

Case report

Three cases of branchial fistula in one family: a rare presentation

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ABSTRACT

Branchial cleft anomalies comprise approximately 30% of congenital neck mass and present as cyst, sinus or fistula. They occur due to disturbance in the maturation of the branchial apparatus during fetal development. They are congenital lesions usually present in childhood, but they are usually diagnosed in later childhood or early adulthood because of enlargement or infection. Branchial cleft fistulae are usually diagnosed earlier than cysts. Correct diagnosis leads to proper management. Complete surgical excision is the treatment of choice. Second branchial cleft and pouch anomalies are commonest amongst all branchial cleft lesions, but complete second branchial cleft anomalies with external and internal opening is rare. Branchial anomalies with family history are also very rare. Here we present a case report of complete branchial fistula with family history which was managed by complete excision of fistula through transcervical and transoral approaches. 3 members of one family in 2 generations presented with branchial apparatus anomalies (father and his two children, elder son and younger daughter). All of them were having branchial fistula on right side of neck since birth.

Keywords: Branchial fistula, Cysts, Step ladder incision

INTRODUCTION

Branchial apparatus was first described by Von Baer (1827). The term branchial fistula was coined by Simpson.¹ Branchial fistula is formed due to persistence of the embryonic second branchial cleft. Branchial anomalies compose approximately 30% of congenital neck mass and present as cyst, sinus or fistula.^{2,3} They are equally common in males and females and usually present in childhood or early adulthood.² Cysts are remnant of the cervical sinus without an external opening. Sinuses are persistence of the cervical sinus with external opening, whereas fistula also involves persistence of the branchial groove with breakdown of the branchial membrane resulting in a pharyngocutaneous fistula.²

Second branchial cleft anomalies are the most common, representing 95% of all branchial cleft malformation.²

Complete history, physical examination and radiological examination help in diagnosing the various types of branchial cleft lesions. Examination usually reveals a congenital opening on the lower neck, anterior to sternomastoid muscle with recurrent clear to mucoid discharge from opening, tract may end blindly forming sinus or may extend up to tonsillar fossa forming complete fistula.³ Sinogram can also be performed to define the track. Complete excision of fistulous tract by combine transoral and transcervical approach is curative treatment for such lesions.

CASE REPORT

A 20 year old female patient came with chief complaints of opening in lower part of neck (Figure 1) since birth and recurrent mucoid to mucopurulent discharge from opening since age of 6 year which was mainly associated with upper respiratory tract infection. Discharge from

opening get relieved after oral antibiotics. On physical examination there was small punctum in the skin at junction of middle two third and lower one third of anterior border of right sternocleidomastoid muscle. When patient presented there was no active discharge from punctum but on pressure above the opening few drops of mucoid discharge were milked out.



Figure 1: Preoperative picture of patient showing the fistula.



Figure 2: CT scan showing the well-defined cyst.



Figure 3: Post op picture showing the well healed scar.



Figure 4: Father with the fistulous opening.

Ear nose throat and general examination was normal. There were no other congenital anomalies. Full blood count and urine examination was normal. On the basis of clinical history and physical examination clinical differential diagnosis were made as 1) second branchial cleft sinus or fistula, 2) third branchial cleft fistula.



Figure 5: Brother with the fistulous opening.

History of similar lesion was also present in father and brother. There was no history of consanguineous marriage in the family. All of them were having branchial fistula on right side of neck since birth.

Ultrasound neck revealed thick wall elongated sinus tract in right side of neck in sub cutaneous plane oriented vertically lying just medial to lower half of sternomastoid muscle, anterior to right lobe of thyroid ending superiorly at right carotid artery bifurcation with an echoic fluid content. Major neck vessels were normal. CECT neck from base of skull to clavicle was done which showed a

cystic lesion along anterior border of lower part of sternomastoid muscle representing type 2 branchial cyst.

Routine examinations were done. Anesthetic fitness was taken and patient was posted for total excision of fistula under general anesthesia.

Surgical procedure

Surgical plan was made which included transcervical and transoral approach with stepladder technique for complete excision of tract. Elliptical incision was taken around the fistula over neck, and blunt dissection was carried out through subplatysmal plane. Soft tissue dissection was done. Soft bluish hue cystic swelling was seen delineating from sternomastoid muscle. Upper flap was raised up to cartilage. Then second incision was made at level of hyoid bone, tract was separated from the surrounding soft tissues and the strap muscle. Tract was going superiorly in the parapharyngeal space toward the tonsillar fossa. Then Boyle-Davis mouth gag was applied to visualize oropharynx. Intraorally, tract was seen extended up to area behind posterior pillar of tonsil which was seen as an indented area when traction was applied to the tract in neck. Incision was applied over that indented area and complete tract was mobilised, brought intraorally and removed. Complete fistulous tract with cannula was pulled through neck, which was of 11.5 cm in length. Oropharyngeal defect was closed with help of 3.0 catgut and both neck wound was closed with help of 3.0 mersilk suture and 3.0 catgut suture. Drain was kept in neck. Postoperative period was uneventful. For initial three days patient was kept nil by mouth and feeding was done through nasogastric feeding tube. Antibiotics and mouth gargles (betadine and hydrogen peroxide) was given for seven postoperative day.

At seventh postoperative day skin sutures was removed and oropharynx was examined for any defect, as skin sutures and oropharynx mucosa was healthy, patient was discharged on seventh postoperative day. Patient was kept on follow up on 15th, 1month and after 3 months. The postoperative follow up was uneventful.

Histopathological report confirmed branchial fistula tract lined with pseudo stratified ciliated squamous epithelium.

The patient had an uneventful recovery and is on regular follow up for last 1 year. Father and son were counselled but denied surgery at present due to some family issues.

DISCUSSION

Pharyngeal arches are mesenchymal derivatives from paraxial and lateral plate mesoderm and appear in 4th and 5th week of development. Pharyngeal arches play a role in the formation of face, ear and neck. They are covered externally by ectoderm, which forms cleft between successive arches and internally by endoderm which

forms pouches between arches.⁵ Anomalous development of these arches may result into cyst, sinuses and fistulae.

Branchial anomalies can be lined with either respiratory or squamous epithelium. Cyst often lined by squamous epithelium, whereas sinus and fistula are more likely to be lined with ciliated columnar epithelium.² Branchial cleft cysts occur three times more often than branchial fistulas.⁴

Second arch anomalies are classified into 4 types.

Type 1: Lesion lies anterior to the sternocleidomastoid muscle and do not contact the carotid sheath.

Type 2: Lesion are the most common and pass deep to the sternocleidomastoid and either anterior or posterior to the carotid sheath.

Type 3: Lesion passes between internal and external carotid arteries and are adjacent to pharynx.

Type 4: Lesion lies medial to carotid sheath close to the pharynx adjacent to the tonsillar fossa 2.

The second branchial cleft and sinuses are encountered along the lower third of anterior border of the sternocleidomastoid muscle and may be bilateral.^{3,6} Complete branchial fistula with external and internal opening is rare.⁷ Preoperative imaging of tract with contrast material delineates the entire tract and aids surgical planning. It also differentiates between sinus and fistula, and helps in complete excision thus minimising the chance of recurrence.⁴ Sometime complete tract cannot be demonstrated because it may be blocked by secretion and granulation. Anatomically fistulous tract passes deep to platysma muscle between second and third pharyngeal arch structures by ascending along the carotid sheath and passing medially between internal and external carotid arteries above the glossopharyngeal nerve and below the stylohyoid ligament.⁵ The fistula may open into pharynx, usually into tonsillar fossa.

It can present at any stage, more commonly in the first and second decade of life. Two to ten percent of them can be bilateral. 6% of the patient with complete fistula can have a family history of branchial fistula anomalies.⁷ Some patient may also present with branchiootorenal spectrum disorder (BORSD) which is characterized by malformations of the outer, middle, and inner ear associated with conductive, sensorineural, or mixed hearing impairment, branchial fistulae and cysts, and renal malformations ranging from mild renal hypoplasia to bilateral renal agenesis..

The definite treatment of branchial anomalies is complete surgical excision of tract, most suitable age for surgery is 2 to 3 year or as early as possible if it is already delayed and among surgical techniques stepladder incision is most accepted method, as it provide better visualisation

of tract near pharynx which is combined with transoral approach for complete excision of tract.^{4,7}

Other modality of treatment includes (a) sclerosing agents which is seldom used today due to the associated inflammatory reaction and the risk of necrosis with perforation into the pharynx, (b) stripping method was described by Taylor and Bicknell in 1977, but this has not been widely used due to great risk of damage to adjacent structures.⁸

Complication of surgery includes recurrence, which could be 3% in fresh cases and up to 20% in second surgical attempts. Other complications include secondary infection, injury to facial, hypoglossal, glossopharyngeal, spinal accessory nerves, injury to internal jugular vein, and hematoma formation. The recurrence rate following surgery varies up to 3% been reported, this is due to incomplete surgical excision.

Familial presentation of branchial fistula anomalies is very rare. Anand et al in his case series presented 7 cases of isolated branchial anomalies distributed in four consecutive generations of a Central Indian family: 6 were males and 1 was female. 3 members had a left-sided branchial sinus, 1 had a right-sided sinus, 2 had bilateral sinuses, and 1 had a right-sided cyst. The family shows that branchial (lateral cervical) cysts and sinuses are inherited as autosomal dominant characters, and that the two anomalies are indistinguishable genetically.⁹

CONCLUSION

Accurate diagnosis, radiological imaging and complete surgical excision results in complete cure of the branchial cleft anomalies.

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