International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Rare Case of Pharyngo Oto Cutaneous Fistula

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Abstract: ***First branchial cleft anomalies are rare congenital anomalies of the head and neck that represent less than 1% to 8% of all branchial cleft defects. These anomalies are thought to result from incomplete obliteration of the clefts that arise between the branchial arches during embryogenesis. The persistence of these structures varies widely anatomically and often misdiagnosed. As a result, several classification schemes have been developed according to anatomic position, number of openings, and the type of tissue lining the tracts. We report an unusual case of a first branchial apparatus anomaly, a fistula communicating between the oropharynx and mastoid with external cutaneous opening in post auricular area.***

Keywords: Branchial cleft, congenital anomaly, facial nerve, 1st branchial cleft anomaly

1. Case Report

A 2 day old full term male child delivered by vaginal delivery of twin pregnancy referred from a private hospital to our institute with Very low birth weight, small left ear and an abnormal opening behind malformed left ear (Figure 1). On examination asymmetric cry facies was seen with malformed small left pinna s/o microtia and fistulous opening in retro auricular region anterior to left Sternocleidomastoid showing copious drainage of saliva, feed through it. No signs of infection were noted. On 2D ECHO examination ASD with mild left to right shunt was seen. This rare association of cardiac anomaly with assymentric facies is s/o Cayler Cardiofacial syndrome. On passing Infant feeding tube through oral cavity, distal end of the tube is seen coming out from the previously mentioned external opening. (Figure 2)

Figure 1: External opening of the fistula

Figure 2: Infant feeding tube coming out of the fistulous opening

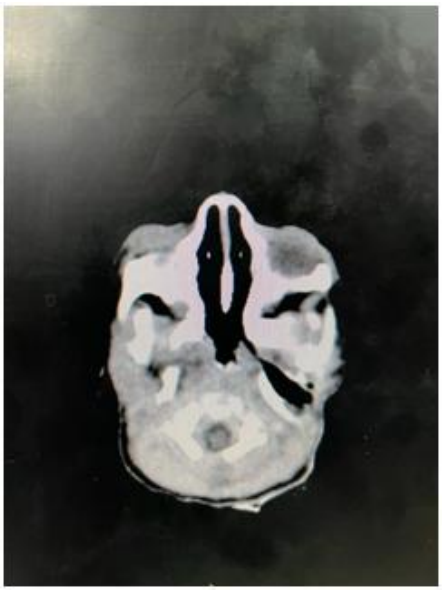

Figure 3: Axial image of fistula extending from oropharynx to left retroauricular area

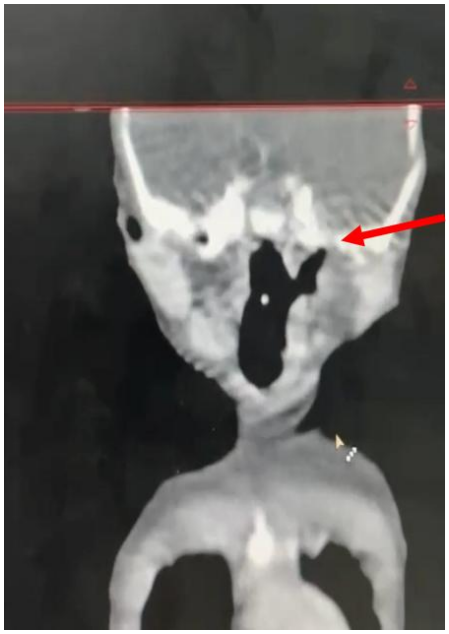

Figure 4: Coronal images of fistula tract

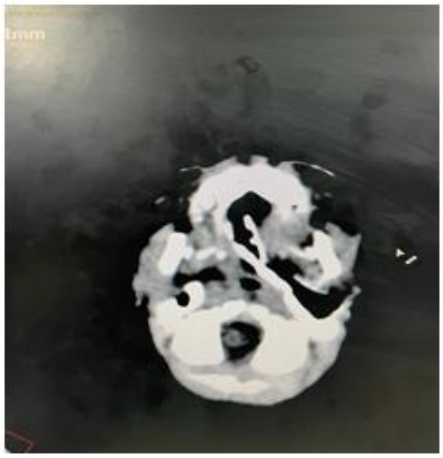

Figure 5: Axial image of fistulous tract post administration of contrast through external opening of fistula

On contrast enhanced CT scan of head and neck region shows left EAC which is not well developed. Left ear ossicles and inner ear appear normal. Infant feeding tube is seen along fistulous tract extending from oropharynx laterally through neck soft tissues traversing anterior to left mastoid with external opening in left retro auricular area on skin surface. (Figure 3, 4, 5). On CT abdomen and pelvis, hemivertebrae seen at L2-L3 levels.

2. Discussion

Buccopharyngeal membrane separates mouth from the pharynx. It disappears by the end of the third week after forming bars at the pharyngeal walls. The mesodermal condensation of these bars is known as branchial arches. The ventrally fused U- shaped arches form the support of pharynx and it consists of six arches of which fifth arch becomes rudimentary. Branchial cleft forms arch laterally and pouch internally. Derivatives of 1 st branchial arch are maxilla, mandible, sphenomandibular ligament, incus, malleus, anterior malleus ligament and muscles of mastication. External auditory meatus (EAC) arises from the groove and eustachian tube and middle ear from the pouch of 1st branchial cleft. 1st branchial cleft is the only cleft that does not obliterate at the end of embryonic development [1]. Only the dorsal part of 1st groove takes part in forming EAC and the ventral groove usually disappears. In rare occasion it can persist and present as preauricular sinus or fistula. Anomalies of the first branchial cleft are rare, accounting for less than 8% of all branchial anomalies [2, 3], with an annual incidence of 1/1 000 000 [4], Arnot (1971) classified 1st branchial cleft anomaly into type 1 and type 2 based on anatomical variant followed by Work in 1972 who took both histology and anatomy into consideration and came up with more detailed classification. In 1980 Olsen et al. further classified the anomaly to cysts, sinuses or fistulas of lobule, canal, post auricular region or angle of jaw. Point of time at which the disorder develops determines whether it's Type 1 or Type 2 anomaly.

Comparison between Arnot and Work Classification on 1st branchial cleft anomaly:

Arnot and Work classification [5, 6]:

Type 1	Cyst or sinus which are ectodermal in origin at parotid gland	Lesion is adjacent to EAC medial to conchae and extends to postauricular crease or supra auricular region. It's ectodermal in origin and usually lies superior to main trunk of facial nerve.
Type 2	Sinus or fistulous tract opens between upper neck and EAC	Cyst, fistula or sinus is usually both ectodermal and mesodermal in origin arising between upper neck and angle of mandible. The lesion passes through parotid gland close to facial nerve in variable relation. The fistula usually opens below angle of mandible.

Various presentation has been reported for type 2 first branchial arch anomaly which includes chronic purulent ear discharge with sinus openings at floor of EAC, fistula or abscess discharging in Poncet's triangle at lateral aspect of neck above hyoid bone and anterior to sternocleidomastoid muscle [7, 8], (the position of sinus or fistula opening at neck varies from pre, infra or post auricle to submandibular region), preauricular cyst, swelling at parotid region or fistulous opening anterior to fossa of rosenmuller at lateral nasopharyngeal wall which

is an extremely rare [9]. Our case is classified as Work's Type 2 based on the location of fistulous tract which is extending posteriorly to post auricular region. Obliteration of ventral part of 1 st branchial arch takes place simultaneously with emergence of parotid gland as ectodermal ingrowth and migration of facial nerve from second branchial arch which explains the close relation between 1 st branchial cleft anomaly with parotid gland and facial nerve [7]. In 2003 Solares, et al. reported on relation of facial nerve to 1 st branchial cleft anomaly in 10 patients whereby 2 of the lesion ran lateral to facial nerve, 7 ran medial and only 1 ran in between branches of the nerve [9]. In general sinus tract runs superficial to facial nerve compare to fistulous tract and in presence of opening at EAC the tract mostly related superficially to facial nerve [10, 11]. In addition to history and clinical presentation, radiographic visualization is important to establish diagnosis and in planning for surgery.

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