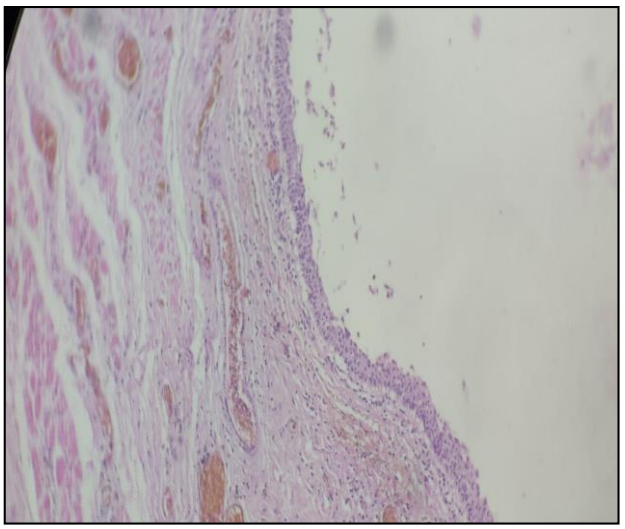

Figure 7: Histopathology-showing tract lined with columnar ciliated epithelium and congested blood vessels.

DISCUSSION

Branchial arches play a role in the formation of face, ear and neck. During embryonic development, the second arch grows caudally and fuses with the 3rd and 4th arches. 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 side.[14]

Among the 2nd arch anomalies branchial cysts are three times more likely than fistulas.[13] The 2nd branchial arch fistula is a rare manifestation of branchial arch anomalies. Complete branchial fistulas with an external and internal opening are rare.[12] Most of the times it is a simple sinus opening that extend up the neck for a variable distance.[11] In our case a unilateral fistula was observed located on the right side of the neck in a two-year-old female. Bajaj in his series reported 50 cases to be unilateral and 12 bilateral.[15] Bilateral branchial fistula and sinuses are rare and only in 6% cases are associated with a family history.[16-18] However, about 2% of sporadic cases may also be associated with an incidence.[19]

Suspicion of a hereditary aetiology should be raised in patients who have bilateral fistulas or preauricular features or a positive family history. These should prompt additional renal and genetic investigation to rule out autosomal dominant conditions such as branchio-otic and branchio-oto-renal syndrome.[12]

Although branchial fistula may occur at any age, they commonly present in first or second decade of life, with an equal gender distribution. Chionh, in his series observed majority of cases presented at the age range of 10-40 years with less than 10 years at presentation in only

15% of cases.[20] Ford et al found 78% of their cases presented by five years of age. There was history of infection and discharge seen frequently and abscesses drainage done for 7%of cases. There was also a right sided preponderance of the lesions.[16] Sampath et al found 70% cases presented above 11 years.[14] Only one patient presented at birth. They also noticed right sided predominance. Second arch anomalies may take several forms; however, malignant lesions are uncommon.[21]

Clinically, cases present as persistent unilateral discharging defect usually at the right side of the neck. In complete fistulas there may be history of drainage on consumptio of fluids.[22] Second branchial cleft anomaly are suspected when fistula or cyst present in the lower anterior or lateral neck. Cysts and fistulas are mostly painless however pain and discharge indicate infection.[15]

Bailey classified the lesions into four types (1929):[23]

Type I-Most superficial and lies along the anterior surface of sternocleidomastoid deep to the platysma, but not in contact with the carotid sheath. Type II-Most common type where the branchial cleft cyst lies anterior to the sternocleidomastoid muscle, posterior to the submandibular gland, adjacent and lateral to the carotid sheath. Type III-Extends medially between the bifurcation of the internal and external carotid arteries, lateral to the pharyngeal wall. Type IV-Lies deep to the carotid sheath within the pharyngeal mucosal space and opens into the pharynx.

In our case report, type I branchial fistula was present deep to platysma, just reaching upto anterior border of sternocleidomastoid muscle.

Though branchial lesions are mostly diagnosed clinically preoperative imaging helps delineate the tract and aid surgical planning. It may also differentiate between sinus and fistula.[13]

Ultrasound of the second branchial cleft cysts shows well-circumscribed, thin-walled and anechoic lesions with compressibility and posterior acoustic enhancement with internal echoes compatible with internal debris.[24]

Fistulograms are commonest investigation and help delineate tract. Sometimes complete tract not be demonstrated as it may be blocked by secretions and granulations.[25]

CT scans show well-circumscribed, low-density cystic masses with a thin wall.[3]

CT fistulogram helps demonstrate the important structures of neck in relation of sinus tract.

MRI is better suited for second branchial cleft cysts with parapharyngeal involvement and for type I first branchial cleft. T1-weighted imaging may turn from low to high signal depending on the proteinaceous content of the cyst while T2-weighted images are typically hyper intense. It provides the relationship of glandular tissue to the mass (e.g., fat planes between the parotid gland and a parapharyngeal mass).[25]

The definitive treatment for branchial fistula is surgical excision, of the sinus tract in its entirety along with the external sinus opening.[26] The approaches described include the transcervical approach, either by a stepladder approach or through a long incision along the anterior border of Sternocleidomastoid muscle.

Stepladder approach described by Bailey in 1933, is the most accepted method, as it provides better visualization near the pharynx. Two incisions in the neck are taken, first, at the external sinus opening and second, the upper incision, made using the hyoid bone as a landmark. This obviates the need for ipsilateral tonsillectomy and also lowers the recurrence rate.[26,27]

Histology often reveals turbid yellow fluid containing cholesterol crystals and stratified squamous epithelium linig.[3]

Complications of surgery include, recurrence, mostly due to incomplete surgical excision, which could be 3% in fresh cases and up to 20% in second surgical attempts.[28] Other complications include secondary infection, injury to nerves (hypoglossal, glossopharyngeal), injury to internal jugular vein and hematoma formation.

In the present study, a bilateral stepladder approach was employed for better surgical access. There was a good surgical outcome and at 1 year follow-up no recurrence was observed (Figure 8).

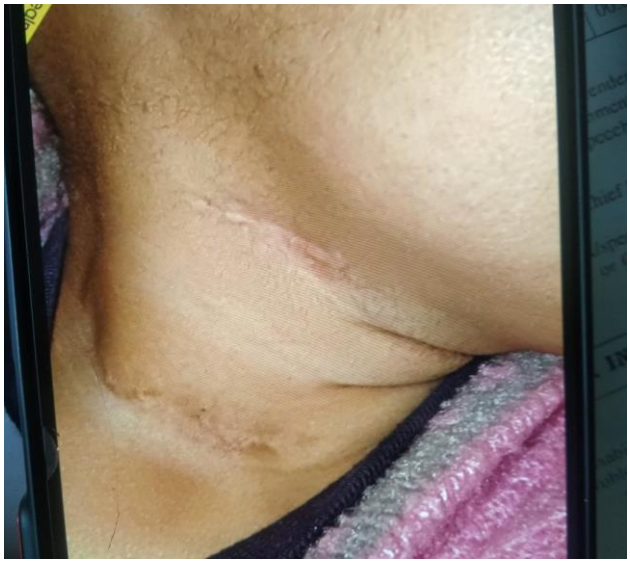

Figure 8: Post op picture (1 year) showing the well healed scar.

CONCLUSION

Complete branchial fistulas are rare. Clinical diagnosis, radiological imaging and complete surgical excision results in definitive cure of the branchial cleft anomalies as these lesions do not regress spontaneously, and they have a high incidence of recurrent infection.

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