MIDLINE NECK FISTULA: 4TH BRANCHIAL CLEFT FISTULA VS INFECTED THYROGLOSSAL CYST

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ABSTRACT

OBJECTIVE: To present a case of a 19 year old female with recurrent mucopurulent discharge draining through midline neck fistula

: To present its clinical presentation, diagnostics and management

STUDY DESIGN: Case Report

SETTING: Tertiary Hospital

PARTICIPANT: One patient

RESULT: This is a case study which described an uncommon location of a 4th branchial cleft fistula in a 19 year old female with recurrent episodes of mucopurulent discharge. Complete ENT examinations were done and CT scan of the neck was requested with an initial impression of a 4th branchial cleft fistula versus an infected Thyroglossal duct cyst. The patient underwent neck exploration to trace the fistula tract. Its location was noted leading to a 4th branchial cleft. Excision of the tract was done and the specimen was submitted to the Department of Pathology for histopathologic finding, which revealed branchialcleft fistula. The patient improved after the operation and was eventually discharged.

CONCLUSION: This paper presented a case of a 19 year old female diagnosed with 4th branchial cleft fistula, presenting with an unusual presentation midline neck fistula draining with mucopurulent discharge. With the aid of CT scan, the tract of the fistula was identified. The patient was subjected to neck exploration and excision of the fistula tract.

KEYWORDS: Midline Neck Fistula, 4th Branchial cleft fistula, Thyroglossal duct cyts

CASE REPORT

Congenital anomalies of the neck occur as a consequence of disruptions in the complex development of the branchial apparatus of the fetus. This is classified according to branchial cleft or pouch of origin and its anatomic relationships. Congenital neck anomalies must be considered in the diagnosis of head and neck masses in children and adults. These include thyroglossal duct cysts, branchial cleft anomalies, dermoid cysts, and midline cervical clefts. It may be in the form of a fistula, sinus, or cyst, based on the completion of development of the anomalous structure[1]. Fistula is referred to as an anomaly when both external and internal openings are present[2] or may be defined as communication between two epithelized surfaces[3].

A detailed understanding of the embryology and anatomy of each of these lesions is essential to provide accurate preoperative diagnosis and appropriate surgical therapy, which are essential to prevent recurrence[2].

The objective of this study is to present a case of a midline fistula, which is unusual course of a 4th branchial cleft anomaly, and to report its management.

A 19 year old Filipino female from New Intramuros, Quezon City presented to the ENT outpatient with a history of fistula with an intermittent draining mucoid discharge in the midline of the lower neck since 6 years old which was associated with productive cough. Patient did not complain of any pain or difficulty of swallowing. The patient consulted a General Physician in a private institution where several antibiotics were given with incision and drainage. However, despite completing the course of antibiotics and Incision and Drainage, the midline neck fistula draining with mucopurulent discharge would recur almost every 2 years. No diagnosis was made nor disclosed to patient. The patient suffered from the condition of having intermittent discharge for 13 years.

One month prior to consult, cough was noted. There was no fever and swelling was noted on the left lower anterior neck which was painful and the fistula on the midline of neck was again draining with purulent mucopurulent discharge. Hence, patient decided to seek consult with an ENT specialist.

The past medical, personal and social and family history were unremarkable.

The clinical examination revealed a fistula with swelling and granulation it which was tender upon palpation and with purulent mucopurulent yellowish discharge in the midline of the anterior lower neck (*See Appendix A, Fig 1*). No cervical lymphadenopathy was present. Other physical examinations were unremarkable.

CT scan of neck with contrast was requested *(See Appendix A, Fig. 2)* which revealed an elongated left paramedian tract with central hypodensity that extends from the left infrahyoid region down and deep into the left

strap muscles near midline and ends in a pouch-like structure at the level anterior to the superior pole of left lobe of the thyroid gland. The length measures 2.6 cm with the superior end pouch measuring 1.5 x 0.8 cm, while the tract within the strap muscle has a diameter of 0.3m and the inferior pouch is 0.9 x 0.7 cm. Another round heterogeneous nodule was also seen at the region of the lower pole of the right lobe of the thyroid gland, which measures 1.0 x 1.2 x 1.4 cm. The impression was Thyroglossal duct cyst vs. 4th branchial cleft cyst, and a consideration of right cystic thyroid nodule.

The patient was subjected for Neck Exploration with excision of cyst under general anesthesia. Curvilinear incision was made along the skin crease to include the fistulous tract. Flaps were elevated on both sides for a better view of the surgical field. *(See Appendix A, Fig. 3).* The Fistula tract was noted at left paramedian area at the lower part of left sternocleidomastoid muscle. The tract was followed and on superior dissection the fistula tract was noted going laterally along the mid sternocleidomastoid muscle level. At this point the fistula tract was noted going medially to the left superior of thyroid lobe. The tract was followed and was noted inferior to the superior laryngeal nerve and superior to the recurrent laryngeal nerve. The superior end of the tract was noted at the hyoid bone. The body of the hyoid bone was cut together with the end of the fistula tract. No tract was noted to enter the pyriform sinus.

A nodule was excised approximately 1.1 cm x 0.5 cm which was seen at lower pole the right thyroid gland.

All specimen were sent to the pathology department along with the fistula tract which measures 6 cm x 1.1cm x 0.9 cm *(See Appendix A, Fig 4*). Histopathological examination revealed a tan elongated partly cystic tissue with lining of pseudostratified ciliated columnar type of epithelium with a few lymphoid aggregates in the wall confirms microscopically a branchial cleft cyst *(See Appendix A, Fig. 5).*

The patient improved and was subsequently discharged.

DISCUSSION

Congenital cervical cystic, sinuses and fistulae must always be considered in diagnosing head and neck masses in children and adults[2]. Complete clinical history and physical examination and knowledge in embryology is always warranted for a proper diagnosis[8]. The diagnosis may easily be established based on the presence of lesion. In our case, the patient presented with a midline neck fistula with recurrent mucopurulent discharge, we can consider an infected thyroglossal duct cyst, as well as an infected dermoid cyst and midline cervical cleft which is found primarily in the anterior midline neck[8], however a third or fourth branchial anomaly may also be found terminating in thyroid gland or paratracheal lesion[3].

Thyroglossal duct cysts are the most common midline congenital neck masses,

accounting for as many as 70% of all congenital neck anomalies. No gender predilection has been reported, and the age of affected patients ranges from birth to 70 years; approximately 50% of patients present before the age of 20 years[5].

An infected dermoid cyst is considered since this is another midline congenital anomaly which results from the entrapment of epithelial elements along embryonic lines of fusion. This presents as painless superficial subcutaneous masses at the anterior neck, this can be close to hyoid and moves with swallowing and tongue protrusion. It gradually increases in size. Infection is rare but the cyst can rupture and may present with granulomatous inflammation. This is commonly diagnosed before 3 years of age[2], however was ruled out because there is no obvious submental swelling[5] in the patient, as commonly seen in patients with infected dermoid cyts. Histologic findings may further rule out this diagnosis.

Another congenital neck anomaly is the midline cervical cleft. This is usually present at birth as cutaneous ulceration with overhanging skin or cartilaginous tag found at the anterior lower midline of the neck. Commonly, a sinus tract is noted which extends downward from the skin and may connect to the sternum or mandible or end in a blind pouch[2]. However this was unlikely the diagnosis since this congenital anomaly is present a birth and was commonly associated with other cleft abnormalities of tongue, lower lip or mandible[2].

The clinical features and usual locations of congenital neck anomalies are summarized in the table (*See Appendix B, Table 1*), which aids in ruling out dermoid cyst and midline congenital cyst.

We can consider branchial cleft anomaly, this said to be a common neck mass and is approximately 30% of congenital masses[2]. It was assumed that the congenital masses result from the cervical sinus of His. This type of neck mass usually presents with a non-tender, fluctuant mass that may become inflamed and abscess during a respiratory infection[9], later with a draining fistula as its presenting sign and symptom. The branchial cleft cyst is commonly located in the medial side of the sternocleidomastoid muscle[10]. Branchial cleft cysts gradually enlarge and frequently are not visible until the second or third decade of life [2]. They are equally common in males and females and usually present in childhood or early adulthood [2] however according to *Mandell*[11] there is a slight female preponderance. There are four branchial anomalies reported, and most branchial anomalies is the second branchial arch and occurs in 95%, while the remaining 5% remaining are from the first and third branchial remnants. Fourth branchial anomalies are said to be rare, with 45 cases reported literature [13] or approximately 1-2% of all branchial anomalies[11].

A fourth branchial anomaly has two distinct clinical presentations, in the first, the lesions was said to be present in neonates as lateral neck cysts or abscesses with

obstructive airway symptoms, while, second group, the lesions were present in children, adolescents, and occasionally adults as recurrent lateral cervical abscesses or as recurrent acute suppurative thyroiditis[11]. According to *Cote*[4], fourth branchial cleft sinuses are frequently diagnosed after recurrent episodes of infection in the sinus tract, the sinus might be misdiagnosed for years, and the patient may undergo multiple procedures before the diagnosis is properly made and treated.

Mandell cited in his literature that 97% of fourth branchial pouch anomalies occur on the left side, its tendency has yet to be explained, however it can be due to the asymmetric development of fourth arch vascular structures. On the other hand, Thyroglossal duct cyst as previously mentioned is located at midline near the level of hyoid bone because the tract passes anterior to the hyoid bone [9]. Seventy-five percent of cases of thyroglossal duct cyst is located in midline or slightly off midline (25%) in the anterior neck; however within 2 cm of midline. Paramedian location is most in thyroglossal cysts and occurs on the left for the reasons not well understood [14].

Clinical history and physical examination is the most important factor in evaluating congenital neck anomalies. In correlation with this case, the clinical presentation of the patient may mislead us in the diagnosis, presenting with a midline fistula however very low in the neck with recurrent mucopurulent discharge, associated with upper respiratory infection. Such midline presentation is uncommon in a 3rd or 4th branchial anomaly, the midline location is said to be the clinical presentation of a thyroglossal duct cyst. Confusion to whether the mass is a third or fourth branchial cleft cyst or an infected thyroglossal duct cyst arises.

CT scan of neck was requested, to trace the fistula tract. It was cited in *Koeller*[8] that radiologic images of 4th branchial may appear as cyst fluid which may vary in signal intensity on T1-weighted images depending on the protein concentration and is typically hyperintense relative to muscle on T2-weighted images. A fourth branchial cleft cleft cyst or fistula is seen connected to pyriform sinus[8]. While a thyroglossal duct cyts manifests as either in the midline of the anterior neck at the level of the hyoid or within the strap muscle just off midline[8]. Current tomography is able to demonstrate the fistula in up to 64% of cases. Barium esophagram can also be helpful with a 50% to 80% sensitivity for third and fourth branchial fistulae [3]. In this case CT scan revealed an elongated left paramedian tract with central hypodensity that extends from the left infrahyoid region down and deep into the left strap muscles near midline and ends in a pouch-like structure at the level anterior to the superior pole of left lobe of the thyroid gland, due to its location the impression was 4th branchial cleft cyst vs Thyroglossal duct cyts. The role of computed tomography is still being defined however it has been reported as helpful in making the diagnosis in some instances[11]. The result of the CT scan of the patient guide us in diagnosis, however regarding Branchial anomalies, it was cited in *Mandell*[11] that both third and fourth branchial anomaly is found

terminating in pyriform sinus, thus tracing the tract is necessary.

Intraoperative finding of our case showed that the fistulous tract was noted laterally along the lower part of sternocleidomastoid and continuous upward until it swings back again at the midline toward the hyoid bone. The tract has no connection from the thyroid gland and there was no attachment to pyriform sinus. *Mandell*[11] reported that third and fourth branchial pouch sinuses may be difficult to distinguish, because both types commonly begin at the pyriform sinus and travel a short distance before ending blindly in the paratracheal or thyroid gland regions. The connection or attachment of pyriform sinus was not observed in this case and probably the tract has finally degenerated or fibrosis might have occurred.

Fourth branchial anomalies usually contain thymic tissue as do cysts and sinuses that result from thymic or parathyroid rests, but only branchial anomalies have the connection to the pyriform sinus. The anatomical variation of the fourth arch anomalies depends on the infected side. On the right *(See Appendix A, Fig. 6)*, the lesion loops around the subclavian artery, pass deep to the internal carotid artery, ascending to the level of the hypoglossal nerve,descending along the anterior border of the SCM to enter the pharynx at the pyriform apex or cervical esophagus. While on the left, the tract descends into the mediastinum, looping around the aortic arch, medial to the ligamentum arteriosus, then ascends in a similar course to the right side. Fourth arch lesions present as lateral cysts in the lower third of the neck. However, a complete fourth branchial fistula has never been clinically described, perhaps because of the extremely convoluted course it would have to take[15].

Fourth pouch sinuses that actually leave the neck and descend below the clavicles into the mediastinum are rare, hence, of key importance in distinguishing a third from a fourth arch sinus is the relationship of the anomalous tract to the superior and recurrent laryngeal nerves. This must be determined surgically. *Mandell*[11] cited that if the tract was found to course inferior to the superior laryngeal nerve (a fourth arch derivative) and superior to the recurrent laryngeal nerve (a sixth arch derivative), a fourth pouch origin is suggested, however if the tract is found to pass cranial to the superior laryngeal nerve and inferior constrictor muscle, a third pouch origin is likely. In this case, intraoperative finding shows that the tract is more likely a fourth branchial anomaly as described by *Mandell*[11] because the tract is inferior to the superior laryngeal nerve and superior to the recurrent laryngeal nerve.

A case report by *Ureta and Ponce*[18] cited the Davies model that shows the tract of a 4th branchial cleft tract, which ascends above the thyroid cartilage, and its internal opening is found in the thyrohyoid membrane. It was also mentioned in the case report that embryologic anatomy involved in predicting

the course of the 4th branchial anomaly is complex and a complete 4th branchial fistula may never be surgically demonstrated since exposure of the tract would be too aggressive and unnecessary.

The histologic findings of the four midline neck congenital anomaly were presented in the table *(See Appendix B, Table 2)*. Branchial anomalies are lined with respiratory or squamous epithelium. Fistulae are more likely to be lined with ciliated, columnar epithelium, while thyroglossal duct cysts reveals a stratified squamous epithelium or ciliated pseudostratified columnar epithelium with ectopic tissue, which is present in 3 to 20%. Lymphoid nodules in the wall of the thyroglossal duct cyst are found 15 to 20% of cases as compared with branchial cleft cyst which is present in about 75% to 80%. . It has been reported in the study of Gilmour that accessory thymic tissue may originate from the fourth branchial pouch[17].

The most precise manner is to distinguish the histologic findings of cellular components of the cyst wall and surrounding tissue, however in some cases the preexisting inflammation of infected thyroglossal duct cyst may produce metaplasia of the lining of a thyroglossal duct cyst, which will make histologic differentiation from a branchial cleft cyst difficult[14]. Immunoperoxidase staining for thyroglobulin may be helpful in such cases[16].

In this study, the histologic finding revealed a lining of pseudostratified ciliated columnar type of epithelium with few lymphoid aggregates, pathologic report revealed a Branchial Cleft Fistula.

CONCLUSION

In conclusion, this paper presented a case of a 19 year old female diagnosed with 4th branchial cleft fistula, presenting with an unusual presentation of midline neck fistula draining with mucopurulent discharge. With the aid of CT scan, the tract of the fistula was identified. The patient was subjected to neck exploration and excision of the fistula tract.

RECOMMENDATION

It is important to remember that 4th branchial cleft fistula is a rare branchial anomaly with recurrent episodes of infections and should be considered in diagnosing a congenital midline neck fistula.

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ILLUSTRATION

APPENDIX A

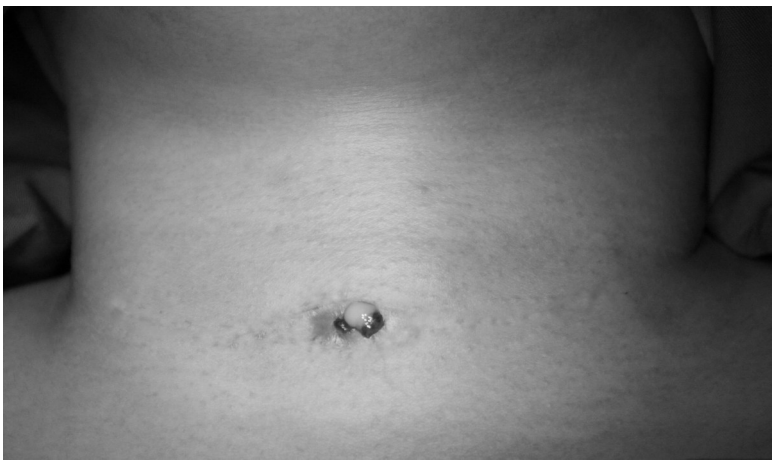
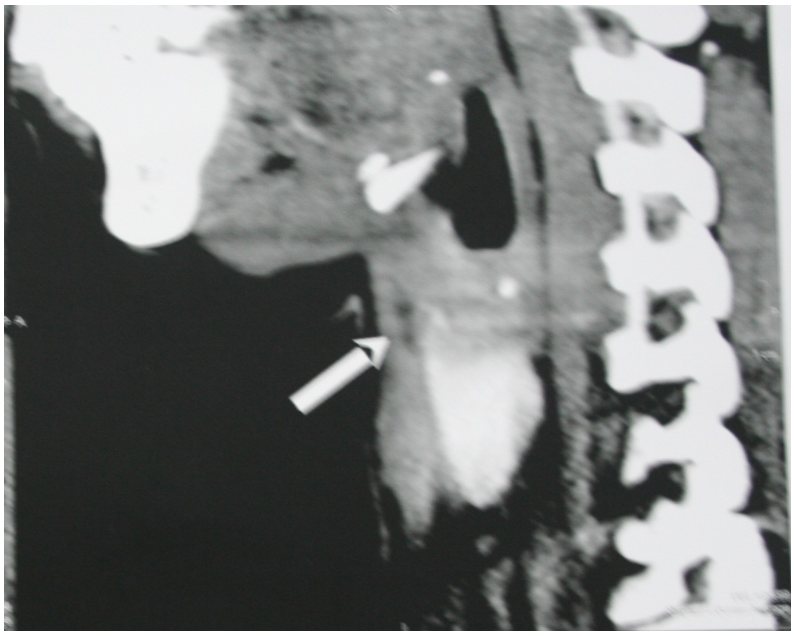


Figure 1. At presentation, granulation tissue are found around the fistula with mucopurulent mucoid discharge at the midline of neck of the patient



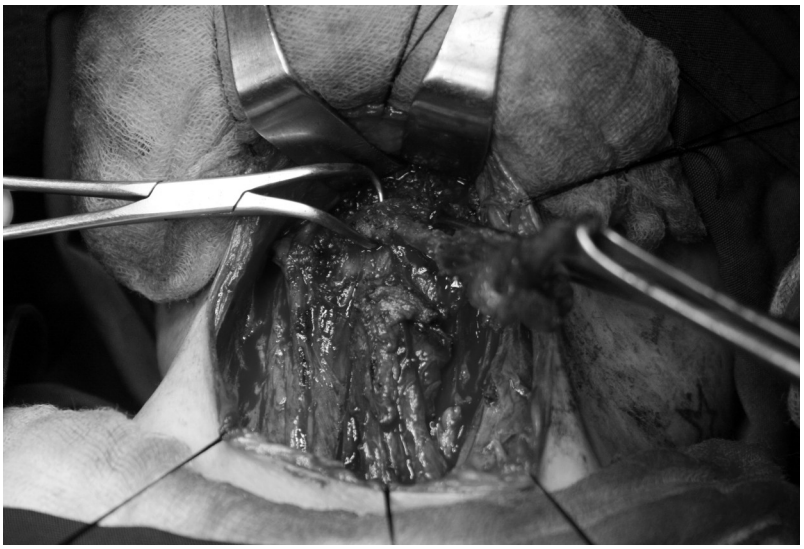


Figure 3. Intraoperative finding shows the fistula tract going from left paramedian of left sternothyroid going medially attached to hyoid bone

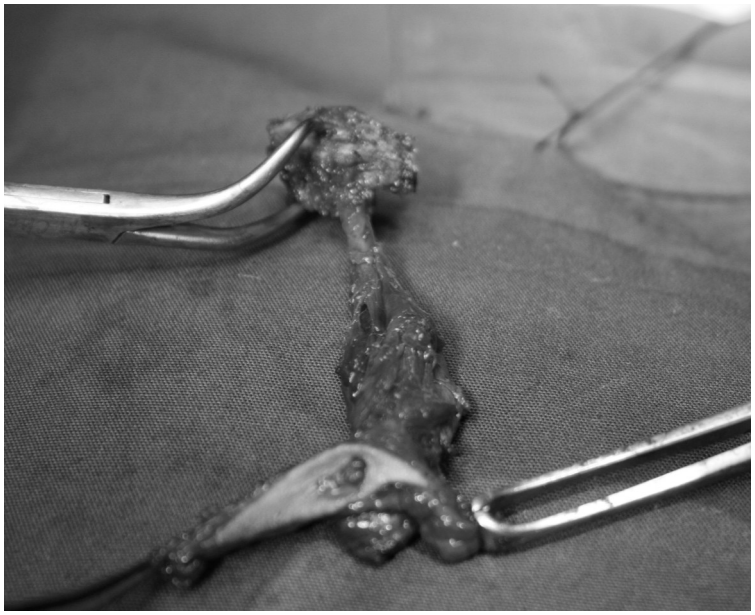


Figure 4. The excised sinus tract of the patient measuring 6 cm x 1.1cm x 0.9 cm

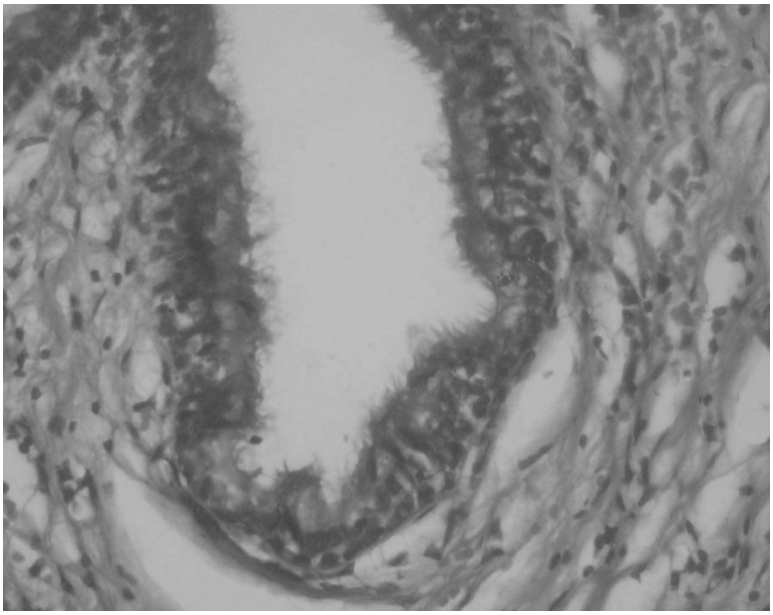


Figure 5. Histopathologic result of the patient revealed branchial cleft cyst showing a lining of pseudostratified ciliated columnar type of epithelium with few lymphoid aggregates.

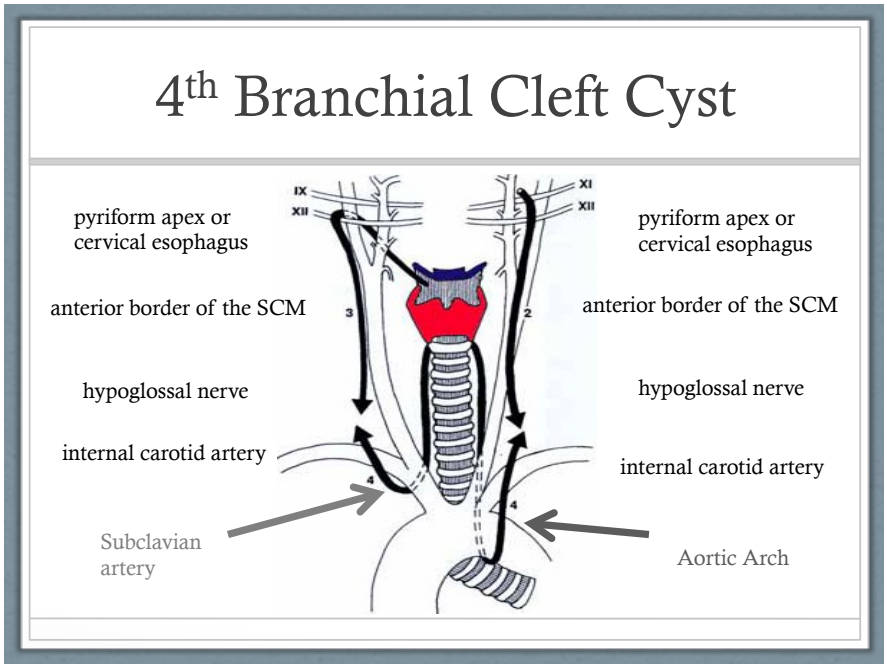


Figure 6. Demonstrating the tract of the right 4th brachial cleft fistula and left 4th branchial cleft fistula

Source: Mandell, David L. MD. Head and Neck Anomalies Related to the Branchial Apparatus .Otolaryngologic Clinics of North America. Volume 33, Issue 6 , Pages 1309-1332, 1 December 2000

APPENDIX A

Table 1. Clinical Features of Congenital Neck Lesions

Lesion	Peak Prevalence (Age)	Sex Predilection	Usual Location
Thyroglossal duct cyst	<10 y	Equal	Hyoid level or below (80%), within 2 cm of midline
Branchial cleft cyst			
First	Middle age	F > M	Parotid, external auditory canal
Second	10–40 y	Equal	Submandibular space, lateral to carotid vessels
Third	10–30 y	...	Left posterior cervical triangle
Fourth	**Any age**	**Female**	**Sinus tract arising from left pyriform sinus; cystic structure located at lower anterior border of SCM**
Dermoid cyst	10–30 y	Equal	Floor of mouth; painless superficial subcutaneous masses at the anterior neck, this can be close to hyoid and move with swallowing; obvious submental swelling
Midline Cervical Cyst	Present at birth		Cutaneous ulceration with overhanging skin or cartilaginous tag found at the anterior lower midline of the neck

Source: *Koeller, Kelly et . al, Congenital Cystic Masses of the Neck: Radiologic Corelation Archives of the EEP.1999; Mandell, David L. MD. Head and Neck Anomalies Related to the Branchial Apparatus .Otolaryngologic Clinics of North America. Volume 33, Issue 6 , Pages 1309-1332, 1 December 2000*

Table 2. Histologic Characteristic of the Four Midline Congenital Anomaly

LESION	HISTOLOGIC FINDING
Branchial Cleft Fistula	**Lined with ciliated, columnar epithelium with lympoid nodules in the wall**
Thyroglossal Duct Cyts	Stratified squamous epithelium or cilitated pseudostratified columnar epithelium with ectopic thyroid tissues; 15-20% lymphoid tissue
Dermoid Cyts	Lined with epithelium with epithelial appendages such as hair, hair follicles or sebaceous glands
Midline Cervical Cyst	Fibrous tissue with interwoven skeletal muscle

Source: *Pilch, Ben MD. Head and Neck Surgical Pathology. 2001. p 8-15. ; Acierno, Stephanie MD et. al. Congenital Cervical Cyst, Sinuses and Fistulae. Otolaryngology Clinics of North America 40(2007)161-176.*