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Isolated First Branchial Cleft Anomalies of the
External Auditory Canal

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Data Collection B
Statistical Analysis C
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Patient: Female, 82-year-old

Final Diagnosis: First branchial arch malformative of the EAC

Symptoms: Mixed moderate hypoacusis and recurrent otitis media

Medication: —

Clinical Procedure: Canaloplasty and Thiersch graft

Specialty: Otolaryngology

Objective: Rare disease

Background: Branchial cleft anomalies are congenital aberrations of the first to fourth pharyngeal pouches. First branchial cleft anomalies are classified into 2 subtypes according to anatomical and histological features. Their diagnosis can be difficult and depends on radiological and histological findings. In contrast, the required treatment is surgical removal, owing to the high risk of infection or malignancy. This case report introduces a first branchial cleft anomaly in an older woman with exclusive involvement of the external auditory canal (EAC).

Case Report: This case report introduces a first branchial cleft anomaly in an 82-year-old woman with exclusive involvement of the EAC. She reported a history of mixed moderate hypoacusis and recurrent otitis media in the last year, without facial nerve involvement. Computed tomography and magnetic resonance imaging were performed to plan surgical treatment, which consisted of canaloplasty and Thiersch grafting. The histopathological examination on operative findings revealed a cystic lesion that was lined by cylindrical epithelium adjacent to the squamous cells, compatible with a diagnosis of first branchial arch malformative residues.

Conclusions: This is the unique case of first branchial cleft anomalies reported in an adult patient that exclusively involved the EAC. The onset of the disease was atypical, and surgery with the total removal of the lesion was the only possible treatment. Histopathology results revealed cylindrical epithelium not represented in the EAC, compatible with first branchial arch malformative residues. This rare condition is a potential diagnostic option that should be considered in the differential diagnosis of cysts of the EAC.

Keywords: Adult • Branchial Cleft Anomalies • Branchial Region • Ear CanalFull-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/936809>

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Background

Branchial cleft anomalies are congenital aberrations that develop from the incomplete obliteration during embryogenesis of the first to fourth pharyngeal pouches. They can result in true fistulae, cysts, or sinus tracts, particularly in the pediatric age group [1].

First branchial cleft anomalies (FBCAs) represent about 5% to 25% of branchial cleft anomalies [1]. FBCAs are usually defined by a smooth, non-tender, fluctuant mass that could involve the parotid gland [2,3] and facial nerve [4,5]. Two types of FBCAs, according to anatomical and histological features, have been identified [6]. Type I includes preauricular masses or sinus tracts localized anteriorly and/or medially to the external auditory canal, laterally to the facial nerve, and are composed only of ectoderm. Type II cysts, with ectoderm and mesoderm, are located at the angle of the mandible and/or submandibular region, lateral or medial to the facial nerve.

The diagnosis of FBCAs can be difficult and depends on radiological and histological findings. In contrast, the required treatment is surgical removal, owing to the high risk of infection or malignancy.

This paper aims to present a rare case of type 1 first branchial arch malformative residues in an older woman, with exclusive involvement of the external auditory canal (EAC). Its clinical and histological findings are described and discussed.

Case Report

An 82-year-old woman presented with a history of hearing loss and recurrent otitis media in the last year. She reported

ear fullness and swelling in the left EAC, and denied any vertigo or tinnitus. The otomicroscopic examination revealed a cystic mass occluding the left EAC. Pure-tone audiometry showed mixed moderate hearing loss, while the right one showed presbycusis.

Computed tomography showed soft tissue occupying the EAC without bone erosion signs (Figure 1A, 1B). These findings were confirmed by magnetic resonance imaging, excluding a connection with the facial nerve and parotid gland. Surgical treatment was planned, and excision of the lesion with canaloplasty and a Thiersch graft was performed. This was later found in the retroauricular region, where the surgeon performed a partial-thickness graft harvested with a knife. The skin was then cleaned, keeping only the epithelium and some papillary dermis, and the thin graft was positioned to cover the bony canal.

A histopathological examination revealed a cystic lesion lined by cylindrical epithelium adjacent to the squamous cells. Hence, a diagnosis of type 1 first branchial arch malformative residues was made (Figure 2A-2D).

After 9 months, the patient was disease-free, and pure-tone audiometry showed a mixed moderate hearing loss that did not worsen after surgical treatment. Therefore, she underwent radiological follow-up.

Discussion

FBCA is a congenital condition based on anomalies in the first branchial apparatus that develop into pharyngeal and oral structures [7]. FBCAs are mainly reported in children, and the diagnosis is frequently delayed and associated with a high rate of

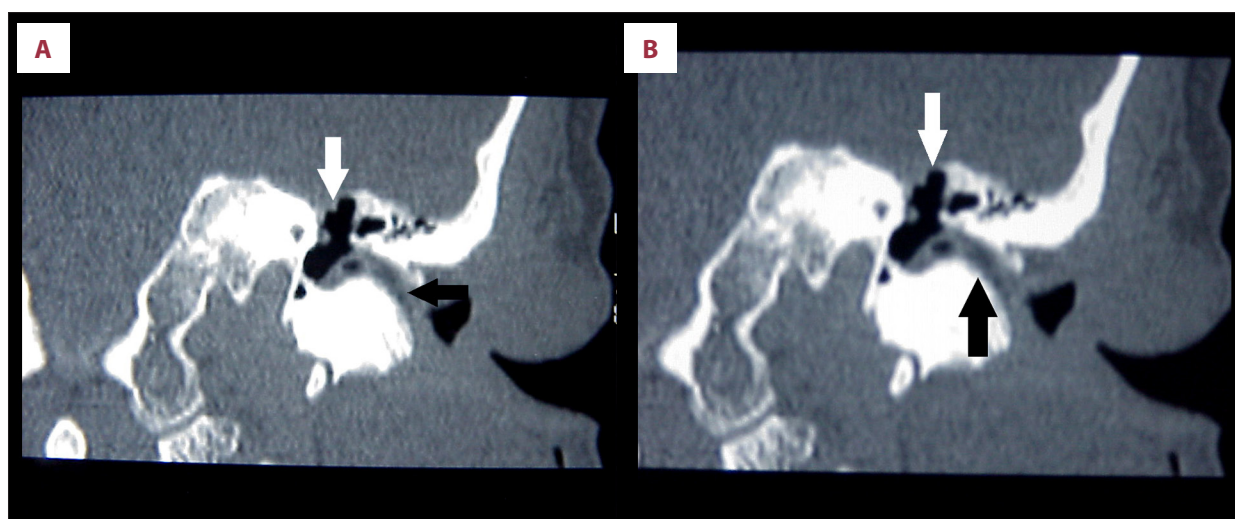


Figure 1. Preoperative computed tomography, coronal view. (A, B) Obliteration of the external auditory canal without bone erosion (black arrow) and no involvement of the middle ear (white arrow).

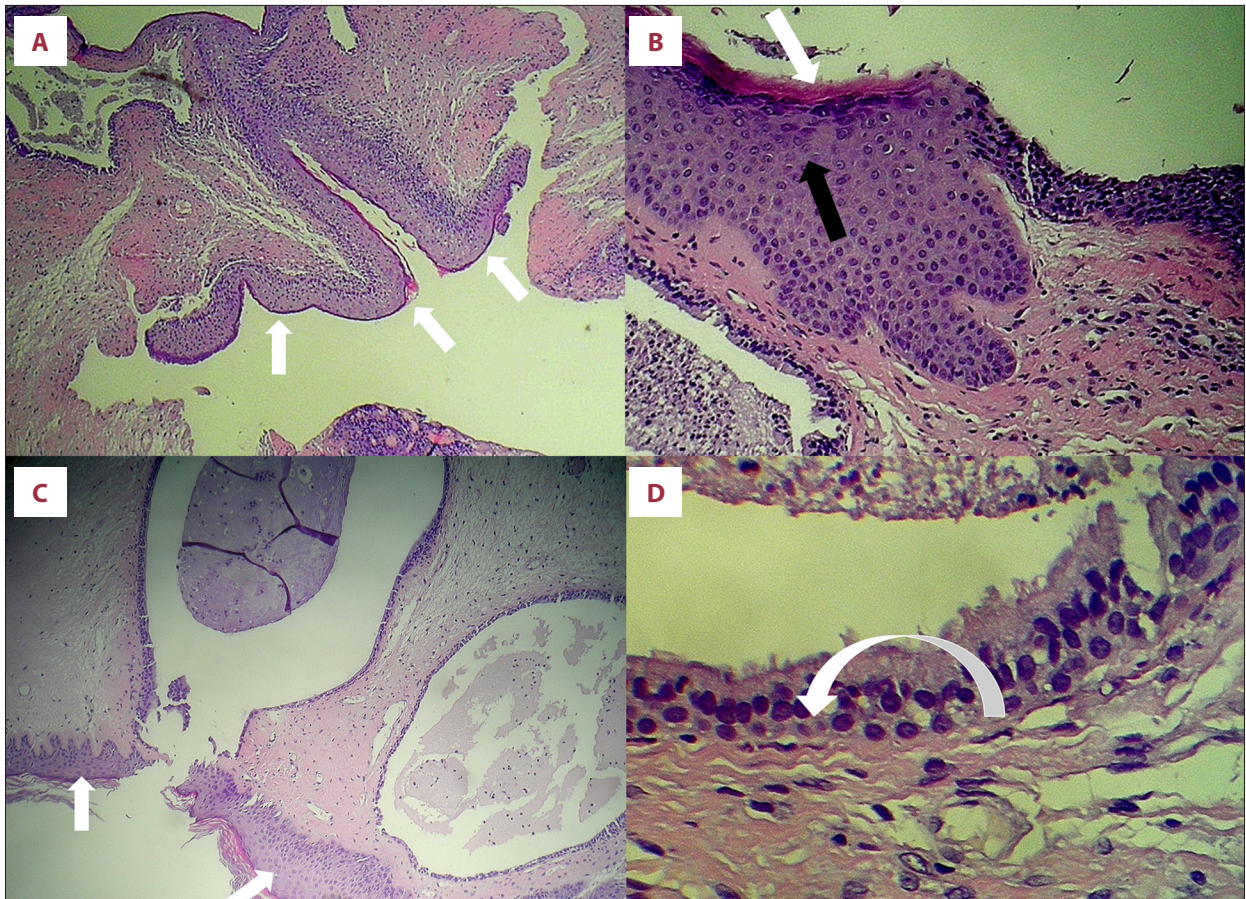


Figure 2. Histological findings, with hematoxylin and eosin staining. (A) Overview of the wall of the cysts (3 white arrows), at magnification $\times 10$. (B) Evidence of lining of squamous (white arrow) and cylindrical epithelium (black arrow), at magnification $\times 50$. (C) Evidence of cysts lined with cylindrical epithelium (white arrows) adjacent to the squamous cyst wall, at magnification $\times 25$. (D) Greater enlargement of the ciliated ciliary epithelium (curved arrow), at magnification $\times 100$.

complications, such as facial nerve paralysis [8]. FBCAs represent 8% of all branchial cleft anomalies. Generally, they result from incomplete obliteration of the clefts that arise between the branchial arches during embryogenesis and are usually located in the neck with other otologic or pharyngeal locations [9,10]. Clinically, FBCAs are characterized by recurrent otorrhea, pain in the parotid region, or a cervical mass anterior to the sternocleidomastoid [5].

In our case, the onset of the disease was unusual as it began in the patient at the age of 80 years, and symptoms were also uncommon, with swelling of the left EAC, without neck involvement [7]. Despite the patient's age, no facial nerve involvement was reported. To the best of our knowledge, there are no studies in the literature of cases of a first branchial cleft with the exclusive participation of the EAC [11].

Preoperative imaging is fundamental to determining the anatomic site and possible difficulties during surgery [2]. In our study, magnetic resonance imaging ruled out any potential

relationship with the parotid gland and eventual connection with the neck. Still, it did not provide adequate information for a definite differential diagnosis in the presence of purulent otorrhea [12]. Moreover, the exclusive EAC involvement ruled out any differential diagnosis with the malignant and benign parotid neoplasms and cysts, parotitis, second branchial cleft anomalies, and various pathological entities of the head and neck lymph nodes.

Surgery with total lesion removal was the only possible treatment [13]. A literature review showed that facial nerve lesions are reported in 13% of cases, and the rate of recurrence is about 3%, with a correct diagnosis [2].

In the present case, histopathological examination surprisingly revealed a first branchial cleft cyst due to the presence of cylindrical epithelium that was not represented in the EAC. This made possible the differential diagnosis of an epidermoid cyst of the EAC. The cylindrical epithelium is located only in the middle ear and not outside when the tympanic membrane is intact.

Moreover, in the present case, the absence of respiratory epithelium and associated lymphoid tissue was fundamental for a differential diagnosis of second branchial cleft cysts.

Conclusions

This is the unique case of type 1 FBCA reported in an adult patient that exclusively involved the EAC. This rare condition represents a potential diagnostic option that should be considered in the differential diagnosis of cysts of the EAC.

References:

1. Coste AH, Lofgren DH, Shermetaro C. Branchial cleft syst. [Updated 2022 Apr 14]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK499914/>
2. Liu H, Cheng A, Ward BB, et al. Clinical manifestations, diagnosis, and management of first branchial cleft fistula/sinus: A case series and literature review. *BMC Surg J Oral Maxillofac Surg.* 2020;78:749-61
3. Han Y, Yang R-Q, Hong L, et al. Unusual presentation of a first branchial arch fistula with maxillofacial infection: A case report. *BMC Surg.* 2021;21:306
4. Quintanilla-Dieck L, Virgin F, Wootten C, et al. Surgical approaches to first branchial cleft anomaly excision: A case series. *Case Rep Otolaryngol.* 2016; 2016:3902974
5. Hinson D, Poteet P, Bower C. Duplicated facial nerve trunk with a first branchial cleft cyst. *Laryngoscope.* 2014;124:662-64
6. Work WP. Newer concepts of the first branchial cleft defects. *Laryngoscope.* 1972;82:1581-93
7. Fanous A, Couloigner V, Gorphe P. Unusual presentation of a first Branchial cleft cyst associated with an abnormal bony canal – a case report. *J Otolaryngol Head Neck Surg.* 2020;49:32
8. Liu W, Liu B, Chen M, et al. Clinical analysis of first branchial cleft anomalies in children. *Pediatr Investig.* 2018; 2(3):149-53
9. Liu Y, Li T, Xue J, et al. First branchial cleft fistula presenting with internal opening on the Eustachian tube: Illustrated cases and literature review. *Int J Pediatr Otorhinolaryngol.* 2012;76(5):642-45
10. Devaraja K, Godkhindi VM, Bhandarkar AM, Lendoye W. First branchial cleft anomaly extending to parapharyngeal space. *BMJ Case Rep.* 2021;14:e244842
11. An extremely rare case of adult with first branchial cleft fistula: Case report. *Ann Med Surg (Lond).* 2021;70:102807
12. Marchioni D, Cuzzola E, Massone F, et al. Congenital postauricular swelling in a child. *Pediatr Dermatol.* 2002;19(3):246-49
13. Guo YX, Guo CB. Relation between a first branchial cleft anomaly and the facial nerve. *Br J Oral Maxillofac Surg.* 2012;50(3):259-63

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