Branchial Cleft Anomaly: An Unusual Tale of Bilateral First and Second Arch

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ABSTRACT

Objective: Combined congenital 1st and 2nd branchial arch abnormalities are rare. The role of tonsillectomy and electrocautery in managing second branchial arch fistulas are discussed.

Case Report: The case report depicts a tale of a 12 month old girl with bilateral neck discharge and bilateral congenital microtia. Imaging confirmed a bilateral 2nd branchial cleft fistula and sinus. She underwent electro-cauterisation of her inner fistulous tract, tonsillectomy followed by excision of her bilateral branchial fistula. A step ladder approach was adopted to reduce surgical morbidity. This is the first report of a 1st branchial cleft anomaly resulting in EAC pathology without a fistula with simultaneous 2nd complete branchial arch fistulae.

Conclusion: A combined approach surgical excision is definitive but ipsilateral tonsillectomy alone is not effective in preventing recurrence of 2nd complete branchial fistula.

KEY WORDS

rare, branchial, arch, cleft, anomaly

INTRODUCTION

Branchial originates from 'Branchia' in Greek which is responsible for the formation of amphibian gills. There are five branchial arches where external invaginations form clefts form the skin while the internal invaginations form pouches responsible for the development of the middle ear, tonsils, thymus and parathyroid glands (Branstetter, Coombs, Phillips, Naul, Levey, 2015). Remnants of these branchial arches may develop into a branchial cleft cyst, sinus or fistula (Prasad, Azeez, Thada, Rao, Bacciu, Prasad, 2014).

Approximately one third (33%) of congenital neck masses and 17% of paediatric neck masses are due to branchial cysts (Prasad et al, 2014), (Table 1). This is a rare case of an infant presenting with congenital abnormalities arising from the 1st and 2nd branchial arches. The authors discuss the role of tonsillectomy and electrocautery in managing second branchial arch fistulas.

CASE REPORT

A 12 month old girl, Chinese in ethnicity presented with bilateral discharging neck since birth. Discharge was whitish and mucopurulent, occasionally foul smelling and often prominent after feeding. She has bilateral congenital microtia (grade II) with right canal stenosis and left canal atresia. ENT assessment revealed non-tender bilateral neck fistulae (Figure 1).

Magnetic Resonance Imaging (MRI) of the neck at 12 months showed a left fluid filled sinus inferior to the angle of the mandible extending superiormedially to the left SCM towards the left palatine tonsils. There was also a right fluid filled sinus of similar position and trajectory to the right tonsils. The external anterolateral neck communications were suggestive of bilateral second branchial cleft fistulae and sinus.

A direct laryngoscopy revealed an internal fistula opening at the superior pole of the right palatine tonsil and midsection of the left tonsil (Figure 2). Bilateral internal openings were diathermised with bipolar. However, parents informed of persistent mucopurulent discharge from bilateral brachial fistulas. Subsequently, she underwent a second procedure at 20 months. Intraoperatively, there was spillage of methylene blue at the left tonsil inferior pole and none the right side. Therefore, a left tonsillectomy was performed and the internal fistula opening was diathermised. An external exploration of the right branchial fistula revealed a fistulous tract at the anterior border of the SCM with a blind ending at the carotid sheath. The right tract was clamped, tied and excised up to the level of the carotid sheath.

Post procedure, the left branchial fistula discharge was persistent despite a significant (approximately 70%) reduction. She underwent an excision of her left branchial fistula at 22 months. Histopathology examination confirmed no malignancy. Periodic outpatient follow-up revealed no further neck discharge.

DISCUSSION

Although branchial arch abnormalities are rare among children, the combination of bilateral 1st and 2nd branchial arch abnormalities are even rare. Second branchial anomaly is more common than the first as...
Table 1. Branchial arch abnormalities and subtypes with associated nervous innervations. Second branchial cleft cyst is the most common, followed by 1st and 3rd [2,3]

<table>
<thead>
<tr>
<th>Brachial Arch</th>
<th>Location of brachial cleft cyst of fistula</th>
<th>Nervous Innervation</th>
</tr>
</thead>
<tbody>
<tr>
<td>First</td>
<td>Type I</td>
<td>Trigeminal (CN V)</td>
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<td></td>
<td>External Auditory Canal</td>
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<td></td>
<td>Parotid Gland</td>
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<td></td>
<td>Type II</td>
<td>Facial (CN VII)</td>
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<td></td>
<td>Angle of mandible</td>
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<tr>
<td></td>
<td>Submandibular gland</td>
<td></td>
</tr>
<tr>
<td>Second</td>
<td>Type I</td>
<td>Vestibulocochlear (CN VIII)</td>
</tr>
<tr>
<td></td>
<td>Anterior to the SCM muscle</td>
<td></td>
</tr>
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<td></td>
<td>Type II</td>
<td>Glosopharyngeal (CN IX)</td>
</tr>
<tr>
<td></td>
<td>Anterior to SCM, posterior to Submandibular gland, lateral to carotid sheath</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Type III</td>
<td>Vagus (CN X)</td>
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<td></td>
<td>Medial to bifurcation on internal and external carotid arteries</td>
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</tr>
<tr>
<td></td>
<td>Type IV</td>
<td>Fifth Spinal Accessory (CN XI)</td>
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<td>Pharyngeal space deep to the carotid sheath</td>
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</table>

**Figure 1.** Discharging anterior neck with no surrounding skin erythema. External openings are anterior to the mid-section of the sternocleidomastoid muscle (SCM) and inferior to the level of the hyoid bone

**Figure 2.** Fistula test was positive for bilateral internal fistulous openings on dilation of bilateral external openings with methylene blue. This indicates a 2nd branchial arch anomaly. Left: Right tonsil; Right: Left tonsil

supported by an earlier case series in Malaysia (Zaifullah, Yunus, See, 2013).

Branchial fistulas are more common than cysts but may occur together, which can be confirmed by contrasted neck CT (Prasad et al, 2014). Cysts may present at later ages compared to fistulas due to the time taken to form a collection and become symptomatic (Teo, Ibrahim, Tan, 2015).

Although 1st branchial cleft anomaly involves the duplication of the External Ear Canal (EAC) membrane (resulting in cysts, sinuses and fistulae); it can also manifest in the absence of a normally patent EAC resulting in atresia or stenosis (Blevins, Byahatti, Karmody, 2003). Belvins et al 2003 reported 3 out of 4 patients had unilateral aural atresia with 1st branchial cleft preauricular lesions. This was supported by Abdollahi Fakhim et al 2014 of a child with congenital middle ear and canal cholesteatoma, EAC stenosis and 1st branchial fistula (Abdollahi Fakhim, Naderpoor, Mousaviagdas, 2014).

The 1st branchial cleft is responsible for the development of the EAC and lateral surface of the tympanic membrane (Work, 1972). Taking into account of our understanding the embryology of the EAC, it is safe for us to assume that our patient does have a bilateral 1st branchial cleft abnormality despite not having a 1st branchial fistula. This may also be considered as a Branchiootoic Syndrome in absence of renal pathology, thus excluding a Branchiootorenal syndrome (Senel, Kocak, Akbıyık, Saylam, Gulleroglu, Senel, 2009).

The combination of a 1st branchial apparatus maldevelopment as manifested by EAC hypoplasia (atresia/stenosis) with a 2nd bilateral branchial fistula makes it a unique case. A search on PubMed and Medline revealed no reported cases of such using keywords Choana; Atresia; Stenosis; Second; Branchial; Anomaly.

Imaging of a branchial fistula incorporates ultrasound, fistulogram, contrasted CT or MRI. Contrasted CT allowed for the evaluation of middle ear structures in view of the patient’s hypoplastic ear canals. MRI allowed for assessment of soft tissue in the neck to ascertain fistula extension to guide intraoperative excision.

Children often do not tolerate fistulograms and are not preferred unless performed intra-operatively (Teo et al, 2015). Moreover intra-operative laryngoscopy should be performed to ascertain the internal opening of a branchial fistula via a fistula test. This is made easier with the use of dye such as methylene blue.

Our patient had a bilateral 2nd Branchial (Type 1) arch anomaly with a complete fistula as confirmed with dilatation with methylene blue. Ford et al 1992 reported only 2 of 106 patients with branchial arch anomalies had a complete fistula, indicating its rarity (Ford, Balakrishnan, Evans, Bailey, 1992). In our patient, our initial attempt to ablate the internal opening resulted in effective masquerade of one of the two internal openings of the fistula (right tonsillar). Even this did not cease the discharge from the external opening.

A unilateral left tonsillectomy still failed to culminate patency of the left branchial fistula. This is consistent with Cheng et al 2012 whom concluded that ipsilateral tonsillectomy does not determine recurrence rate. In that study, 14/36 (43.8%) underwent unilateral tonsillectomy with excision of branchial fistula, but only one non-tonsillectomy case developed recurrence which was not significant (Cheng, Elder, 2012).

Electro-cauterisation is not effective in treating branchial fistulas as the tract will remain patent because the internal opening may fail to scar and close. However, electro-cauterization has been effective in treating 3rd and 4th branchial sinuses (Derks, Veenstra, Oomen, Speleman, Stegeman, 2016). A negative fistula test as seen in the right tonsil could be secondary to granulation tissue or thick secretions resulting in a false negative test (Ang, Pang, Tan, 2001).

The young patient was treated conservatively with multiple procedures to reduce surgery associated morbidity, given her age. Surgical excision is gold standard in treating branchial fistulae. A combined approach surgical excision and laryngoscopy with fistula tests ensure complete excision (Jovic, Saldanha, Kuo, Ahmad, 2014). As a result,
1st and 2nd Branchial Cleft Anomaly

recurrence rate can be reduced. A step ladder approach encompasses a series of incision starting with the skin from the external opening down to the carotid bifurcation may be adopted (Prasad et al, 2014).

CONCLUSION

Literature review revealed only five reported cases of bilateral simultaneous 1st and 2nd branchial arch abnormality. Although combined approach surgical excision is definitive, ipsilateral tonsillectomy alone is not effective in preventing recurrence of 2nd complete branchial fistula.

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REFERENCES