Review Article

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Branchial fistula: review of literature and case report

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ABSTRACT

The branchial arches are the embryological precursors of the face, neck and pharynx. Second most common congenital lesions of the head and neck in children are the anomalies of the branchial arches, with second arch anomalies by far the most common. Clinically, these congenital anomalies may present as cysts, sinus tracts, fistulae or cartilaginous remnants with typical clinical and radiological findings. We report the case of 11-month-old male child with congenital Type I Second branchial fistula on the left side of neck.

Keywords: Branchial cyst, Branchial fistula, Branchial apparatus, Congenital cervical lesions

INTRODUCTION

Anomalies of branchial arches are uncommon anomalies which represent the embryological precursors of face, neck and pharynx. They are the second most common congenital lesions of head and neck in children and account for approximately 17% of all paediatric cervical masses. ¹⁻³ The branchial arches develop in the fourth week of gestation and are the embryological precursors of ear and muscles, blood vessels, bones, cartilage and mucosal lining of the face, neck and pharynx. In total, six pairs of branchial arches are formed on either side of the pharyngeal foregut in cranio-caudal succession. 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. ²⁻⁴

Thus, depending on the anatomic location, branchial anomalies have been classified into first, second, third and fourth arch anomalies. Second branchial arch anomalies are the most common accounting for approximately 95% of cases. First branchial arch anomalies account for 1-4% cases; third and fourth

branchial arch anomalies are extremely rare.²⁻⁵ 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.^{1,2}

The branchial apparatus was first described by Von Baer while its anomalies were first described by von Ascheroni.⁶ The branchial fistula is an uncommon anomaly of embryonic development of branchial apparatus. Amongst these, anomalies of second branchial arch as well as pouch are common. They represent 90-95% of branchial anomalies.⁷

Second branchial cleft anomalies

During embryonic development, the second arch grows caudally and it covers the second, third, and fourth branchial clefts. The cervical sinus of His is formed by the fusion of this second arch with the enlarging epipericardial ridge of the fifth arch. The edges of

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cervical sinus in the due course fuse and hence in life no defect is seen.

However, it is the persistence of intervening ectoderm that gives rise to branchial cyst. The branchial fistula results from breakdown of the endoderm, usually in the second pouch. In the normal course a persistent fistula of the second branchial cleft and pouch passes from the external opening in the mid or lower third of neck in the line of the anterior border of the sternocleidomastoid muscle, deep to platysma, as in our case, along the carotid sheath. The tract then passes medially deep between the internal and external carotid arteries after crossing over the glossopharyngeal and hypoglossal nerves. Finally, it opens internally in the tonsillar fossa.⁴ Second branchial cleft anomalies most commonly present as cysts followed by sinuses and fistulae.8 They have previously been classified into four different sub-types by Bailey in 1929:9

- 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
- Types I-III are the most frequently occurring second arch anomalies, with type II being the most common.

Clinical presentation and diagnosis

A fistula or cyst in the lower anterior or lateral region of the neck is most likely to represent a second branchial cleft anomaly. Fistulae are usually diagnosed in infancy/childhood with drainage of secretions or purulent material from an opening at the anterior border of the sternocleidomastoid within the lower third of the neck.

Cysts are most often diagnosed as a painless, compressible lateral neck mass in a child/young adult that may become tender and/or increase in size if they become infected.8 Fistula presents with mucoid discharge from the anterior aspect of neck.

In addition, presence of infection may lead to formation of an abscess and signs of inflammation at the site of opening. Although branchial fistula may occur at any age but commonly they present in first or second decade of life. ¹⁰ Most of the times it is a simple sinus opening that extend up the neck for a variable distance. Complete branchial fistula with internal opening into tonsillar region is rare. The diagnosis is most often clinical and radiological investigations are rarely asked for.

Radiological studies

Fistulogram: It delineates the tract and it is often the commonest investigation available. A complete fistula demonstrable by a fistulogram is uncommon. CT fistulogram with reformatted images delineates the relation of sinus tract to that of important structures of neck. It also helps in classifying the type of lesion, provides a roadmap for surgeon prior to surgery.

MRI: It is better suited in the assessment of deep tissue involvement. Most advantageous for Type I first branchial cleft cysts and for parapharyngeal masses that may be second branchial cleft cysts. On T1-weighted imaging, they may turn from low to high signal depending on the proteinaceous content of the cyst, but are typically hyperintense on T2-weighted imaging. It provides the relationship of glandular tissue to the mass (e.g. fat planes between the parotid gland and a parapharyngeal mass) and hence acts as a roadmap prior to surgery.¹²

CT Scan: They are well-circumscribed, low-density cystic masses with a thin wall. The mural thickening is attributed to the response of lymphoid tissue. Mural thickening and enhancement varies with inflammatory change and typically occurs in the setting of infection. A tissue 'beak' between the internal and external carotid arteries is pathognomonic of Bailey type III cysts.¹³

Ultrasound (US): Second branchial cleft cysts are typically well-circumscribed, thin-walled and anechoic with evidence of compressibility and posterior acoustic enhancement. They may contain internal echoes compatible with internal debris.¹⁴

Histology: They are filled with a turbid yellow fluid containing cholesterol crystals and are lined by stratified squamous epithelium.

Treatment

The treatment of choice for branchial fistula is Surgical Excision. Surgical management involves complete surgical excision encompassing the external sinus opening with dissection of the sinus tract. Several surgical approaches have been described for the management of a branchial fistula. These include:

- Transcervical approach, either by a Stepladder Approach or Through a long incision along the anterior border of Sternocleidomastoid muscle
- Combined pull through technique.

The standard surgery for a second branchial arch fistula is the Stepladder Approach originally described by Bailey in 1933 with two incisions in the neck that gives exposure of the fistula tract with less tissue dissection. The higher incision should be bigger than the lower one because the fistula tract is deeper in location in the vicinity of important neurovascular structures.

Case report

11-month-old male child was brought to the surgery OPD of ESIC medical college with the chief complaints of swelling in the lower part of neck on the left side since birth. It was associated with discharge of mucoid material from the swelling from last 6 months.

There was no history of increase in the discharge while taking meals or with hot bath. There was no history of any other swelling in the neck or any other systemic symptoms. The child had achieved normal milestones upto his age. On examination there was small pin point opening in the lower part of the neck left side at anterior border of sternocleidomastoid muscle, at its lower 1/3rd and 2/3rd junction, situated 3cm above medial end of left clavicle. Surrounding the opening there was hyperpigmented area with no active discharge present (Figure 1) (Figure 2).



Figure 1: Type-I branchial fistula.



Figure 2: Another view of Type I branchial fistula.

USG neck was done which showed well defined cystic lesion over lower part of neck with hyperchoic foci measuring 3X2 cm over lower part of sternocleidomastoid muscle.

Patient was planned for exploration and excision of fistula under general anesthesia. External opening of the fistula was cannulated with 24G needle and methylene

blue dye was injected, which filled the tract and cyst cavity underneath. A transverse elliptical skin incision was made around the external opening, (Figure 3) subplatysmal flaps were raised and the tract was found to be ending just below the platysma, along the anterior surface of sternocleidomastoid muscle. Grossly 3X2 cm sized tract was excised with mucoid material inside its cavity. The skin was closed in layers and pressure dressing was done. Histopathology confirmed tract lined by keratinoid stratified squamous epithelium (Figure 4).



Figure 3: Superficial wound reaching deep to play small upto anterior border of Sterno Cleidomastoid Muscle.

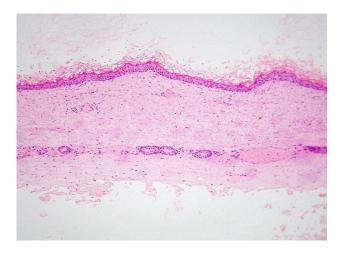


Figure 4: Keratinized stratified squamous epithelium lining the tract.

DISCUSSION

During embryonic development, the second arch grows caudally, enveloping the third, fourth, and sixth arches and fusing with skin caudal to these arches, forming a deep groove (cervical sinus). The edges of this grove then meet and fuse. The ectoderm within the fused tube then disappears.

Persistence of the ectoderm gives rise to a branchial cyst. A branchial fistula results from the breakdown of the endoderm. A persistent fistula of the second branchial cleft and pouch usually has its external opening in the neck near mid or lower part of SCM muscle. As it

ascends it pierces platysma. At the level of hyoid, it curves medially and passes between the external and internal carotids in relation to the hypoglossal and glossopharyngeal nerves. It opens in to the oropharynx usually in the intratonsillar cleft of palatine tonsil. In series of 98 cases by Ford et al.⁷ 78% presented by the age of five years and in vast majority there was history of intermittent discharge and infection of neck sinus since birth. In seven percent there was history of incision and drainage of an associated neck abscess. In his series 60% sinus opening was on the right side and 40% on left. In Sampath et al series of 17 cases 70% presented at age above 11 years.¹⁶ Only one patient presented at birth. 64.7% occurred on the right side and 35.3% on the left.

Second arch anomalies may take several forms. There may be only a simple sinus opening that extends up the neck for a variable distance. Branchial fistulas commonly present with persistent mucoid discharge from an opening in the skin of the neck. But rare and unusual presentation have also been documented. They have been documented as to present as parapharyngeal mass located in the supratonsillar fossa and extending to the lateral nasopharynx.¹⁷ Exceedingly rarely, a branchial cleft anomaly may be found to be malignant on presentation.¹⁸ A complete branchial fistula with external and internal opening is rare. The completeness of a fistula is diagnosed by a dye test in which methylene blue is injected through the outer opening and appears in the throat. A negative preoperative outcome on the test might become positive under general anaesthesia because of muscle relaxation.

Occasionally, the fistula tract may be blocked by secretion or granulation giving negative fistula test.¹⁹ In many a case, saliva is seen dribbling from the neck opening, which itself proves the completeness of the tract. In the 62 pediatric second branchial cleft anomalies, Bajaj et al.⁸ reported 50 of them to be unilateral and 12 to be bilateral. Several surgical approaches have been described for the management of a branchial fistula. The stepladder approach was described in 1933.20 The fistulous tract can be approached through a series of stepladder incision first encompassing the sinus opening and second overlying the carotid bifurcation. Subsequently the parapharyngeal portion of the fistula can be approached perorally after tonsillectomy. A wide cervicotomy incision (hockey stick) can also be used which allows for adequate exposure of neck structure for accurate dissection.²¹ In our case report Type I branchial fistula was present deep to platysma, just reaching upto anterior border of sternocleidomastoid muscle.

CONCLUSION

Abnormal development of Branchial apparatus lead to formation of different anomalies which remain asymptomatic and present later in life as cysts, sinuses or fistulae in the neck. Surgery is the treatment of choice for these lesions due to the fact that these lesions do not

regress spontaneously, and they have a high incidence of recurrent infection.

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