

CASE REPORTS

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First branchial cleft fistula: a case report and literature review

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Abstract

Background: First branchial cleft fistula is a rare malformation of the neck and head. The purpose of this report is to showcase a method of treating first branchial cleft fistula while preserving the facial nerve.

Case presentation: This case report presents an 8-year-old female with a type 2 first branchial cleft fistula. The treatment process follows multidisciplinary approach with the aid of health professionals from different departments, pre-op magnetic resonance imaging (MRI) scanning to map the fistula, and the use of nerve monitoring device (NIM).

Conclusion: The procedure was successfully done while preserving the facial nerve.

Keywords: First branchial cleft, First branchial cleft fistula type 2, Otorhinolaryngology, Pediatric surgery

Background

The first branchial cleft is an extremely rare occurrence that manifests as cysts, sinuses, or fistulas. From all branchial abnormalities, the first branchial cleft fistula makes up almost 4% of the branchial abnormalities [1]. It can be distinguished based on the histological and anatomical traits. First branchial clefts can be caused by the inadequate removal or closure of the ectoderm [2]. The location of the fistula can be sufficiently determined through magnetic resonance imaging (MRI scans).

Case presentation

An 8-year-old female patient presented with a mass on the left side of her neck with a history of sinus discharge. The cystic lesion was first noted months after birth, but it was smaller in size. The patient has no history of hearing loss and took an otologic examination, and the evaluation results were normal and reveal that no effect on the tympanic membrane. The patient ran MRI scans, and scans showed a fistulous tract extending along the left side of the neck to the middle ear (Fig. 1). Surgery was the

preferable choice of treatment. The patient underwent surgical excision to extract the cyst. The operation followed a multidisciplinary approach with the aid of anesthesiologists, ENT doctors, radiologists, and pediatric surgeons. During operation, methylene blue dye was injected in the cervical opening of the fistulous tract to mark the fistula. An elliptical incision was done around the fistula. It extended along the left mandible to the ear. The branches of the facial nerve were superficial to the fistula. Throughout the operation, nerve monitoring device was used to detect the facial nerve. Also, ENT surgeon attended the procedure to double-check the integrity of the facial nerve. The fistula has been extracted with no complications. The postoperative sessions revealed that the facial nerve function was normal. There were no signs of drooping or difficulty controlling facial muscles. No complications or visible recurrences of sinus or any residue were noted in a 2-year follow-up (Fig. 2).

Discussion

From all branchial cleft anomalies, the most common defect arises from the second branchial arch, which makes up almost 95% of defects, then the first arch of less than 6% of defects. Embryologically, after the third week of embryonic development, six pairs of branchial

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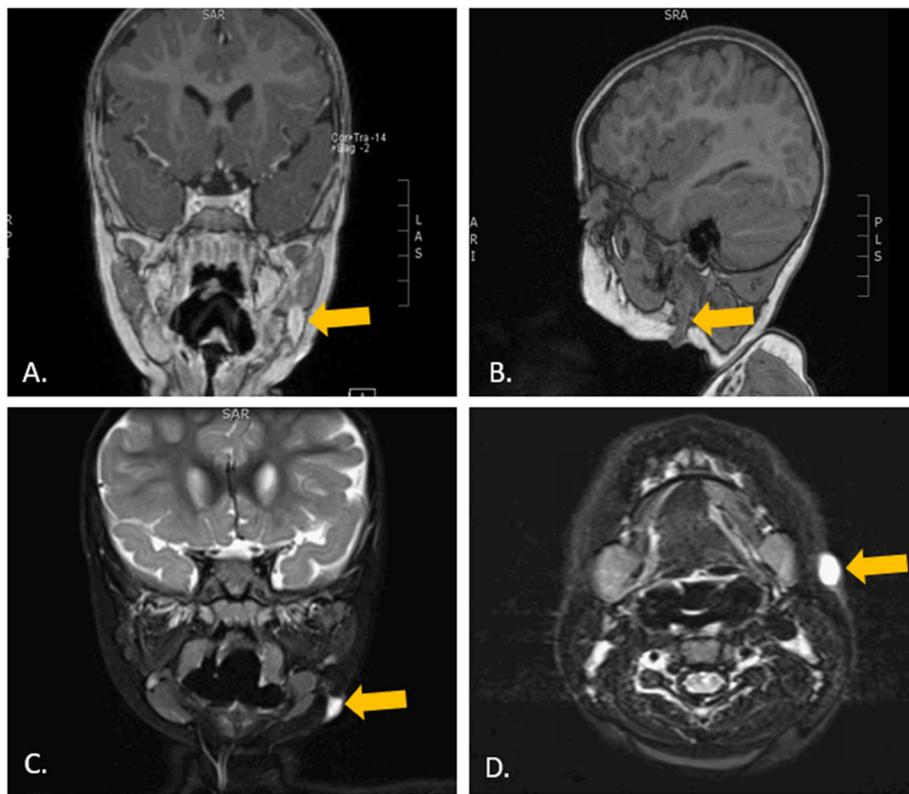


Fig. 1 There is evidence of a branchial cleft fistula measuring 1.1 cm that is directed toward the head and medial to the pinna of the ear. MRI scan T-1. **A** Coronal cut. **B** Sagittal cut. **C** Coronal cut. **D** Axial cut

arches and clefts start to appear in the neck and head of the embryo. Various cartilages and vessels will start to form from these arches. First branchial arch appearance is associated with the embryonic growth of parotid. Additionally, it is the precursor of the formation of the maxilla that arises from the first arch, and the first pharyngeal pouch forms the middle ear, eustachian tube, and the mastoid antrum. During the sixth gestation, these arches start to merge and disappear; however, in

some rare cases, the first branchial arch will not completely disappear [3], thus, resulting in the first branchial cleft anomaly. Branchial cleft cysts, fistulas, and sinuses may occur in any age group, but the first and second decades of life are the most common [4].

First branchial cleft has two distinctive types—type 1 and type 2—and the classification is based on the anatomical and histological location of the cleft by the studies of Arnot and Work, respectively. Arnot’s type 1

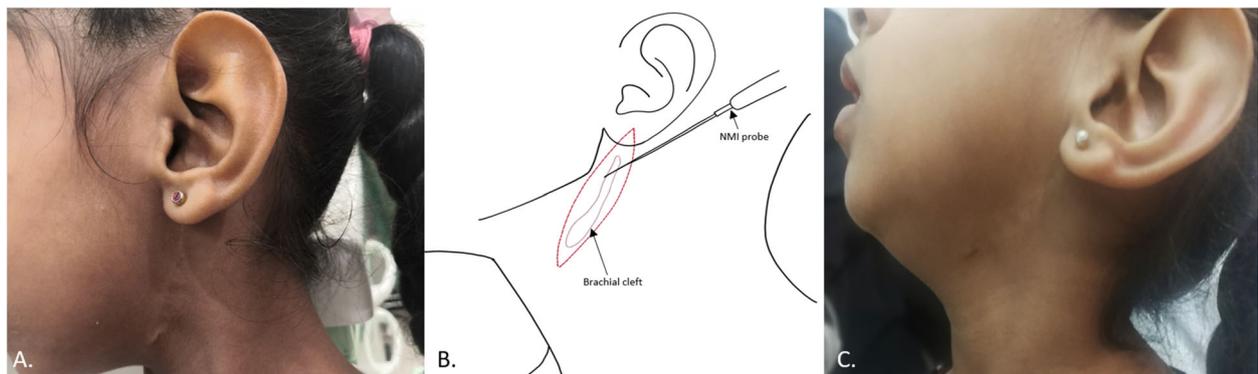


Fig. 2 **A** Pre-operative clinical photo showing the fistulous mass. **B** Schematic diagram of intra-operative with the aid of NIM. **C** Postoperation during follow-up sessions showed no signs of discharge or recurrence

lesion has sinuses or cysts found on the parotid gland, while type 2 lesion is located between the external auditory canal (EAC) and the upper neck. According to Work, type 1 is the duplication of the membranous external ear canal parallel to the normal ear canal, whereas type 2 is the duplication of the membranous external auditory canal and the pinna, which is located in the upper neck near the angle of the mandible [5]. In our case, the histological and anatomical properties demonstrated type 2 first branchial fistula.

Due to its rarity, the first branchial cleft fistula is usually falsely diagnosed and mistreated [6]. Early diagnosis and proper treatment are necessary for avoiding recurrences and preventing damage to the facial nerve. A branchial cleft cyst is a fluctuant mass that should be removed once diagnosed, for the cyst can spread and harden over time and be a source of infection and drainage [7, 8]. In case of leaving any tissue residue, it will have a high chance of recurrence. The chief constraint was preserving the facial nerve (marginal mandibular nerve). The facial nerve and the first branchial cleft are closely related more than any other type of branchial defect. This correlation makes it indispensable to use a nerve monitoring device (NIM) routinely throughout the operation. Using NIM can lower the risks of recurrence and damage to the facial nerve. In case of not using NIM, it can risk damaging facial nerve which can lead to facial nerve palsy, infections, recurrence, and negatively impact the patient. Also, MRI scans are extremely beneficial in the identification of cysts by providing a high degree resolution of soft tissues and major neck vessels [9]. The proximity to the facial nerves and the uncommonness of the anomaly results in high rates of complications. Complication rates were lower when otolaryngologists performed surgeries because of the higher experience with branchial anomaly surgeries than other surgeons. Consequently, it is highly recommended to seek specialized centers [10].

Conclusions

In conclusion, first branchial cleft is a rare occurrence that is often misdiagnosed which can lead to insufficient treatment and delay of proper treatment. Preserving the facial nerve is the main goal of the operation. Using nerve monitoring device is essential to the succession of the operation.

Abbreviations

NIM: Nerve monitoring device; MRI: Magnetic resonance imaging; ENT: Ear, nose, and throat; EAC: External auditory canal

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Authors' contributions

RB and OB selected the case report. RB did the extensive literature review and interpreted the patient data regarding the diagnosis of the first branchial cleft sinus. RB and OB performed the case history and presentation and was a major contributor in writing the manuscript. RB and AB did the final revision and correction of the manuscript. All authors contributed to the conception and design of the report and/or the acquisition and interpretation of data. Drafts were revised critically for important intellectual content and the final version approved by all. All agree to be accountable for all aspects of the work and have read and approved the final manuscript.

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Availability of data and materials

The datasets generated and/or analyzed during the current study are not publicly available due [patient confidentiality policy in our institution] but are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Formal written consent was obtained from the patient's parent.

Competing interests

Author RB declares that she has no conflict of interest.

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References

1. Work WP. Newer concepts of first branchial cleft defects. 1972 *Laryngoscope*. 2015;125(3):520-532. doi: <https://doi.org/10.1002/lary.25202>. Epub 2015 Feb 11. PMID: 25677102.
2. Cho SI. A Case of First Branchial Cleft Fistula Presenting with an External Opening on the Root of the Helical Crus. *Case Rep Med*. 2018;2018:4215802. doi: <https://doi.org/10.1155/2018/4215802>. PMID: 29560006; PMCID: PMC5831870.
3. Faruque O, Wischhusen JD, Reckley LK, Rooks VJ, Liming BJ. First branchial cleft fistula (Work Type 2) with an internal opening to the Eustachian tube: Case report and review of literature. *Radiol Case Rep*. 2019;14(7):819-824. doi: <https://doi.org/10.1016/j.radcr.2019.03.036>. PMID: 31049118; PMCID: PMC6484227.
4. Al Sukhun J, El Naggar M. Unusual Presentation of a Large Multilocular Second Branchial Cleft Cyst. *J Craniofac Surg*. 2019;30(6):1772-1773. doi: <https://doi.org/10.1097/SCS.00000000000005506>. PMID: 31033768.
5. Liu Y, Li T, Xue J, Jia J, Xiao S, Zhao E. First branchial cleft fistula presenting with internal opening on the Eustachian tube: Illustrated cases and literature review. *Int J Pediatr Otorhinolaryngol*. 2012;76(5):642-645. doi: <https://doi.org/10.1016/j.ijporl.2012.01.028>. Epub 2012 Feb 14. PMID: 22341630.
6. Liu H, Cheng A, Ward BB, Wang C, Han Z, Feng Z. Clinical Manifestations, Diagnosis, and Management of First Branchial Cleft Fistula/Sinus: A Case Series and Literature Review. *J Oral Maxillofac Surg*. 2020;78(5):749-761. doi: <https://doi.org/10.1016/j.joms.2019.12.017>. Epub 2020 Jan 7. PMID: 32008991.
7. Lee HJ, Kim EK, Hong S. Sonographic detection of intrathyroidal branchial cleft cyst: a case report. *Korean J Radiol*. 2006;7(2):149-151. doi: <https://doi.org/10.3348/kjr.2006.7.2.149>. PMID: 16799277; PMCID: PMC2667589.

8. Akakpo K, Luck K, Chun RH. Use of a Cervicofacial Flap in Closing Large Defects Following Wide Local Excision of Complicated Preauricular Lesions. *Ann Otol Rhinol Laryngol*. 2020 9:3489420932605. doi: <https://doi.org/10.1177/0003489420932605>. Epub ahead of print. PMID: 32517509, 12, 3489420931167.
9. Liu W, Chen M, Hao J, Yang Y, Zhang J, Ni X. The treatment for the first branchial cleft anomalies in children. *Eur Arch Otorhinolaryngol*. 2017;274(9): 3465–70. <https://doi.org/10.1007/s00405-017-4648-y>.
10. Moroco AE, Saadi RA, Patel VA, Lehman EB, Wilson MN. Postoperative Outcomes of Branchial Cleft Cyst Excision in Children and Adults: An NSQIP Analysis. *Otolaryngol Head Neck Surg*. 2020;162(6):959-968. doi: <https://doi.org/10.1177/0194599820915468>. PMID: 32484763.

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