Bangladesh J Otorhinolaryngol 2011; 17(1): 72-75

Case Report

A rare variant of first branchial cleft fistula

AFM Ekramuddaula[1], Hasan Zafar Rifat[2], Mazharul Shaheen[3]
Mohammad Abdus Sattar[4], Mohammad Idrish Ali[5]

Abstract:

Objective: To report an extremely rare variant of first branchial cleft anomaly.

Case report: A-13-year old boy presented with the chief complaint of persistent right aural discharge since infancy. On examination, a 1 mm fistulous opening was present in the floor of cartilaginous portion of external auditory canal. And scanty watery discharge came out through this opening when the patient asked to swallow. Sinogram through discharging meatal opening showed a thick irregular tract about 5 cm length traversing the soft tissue of neck extending from the floor of right external auditory canal to the lateral pharyngeal wall and spillage of contrast into the oral cavity.

Conclusion: This was an extremely rare variant of first branchial cleft fistula. Due to the rarity of this disease, clinical misdiagnoses and inappropriate management may lead to recurrence and complications. For better prognosis, appropriate diagnosis and surgical planning are very important.

Key words: *First branchial anomaly, branchial cleft fistula, branchial cyst, fistulography.*

Introduction:

First branchial cleft anomalies - which include branchial cysts, sinuses, and fistulas-account for less than 8% of all branchial cleft anomalies and its annual incidence being approximately one per 1000000.[1] Such anomalies are the result of incomplete obliteration of the clefts that arise between the branchial arches during embryogenesis, producing persistent anomalous structures with considerable variation in their anatomical course.[2] Due to high degree of variability, several different classifications have been published, based on the anatomical position, number of openings and type of tissue lining the tract.[3] This present paper report a case, in which a tortuous fistulous tract was found between the floor of external auditory canal and superior pole of tonsil, diagnosed by fistulography and injection of colored drink through opening of external auditory canal. An extensive literature search revealed that this variant of first branchial cleft anomaly has not been previously reported.

1. ENT-Head & Neck Surgeon, Popular Diagnostic Center, Uttara, Dhaka, Bangladesh.
2. Consultant, ENT, Victoria Hospital (Sadar), Naryanganj, Bangladesh.
3. Assistant Professor of ENT, Sylhet MAG Osmani Medical College, Sylhet, Bangladesh.
4. Assistant professor of ENT, BSM Medical University, Shahbag, Dhaka, Bangladesh.
5. Medical Officer, Department of ENT, BSM Medical University, Shahbag, Dhaka, Bangladesh.

Address of Correspondence: Dr. AFM Ekramuddaula, ENT-Head & Neck Surgeon, Popular Diagnostic Center, Uttara, Dhaka, Bangladesh.

Case report:

A 13-year-old boy presented with the chief complaint of persistent right aural discharge since infancy. The discharge, was aggravated

during intake of meal and was usually thin watery but occasionally thick, coloured.

On examination, a 1 mm fistulous opening was present in the floor of cartilaginous portion of external auditory canal. And scanty watery discharge came out through this opening when the patient asked to swallow. Examination of oral cavity after pouring of coloured drink through the fistulous meatal opening revealed spillage of coloured material from the tonsillar fossa close to upper pole of tonsil.

A sinogram was done by introducing contrast media through discharging meatal opening, which showed a thick irregular track about 5 cm length traversing the soft tissue of neck extending from the floor of rt external auditory canal to the lateral pharyngeal wall and spillage of contrast into the oral cavity.(figure-1, 2)

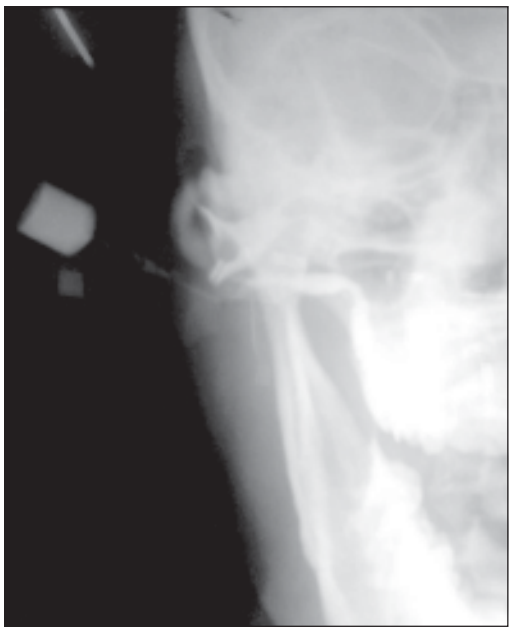

Figure-1: *Sinogram of meatal discharging opening showing spillage of contrast in the oral cavity.*

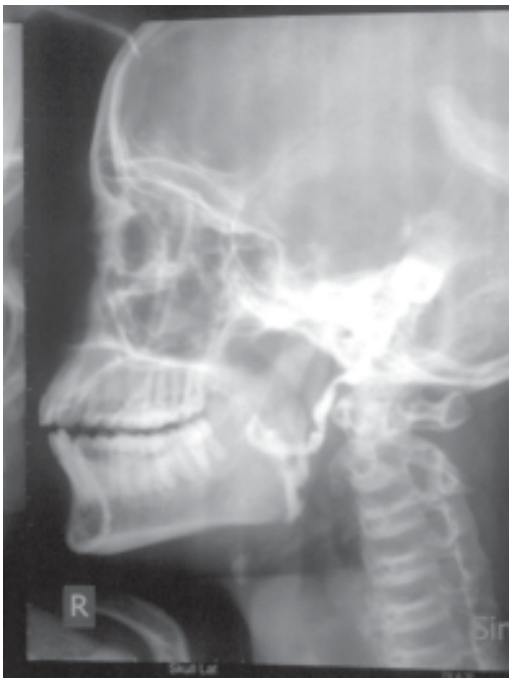

Figure-2: *Sinogram of meatal discharging opening showing a thick irregular tract extending to lateral pharyngeal wall.*

We planned for complete resection of sinus tract through lateral cervical or transparotid approach. For this purpose, we advised for computed tomogram fistulography using diatrizoate meglumine dye. However, after the patient's attendants counselled regarding the nature of the problem, its surgical treatment and the likely results of surgery versus conservative treatment & follow up, they opted to defer surgery until it became unavoidable.

Discussion:

First branchial cleft anomalies are rare congenital malformations, with only about 200 cases reported in literature.[4] There rarity and diverse presentations have frequently led to misdiagnosis and inappropriate treatment. In a trend towards specialisation /subs-pecialisation, first branchial cleft anomalies, with their varied clinical manifestations, may possibly present to an otology, head and neck

surgery and even general paediatric surgery practice.

Branchial apparatus was first described by Baer in1827. Five pairs of branchial arches appear at about the 4th week of fetal life. The maxilla and mandible as well as the parenchymal part of parotid gland developed from the first branchial arch. At the 6th week of fetal life, the ventral portion of the first branchial groove disappears gradually as the first and second branchial arches fuse together. The dorsal portion of the first brachial groove deepens and develops into the external auditory canal and the outer layer of the ear drum, while the dorsal part of the first branchial pouch becomes the Eustachian tube and the middle ear. The inner layer of the ear drum comes from the endoderm of the first pouch. In the process of development, the facial nerve migrates upwards. Therefore, the anatomical relationship between the branchial cleft anomalies and the facial nerve sometimes may vary greatly.

In 1970's, Arnot[5] and Work[6] classified the first branchial cleft anomalies into two types based on anatomical and histological features respectively. According to the classification of Work, the type I anomaly is less common than the type II anomaly. It is of ectodermal origin and considered to be a duplication of the membranous external auditory canal. It generally occurs medial to the concha of the auricle and culminates in a "cul-de-sac" on a bony plate at the level of the mesotympanum. The type II anomaly is of both ectodermal and mesodermal origin and may contain cartilage. It consists of a fistula running from the floor of the external auditory canal to the upper neck near the submandibular angle. A case combined with some characteristics of both type I and II anomalies was reported[7]. Volaris and Pahor[8] proposed a variant of the fistula tract extending from the ear canal to the mastoid and sternocleidomastoid muscle as type III anomaly. S Ramnani et al,[9] reported a variant of fistula which extend from the external inframeatal opening to the lateral nasopharyngeal wall, anterior to the fossa of Rosenmuller and its surgical excision was postponed due to difficult approach as well as patient's attendant disinterest. Clinically, a type II anomaly usually presents as a recurrent abscess or a mass with a sinus orifice in the unilateral upper neck as well as frequent otorrhea and a sinus orifice in the ear canal floor. Sinus orifices and fistula tracts may be found in the middle ear, paraphayngeal space,[10,11] Eustachian tube, concha cavity, or retro-auricular skin area.[10]

Although congenital in origin, first branchial cleft anomalies can present later in life, at a mean age of 18.9 years (range, 20 days to 82 years)[2], approximately twice as often in woman (69%) as in men (31 percent). Fistulae occur more frequently on the left side (64 percent), while sinuses show no side preference.[2]

The clinical picture of first branchial cleft anomalies usually overlaps with those seen in other more common disease. Otorrhoea is the most frequent otological symptom and the condition should be suspected if recurrent/ chronic otorrhea is present in the absence of chronic otitis. A sinus/fistula opening in the external auditory canal is present in only 44% of patients, and even if such an opening exists, it may not necessarily appear obvious.[12] In 2 out of 3 cases reported by a study[12] first branchial cleft anomalies are associated with a myringeal web, an epidermal structure which extends from the floor of the external auditory canal to the umbo of the tympanic membrane. This varied clinical presentation along with its rarity, makes misdiagnosis common, leading to inadequate and inappropriate treatment. The resultant recurrent infections and repeated surgery lead to increased scarring and higher risk of iatrogenic facial palsy during its subsequent surgical removal.

Imaging studies are useful in aiding diagnosis. CT scan can confirm the diagnosis by showing the tract near the external auditory canal to be wide and surrounded by cartilage.[12] A fistulogram is a useful diagnostic tool in the case of a sinus or fistula without signs of cyst formation or inflammation.[13]

The aim of treatment is to remove the lesion completely and safely with preservation of the facial nerve and to reconstruct otological structures if necessary . The best chance of achieving this is through early diagnosis at primary presentation and the availability of the necessary surgical expertise. Operation plans should be postponed in infants younger than 6 months old due to their undeveloped auricular cartilage and mastoid.[14] Generally, the complications include recurrence(12.5%), infection (7.7%), facial palsy (2.9%), ear canal stenosis (0.96%), ear drum perforation (0.96%), and keloid formation.

Our patient was male and had his fistula opening in the floor of right external auditory canal. Fistulography revealed a long tortous fistulous tract extending external auditory canal through soft tissues of neck to the oropharynx. As the tract course was in the area formed by the first branchial arch, the lesion appeared to be a rare variant of first branchial arch anomaly. The great variability in the course of these lesions, and the necessity of complete excision during treatment, makes CT radiographic visualision crucial for definitive treatment.[3] Complete excision of patient's tract would require careful handling of cranial nerves, the facial nerve in particular, along with other vital structures.

References:

1. Arndal H, Bonding P. First branchial cleft anomaly. Clin Otolaryngol 1996; 21: 203-207.
2. D'Souza AR, Uppal HS, De R, Zeitoun H. Updating concepts of first branchial cleft defects: A literature review. Int J Pediatr Otorhinolaryngol 2002; 62:103-109
3. Whestone J, Branstetter BF, Hirsh Behulsing M. Fluoroscopic and CT fistulography of the first branchial cleft. AJNR Am J Neuroaudiol 2006; 27: 1817-19.
4. Wittekindt C, Schondorf J, StennetE, JungFrazer JE. The nomenclature of diseased states caused by certain vestigial structures in the neck. Br J Surg 1923; 11: 131-136.
5. Arnot RS. Defects of the first branchial cleft. South Afr J Surg 1971; 9: 93-98.
6. Work WP. Newer concepts of first branchial cleft defects. Laryngoscope 1972; 82: 1581-1593.
7. Lim LH, Hartnick CJ, Willging JP. A rare case of combined type I and II first branchial sinus. J Pediatr Surg 2003; 38: 12-13.
8. Violaris NS, Pahor AL. A variation of first branchial cleft anomalies. J Laryngol Otol 1993; 107:625-626.
9. S Ramnani, V Mungutwar, NK Goyal, A Bansal. A rare variant of first branchial cleft fistula. J Laryngol Otol 2009; 123:1387-1389.
10. Mukherji SK, Tart RP, Slattery WH, Stringer SP, Benson MT, Mancuso AA. Evaluation of the first branchial anomalies by CT and MR. J Comput Assist Tomogr 1993; 17: 576-581.
11. Shirley DM, Santos PM: An unusual first branchial cleft defect. Otolaryngol Head Neck Surg 1995; 113: 829-832.
12. Sichel JY, Halperin D, Dano I, Dangoor E. Clinical update on type II first branchial cleft cysts. Laryngoscope 1998; 108: 1524-1527.
13. Stokroos RJ, Mianni JJ. The double auditory meatus-rare first branchial cleft anomaly: clinical presentation and treatment. Am J Otol 2000; 21: 837-41.
14. Triglia JM, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN. First branchial cleft anomalies: A study of 39 cases and a review of the literature. Arch Otolaryngol Head Neck Surg 1998; 124: 291-295.