Research Article

RECURRENT FOURTH BRANCHIAL ARCH FISTULA – A REVERSE SURGICAL APPROACH

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A R T I C L E I N F O	A B S T R A C T
	Objective: To present a case of fourth branchial arch fistula with five episodes of recurrences with a late diagnosis in second decade of life, which was managed with a non conventional, reverse surgical technique. **Method:** A thorough case report with a review of available literature about fourth branchial arch anomalies is presented. **Result:** Diagnosed with laryngeal endoscopy and barium swallow esophagogram, complete excision of fistula tract was done via a reverse surgical technique. Histopathology revealed Branchial fistula tract with features of chronic inflammation. **Conclusion:** Because of distorted anatomy, intervention in recurrent forth branchial arch fistula needs to be individualized. A reverse surgical approach during a quiescent period comprising complete excision of the fistula tract starting at the piriform sinus end and dissecting downward along the fistula tract upto skin end, combined with transfixation of both ends and division of thyroid isthmus is recommended for treatment of recurrent as well as primary cases for complete removal of disease and preventing recurrence.

INTRODUCTION

Fourth branchial arch anomalies are rare, comprising only about 1-4% of all branchial arch anomalies [1]. Even its existence was questioned before Tucker and Scolnick in 1973 described the first case of fourth branchial cleft anomaly [2]. Moreover, literature is studded with prevalence of fourth branchial arch sinuses, with no documentation of true fistula till date. Most cases manifest in late childhood with recurrent deep neck space infection and/or abscess or acute suppurative thyroiditis [3], almost always on left side [4], but often remains misdiagnosed followed by repeated ventures of incision and drainage. Hence, when it presents as a fistula with fourth branchial arch origin, possibility of pseudofistula cannot be excluded. The complex tortuous developmental model of a true left fourth branchial arch fistula with its consecutive looping of arch of aorta and hypoglossal nerve before reaching skin probably precludes the fistula formation, though theoretically possible.

Case report

A 20 years old male presented in out-patient department with complaint of on and off purulent discharge with pain from a skin opening over left side of front of neck since his 9 years of age.

Patient had history of occasional passage of clear fluid through the skin opening during drinking water, not associated with episodes of coughing or chocking or difficulty in deglutition. There was no history of neck trauma, skin lesion, use of topical medicine. He was non-diabetic, immunocompetent.

The first presentation was a swelling with pain at the same site with purulent discharge, which was treated with incision and drainage in regional government hospital, though no official document was available. He was symptom-free for next 7 years. Then again he developed the same and underwent incision and drainage again in the same hospital. When the symptoms recurred again after one year, on visiting another hospital he was diagnosed as branchial fistula and excision under general anaesthesia was done. But it recurred again after one year, again managed by same procedure. When it recurred again after one year, he presented to our out-patient department.

On examination, a cutaneous fistula with surrounding granulation tissue was seen at the lower end of anterior border of left sternocleidomastoid muscle with purulent discharge with scars around as witness of previous surgical ventures.

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Figure 1 -A cutaneous fistula with surrounding granulation tissue at the lower end of anterior border of left sternocleidomastoid muscle with purulent discharge with surrounding scars.

Oral cavity and oropharynx with both tonsillar fossae revealed no abnormality. Indirect and direct (via fibre optic laryngoscope) laryngoscopy showed a slit like opening in the apex of left pyriform fossa. General and systemic examination were found to be normal.

A barium swallow Esophagogram in erect position shows passage of radio-opaque dye from left pyriform fossa through an irregular, but distinct tract, parallel and anterior to esophagus, reaching upto the skin of front of neck at the level of body of 7th cervical vertebra.

Figure 2a Bariumswallow Esophagogramin erect position lateral view shows passage of radio-opaque dye from left pyriform fossa through an irregular, but distinct tract, parallel and anterior to esophagus, reaching upto the skin of front of neck at the level of body of 7th cervical vertebra.
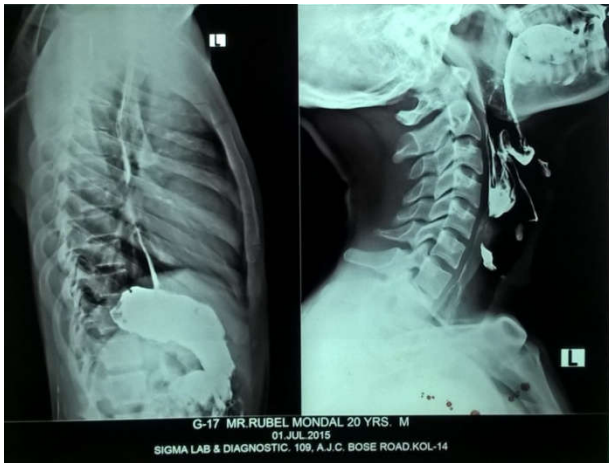

Figure 2b- A barium swallow Esophagogram in erect position Antero-posterior view shows passage of radio-opaque dye from left pyriform fossa through an irregular, but distinct tract, parallel and anterior to esophagus, reaching upto the skin of front of neck at the level of body of 7th cervical vertebra.

CT scan of neck shows a fistulous tract from apex of left pyriform fossa, running along postero-medial to left lobe of thyroid, and through the strap muscles reaching a cutaneous opening surrounded by granulation tissue.
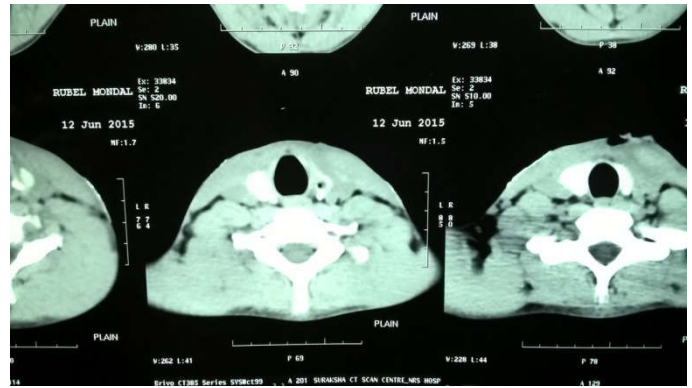

Figure.3a CT scan of neck shows a fistula tract running along postero-medial to left lobe of thyroid.

Provisionally diagnosed as recurrent fourth branchial arch fistula, the treatment strategy taken was another approach – excision of tract in a reverse technique.

Figure 3c CT scan of neck shows a fistula tract reaching a cutaneous opening surrounded by granulation tissue.

Under general anaesthesia, a hypopharyngoscopy was done to pass an infant feeding tube through the opening in the apex of left pyriform fossa.
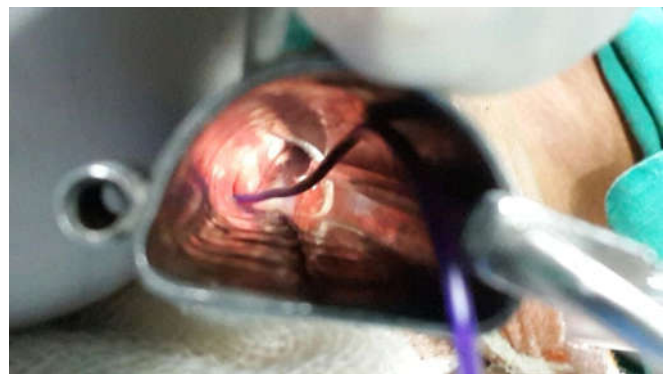

Fig 4 Hypopharyngoscopic picture showing passing of an infant feeding tube through the opening in the apex of left pyriform fossa.

Then patient was kept in position – supine with neck extended, head turned to right. A hockey- stick like incision was given - standard Kocher's neck incision (involving the fistula opening in an ellipse) with extension along the anterior border of left sternocleidomastoid muscle up to upper border of thyroid cartilage.Dissection was forwarded by Platysmal flap elevation and deep fascia dissection. Thyroid cartilage was reached after splitting strap muscles. Inferior constrictor muscle and

perichondrium of thyroid cartilage was retracted to expose the position of pyriform fossa. The upper end of fistula tract identified and dissected downward postero-medial to left lobe of thyroid (sandwiched between isthmus and left lobe of thyroid).
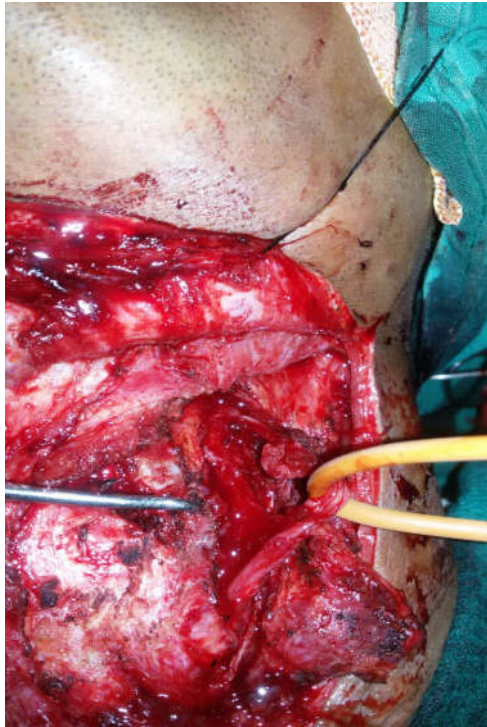

Figure 5 Intra-operative picture showing fistula tract being identified and dissected downward postero-medial to left lobe of thyroid (sandwiched between isthmus and left lobe of thyroid).

Isthmus was secured and divided.

Figure 6 Intra-operative picture showing Isthmus being secured and divided.

Soft tissue dissection was cantinued downward along the tract upto the cutaneous end. Transfixation suture was given in upper end and was excised.
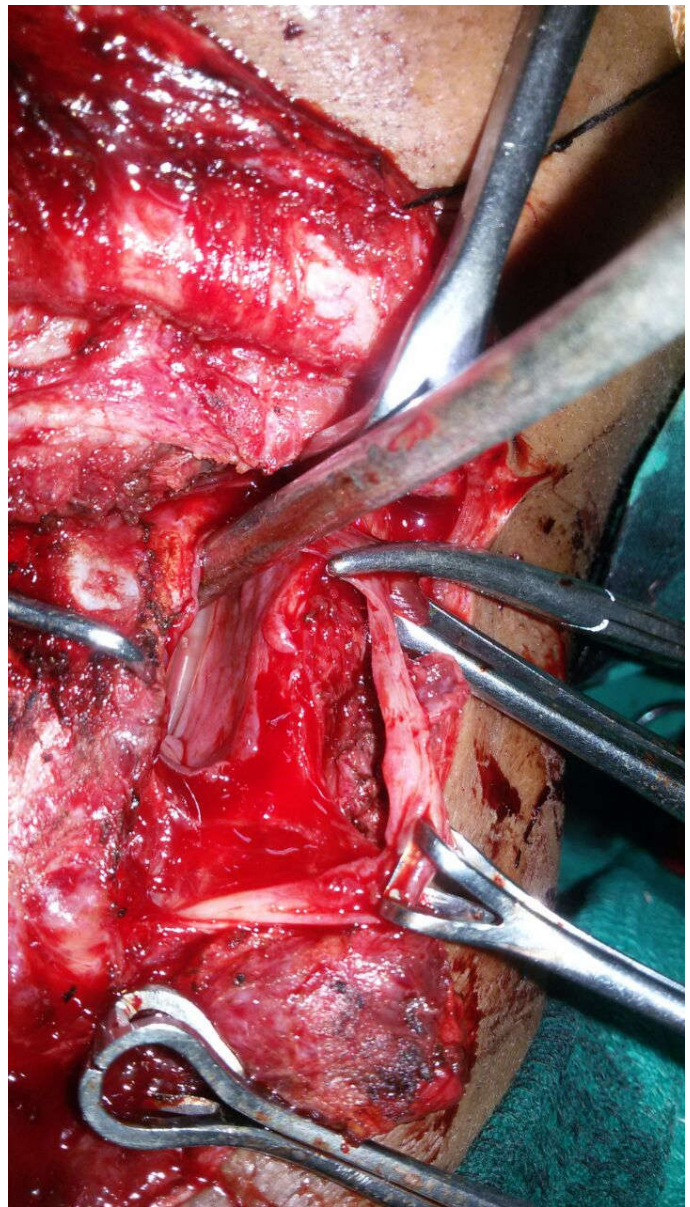

Figure 7- Intra-operative picture showing transfixation suture being given in upper end for excision

Lower end was excised along with the skin with granulation tissues around. pyriform fossa was repaired by extramucosal interrupted sutures. Neck was closed in layers. Post-operative recovery was uneventful.Histopathology report of surgical specimen showed focal area of stratified squamous lining of tract wall with rest of lining ulcerated with necrosis & inflammation. Deeper fibro-collagenous tissue contained dilated & congested blood vessels with sparse, focal chronic inflammatory cells. No granuloma or evidence of malignancy seen - suggestive of residual branchial fistula tract.

On follow up the patient is asymptomatic with no signs of recurrence. Barium swallow Esophagogram in erect position on 14th postoperative day shows absence of any fistula tract.

DISCUSSION

The branchial apparatus develops around the fifth Intrauterine week. It consists of six mesodermal arches with intervening ectodermal invaginations called "clefts" laterally and endodermal invaginations called "Pouches" medially. The fifth arch eventually gets obliterated. By sixth intrauterine week, the second arch grows downward covering the third and fourth

clefts to form cervical sinus of His, which gradually obliterates with development of embryo. Failure of obliteration of this sinus with rupture of interposed mesodermal branchial plate leads to formation of branchial fistula with a skin opening connected internally to an opening in tonsillar fossa, base or apex of pyriform fossa, for involving 2nd, 3rd or fourth arch respectively [3].

The most accepted analysis of route of fourth branchial arch fistula by Liston in 1981 shows that the tract starts at the apex of pyriform fossa, passes downward to leave the pharynx caudal to superior laryngeal nerve, cricothyroid muscle and thyroid cartilage. Then the tract passes downward lateral to trachea and recurrent laryngeal nerve. On the left side, the tract goes further downward, loop anteriorly under the arch of aorta, takes a course upward just in front of common carotid artery, loops over hypoglossal nerve, then descends and opens in skin over lower part of anterior border of sternocleidomastoid in neck. On the right side, the tract follows nearly the same path, except that it hooks around right subclavian artery. Due to this complexity, fourth branchial arch fistula, though theoretically possible, no documentation with a whole tract is found till date [6]. However, iatrogenic or spontaneous rupture of abscess may lead to formation of a pseudofistula [7].

Our case presented with a discharging fistula over left side of front of neck since 9 years of age, but diagnosed as fourth branchial arch fistula after 11 years. Most cases manifest in late childhood with recurrent deep neck space infection and/or abscess or acute suppurative thyroiditis [3], almost always on left side [4], but often remains misdiagnosed followed by repeated ventures of incision and drainage. The intervals from the onset of symptoms to correct diagnosis ranged from 3 years to 34 years with an average of 20 years [5].

Diagnosis was difficult due to long history along with repeated surgical intervention. High level of suspicion was required to recommend for a direct laryngoscopy. Direct laryngoscopy remains the key to diagnosis in our case, which revealed the opening at apex of pyriform fossa. This was complemented by barium esophagogram and Computed tomography of neck. A variety of investigation options are available in literature. Though barium esophagogram and CT scan are well accepted means, they may fail to demonstrate any fistula tract in face of acute inflammation [8]. If combined with Valsalva manoeuvre, due to opening up of sinus, these means can provide with satisfactory image of tract [9]. But, direct laryngoscopy provides relevant information even in episode of acute inflammation regarding piriform fossa opening [10]. The barium esophagogram and direct laryngoscopy were found to be the most useful diagnostic tools [4]. Multiple options for intervention is available with respective recurrence rates. Open neck surgery is recommended as complete excision of tract including piriform sinus during a quiescent period [7]. Identification of inner opening can be facilitated by direct endoscopic placement of a Fogarty catheter [3] or injection of methylene blue through cannulation into the lumen before surgical exploration [11]. Resection or retraction of posterior border of the thyroid ala is must for adequate exposure. Retrograde excision, beginning at the piriform apex, ensures complete removal of the tract and protection of the recurrent laryngeal nerve [7]. This procedure may be combined with partial thyroidectomy [4,12], which shows the minimum recurrence rate in available literature. Endoscopic options include endoscopic transpharyngeal excision with a CO2 Laser

[13], chemocauterisation with 10% trichloroacetic acid, endoscopic monopolar diathermy [14].

A review and analysis of published cases of congenital fourth branchial arch anomalies shows recurrence rates of different treatment options: incision and drainage, 89%; open neck surgery and tract excision, 15%; endoscopic cauterization of the sinus tract opening, 15%; and open neck surgery with partial thyroidectomy, 8%. Complications after surgery occurred primarily in children 8 years or younger [4]. Planning for intervention was crucial for this patient, because repeated recurrences rendered it obligatory to excise the whole of fistula tract including the piriform fossa ending. The routine excision procedure for any branchial fistula starts with a neck incision with dissecting upward. But here we had to deal with some specific issues. First, lower end of fistula tract was excised twice with recurrence leading to loss of original anatomical architecture. Second, Upper opening of fistula tract was visible in piriform fossa – never been distorted surgically. Third, this persistent upper end remains the reason behind recurrence. So, if dissection is started from upper end, delineating the tract downward, the whole of tract can be approached and excised. So after passing an infant feeding tube through the opening in piriform fossa with methylene blue dye, the excision was done in a reverse process. The literature shows one approach that is similar to our approach to some extent [7], where the authors suggest that a retrograde excision ensuring complete removal by beginning at pyriform apex. Resection or retraction of posterior border of thyroid ala was also recommended for adequate exposure. But it differs from our approach at certain points. It was recommended for fourth branchial pouch sinus, not fistula. So their dissection ended near level of thyroid gland. But we could not stop at retracting the thyroid ala only, but to divide the isthmus of thyroid gland to delineate it downward.

Advantages of this approach are, first, the both ends of the fistula tract is identified, hence no chance of leaving any end for recurrence. Second, the we can avoid entering through the same skin end of fistula with distorted soft tissue architecture precluding complete removal. Third, transfixation of piriform end of fistula ensures cure without recurrence, literature also suggests closure of opening in pyriform fossa by surgery or other means favourable [7,13,14]. Fourth, it does not require removal of respective thyroid lobe, rather only division of isthmus suffice to dissect the node. Fifth, it does not involve resection of thyroid cartilage, as suggested by some authors [7], thus decreasing morbidity.

As usual some disadvantages are also there. First, large neck incision leads to large scar, and thus, worse cosmesis. Second, complications and morbidities associated with soft tissue dissection of neck and division of thyroid isthmus lies there, like, recurrent laryngeal nerve palsy, bleeding and pharyngocutaneous fistula. Though we performed this approach in a case of recurrence, we recommend its use in primary cases also, as the points suggested as advantages apply for primary cases too and precludes recurrence. Histopathology shows sinus tract is usually lined by pseudostratified ciliated columnar epithelium [3] or stratified squamous non-keratinised epithelium [15]. Our surgical specimen showed focal area of stratified squamous lining of tract wall, rest of the necrosis and inflammation can be explained by history of recurrent infection.

CONCLUSION

Because of distorted anatomy, intervention in 4th branchial arch fistula may differ from conventional approach.. A reverse surgical approach during a quiescent period comprising complete excision of the fistula tract starting at the pyriform sinus end and dissecting downward along the fistula tract upto skin end, combined with transfixation of both ends and division of thyroid isthmus is recommended for treatment of recurrent as well as primary cases for complete removal of disease and preventing recurrence.

References

1. Nicollas R., Guelfucci B., Roman S., Triglia J. M. Congenital cysts and fistulas of the neck. *International Journal of Pediatric Otorhinolaryngology*. 2000;55(2):117–124.
2. Tucker HM, Skolnick ML. Fourth branchial cleft (pharyngeal pouch) remnant. *Trans Am Acad Ophthalmol Otolaryngol*. 1973 Sep-Oct;77(5): ORL368-71.
3. Godin MS, Kearns DB, Pransky SM, Seid AB, Wilson DB. Fourth branchial pouch sinus: principles of diagnosis and management. *Laryngoscope*. 1990 Feb; 100:174-8.
4. Nicoucar K., Giger R., Pope H. G., Jr., Jaecklin T., Dulguerov P. Management of congenital fourth branchial arch anomalies: a review and analysis of published cases. *Journal of Pediatric Surgery*.2009;44(7):1432–1439
5. Lee FP. Occult congenital pyriform sinus fistula causing recurrent left lower neck abscess. *Head Neck*. 1999 Oct;21(7):671-6.
6. Jeyakumar A, Hengerer AS. Various presentations of fourth branchial pouch anomalies. *Ear Nose Throat J.* 2004 Sep;83(9):640-2, 644.
7. Rosenfeld RM, Biller HF. Fourth branchial pouch sinus: diagnosis and treatment. *Otolaryngol Head Neck Surg*. 1991 Jul;105(1):44-50.
8. Nicollas R., Guelfucci B., Roman S., Triglia J. M. Congenital cysts and fistulas of the neck. *International Journal of Pediatric Otorhinolaryngology*. 2000;55(2):117–124.
9. Miyauchi A., Tomoda C., Uruno T., Takamura Y., Ito Y., Miya A., Kobayashi K., Matsuzuka F., Fukata S., Amino N., Kuma K. Computed tomography scan under a trumpet maneuver to demonstrate piriform sinus fistulae in patients with acute suppurative thyroiditis. *Thyroid*. 2005;15(12):1409–1413.
10. Choi S. S., Zalzal G. H. Branchial anomalies: a review of 52 cases. *Laryngoscope*. 1995;105(9):909–913.
11. Ford GR, Balakrishnan A, Evans JNG, Bailey CM. Branchial cleft and pouch anomalies. *The Journal of Laryngol & Otol* 106:137–143.
12. Hallak B, Bouayed S, Leishman C, Sandu K. Residual fistula of fourth branchial arch anomalies and recurrent left-side cervical abscess: clinical case and review of the literature. *Case Rep Otolaryngol*. 2014; 2014:931279.
13. Parker KL, Clary MS, Courey MS. The endoscopic approach to a fourth branchial pouch sinus presenting in an adult. *Laryngoscope*. 2013 Nov;123(11):2798-800.
14. Wong PY, Moore A, Daya H. Management of third branchial pouch anomalies - an evolution of a minimally invasive technique. Int *J Pediatr Otorhinolaryngol* 2014; 78(3):493-8.
15. Takimoto T, Yoshizaki T, Ohoka H, Sakashita H. Fourth branchial pouch anomaly.*J Laryngol Otol*. 1990 Nov;104(11):905-7.

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