International Journal of Pediatric Otorhinolaryngology Extra 12 (2016) 1–4

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology Extra

journal homepage: www.elsevier.com/locate/ijporl

Case report

First branchial cleft anomaly presenting with a complete duplication of the external auditory canal—A photo anatomic review

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ARTICLE INFO

Article history:
Received 3 November 2015
Received in revised form 15 December 2015
Accepted 16 December 2015
Available online 31 December 2015

Keywords:
First branchial cleft anomaly
Duplication external ear canal

ABSTRACT

First branchial cleft anomalies are an uncommon group of congenital malformations. Anatomically complex in origin, they present with an impressive array of signs and symptoms. There are several classification systems available, rendering them challenging in terms of preoperative assessment and surgical planning. The objective of this report is to describe the presentation, surgical treatment, and outcome of a child presenting with a complex cerumen containing duplication of the external ear canal running medial to the facial nerve. Diagnostic and management pitfalls in the approach to pediatric patients with first branchial anomalies are outlined. A review of the literature is presented.

1. Case history

A 4-year-old previously healthy boy presented with recurrent left neck abscesses, palpable persistent submandibular swelling and multiple previous incision and drainage procedures. Ultrasound performed early in his clinical course, demonstrated a phlegmon-like lesion with inflammatory change. T2 weighted MRI revealed a hyperintense tubular lesion within the left parotid gland with a tract extending cephalad to the anterior wall of the external auditory canal Peripheral enhancement was seen on contrast-enhanced, fat-suppressed T1 weighted images, suggesting the presence of inflammation. Decreased diffusivity within the lesion on diffusion-weighted images raised the possibility of purulent material.

Total left parotidectomy with excision of the lesion was planned. At surgery, a modified Blair skin incision was made and extended inferiorly. A large duplication of the cartilaginous external auditory canal was identified proximally tracking medial to the main trunk of the facial nerve towards the angle of the mandible. The tract was carefully dissected from the external auditory canal and the facial nerve. In order to free the portion of the tract that ran deep to the facial nerve, the tract was transected. Transection revealed a hair and skin lined tubular structure.

Histopathological assessment demonstrated a semicircular structure supported by elastic cartilage, and lined with squamous epithelium containing dermal appendage structures, sebaceous units, eccrine ducts, acellular keratin and occasional hair shafts which was consistent with a duplication of the external auditory canal or a Work type 2 branchial cleft anomaly. The patient recovered well and demonstrated no evidence of recurrence at subsequent outpatient follow up.

2. Discussion

The branchial apparatus occurs in all vertebrates during embryogenesis [1]. In the fourth week in utero, the fetus possesses six pairs of branchial arches, which form primordial structures of the face and neck. An outer ectodermal cleft and an inner endodermal pouch separate each arch [2]. During normal development, the first branchial arch forms the mandible, part of the upper jaw and parts of the inner ear [3]. The anterior parts of the first and second arches fuse together, obliterating the portion of the first branchial cleft between them. The remaining part of the first cleft forms the external auditory canal, whilst the inner pouch becomes the middle ear cavity and the eustachian tube [4].

Branchial anomalies arise when the branchial arches and their associated clefts or pouches fail to regress or develop normally [5]. Anomalous development may result in the formation of pits, sinuses, fistulae, cysts, or rarely complete duplication of the entire external auditory canal. Whilst most branchial cleft anomalies involve the second branchial complex, true first branchial cleft anomalies are relatively uncommon. They account for less than 8%

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http://dx.doi.org/10.1016/j.pedex.2015.12.001
1871-4048/

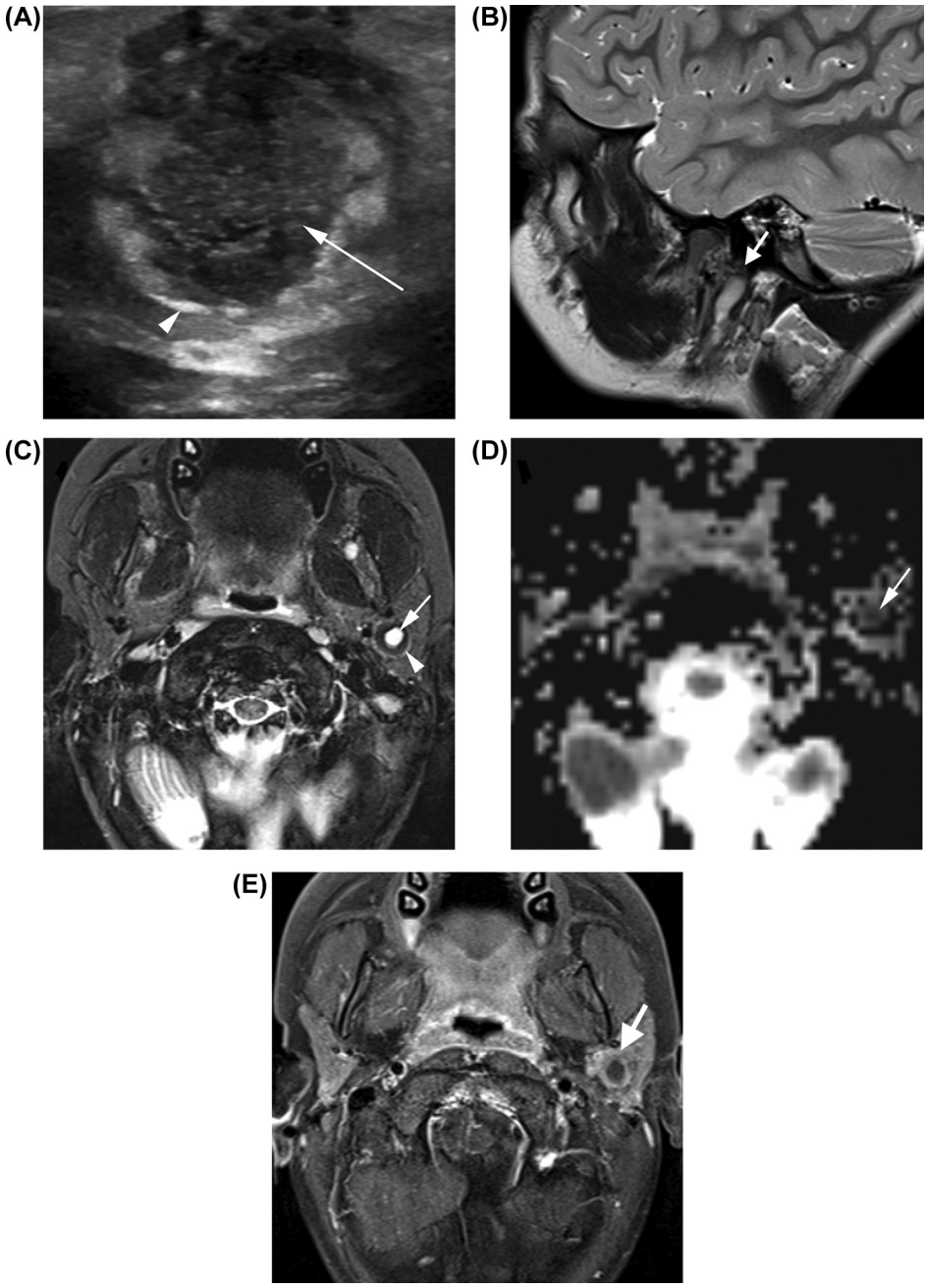

Fig. 1. (A) Ultrasound image demonstrates an irregularly shaped heterogeneous lesion (arrow) that contains multiple moderately echogenic mural foci (arrowhead), surrounding edema and inflammatory change. (B) Sagittal T2 weighted MR image shows a tubular lesion (arrow) along the posterior aspect of the left parotid gland. (C) Axial T2 short tau inversion recovery image demonstrates the hyperintense lesion (arrow) with hypo intense mural thickening (arrowhead) within the left parotid gland. (D) The diffusion weighted image apparent diffusion coefficient map reveals decreased diffusivity within the lesion (arrow) attributable to the keratinaceous content. (E) Axial fat-suppressed T1 weighted image shows peripheral mural enhancement (arrow).

of all branchial anomalies and have an estimated incidence of about one per million per population per year [6,7]. Interestingly, first branchial cleft anomalies are reportedly almost twice as common in females. Fistulas occur more frequently on the left side, but sinuses show no lateral pattern of preference. Duplication of the external auditory canal is likely the rarest of all first branchial cleft anomalies. Because of the paucity of cases, the true incidence of this particular subset of lesions is unknown.

Because of often-ambiguous presentations, misdiagnosis, inappropriate treatment and a high incidence of recurrence are common [8]. A number of systems have been developed in an attempt to assist diagnosis. The Work classification system based upon both anatomical and histological features is the most widely used, dividing lesions into Type 1, which are purely ectodermal, and Type 2 containing ectodermal and or mesodermal components. Type 2 lesions develop into cysts, sinuses or fistulae, and can contain either skin appendages or cartilage [9]. Arnot proposed an alternative classification system, dividing lesions into two types (confusingly also named Type 1 and Type 2). Arnot defined Type 1 lesions as those presenting with a lesion within the parotid gland due to buried cell rests in the anterior part of the arch, and Type 2 lesions as those presenting with a sinus ending at the external auditory canal arising from incomplete closure of the cleft itself [10]. Subsequently, Olsen classified branchial anomalies as cysts,