The Journal of Laryngology & Otology
June 2003, Vol. **117,** pp. 508–510

An undescribed first branchial cleft anomaly

Jason Gabriel Rockey, M.R.C.S., D. Gareth John, F.R.C.S.*, John Herbetko, F.R.C.R.†

Abstract
A variant of a type 2 first branchial cleft anomaly, in which accessory ossicles were found, is described. There follows a discussion of the classification of first branchial cleft abnormalities and how this particular case falls outside the standard classification. CT scanning is mentioned as the investigation that is most useful for defining these abnormalities.

Key words: Branchial Clefts; Embryology; Tomograph, X-Ray Computed

Introduction

The external ear canal is formed by the dorsal end of the first branchial cleft deepening into the mesoderm between the first and second arches, at about five weeks.[1] This is mainly composed of ectoderm, which meets with the endoderm of the first branchial pouch; together with the intervening mesoderm, this forms the tympanic membrane.

Anomalies of the external canal can thus be classified embryologically into aplasia, atresia, stenosis and duplication.[2] The particular anomaly in this case is of the duplication type, which was originally divided into type 1 and type 2 anomalies by Work.[3] Type 1 was considered to comprise ectoderm only and type 2 was made up of the skin (ectoderm) and cartilage (mesoderm).

Case report

A 51-year-old woman presented to her GP complaining of a left-sided facial weakness of five days duration. He referred her to the ENT department.

On examination in the clinic, she had a House-Brackman grade II left facial weakness. She had pits in the floor of both her external ear canals approximately midway between the external auditory canal and the tympanic membrane. The pits measured approximately 2 mm across. Her tympanic membranes and the rest of her ENT examination were normal. Neurological examination, including assessment of her balance was also normal. Her pure tone audiogram showed a mild bilateral sensorineural hearing loss. It was felt a computerized

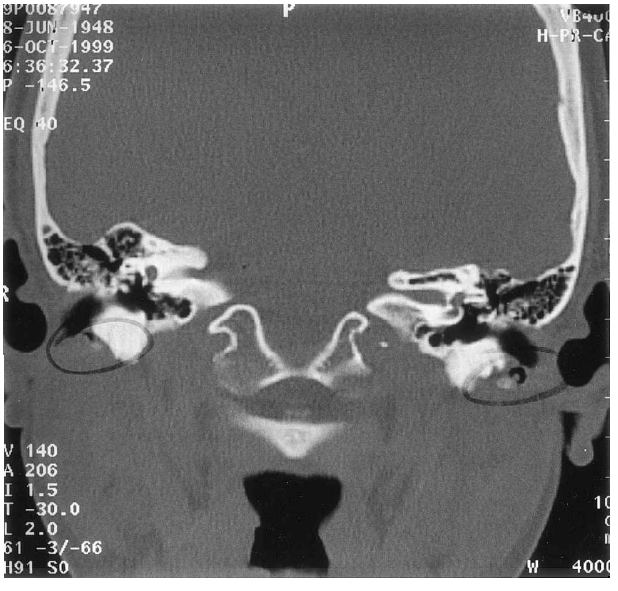

Fig. 1
Coronal section showing anomaly just lateral to bony external canal.

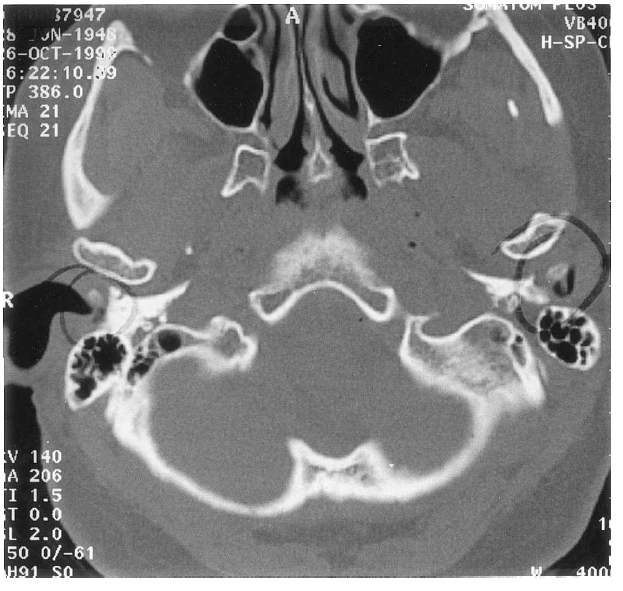

Fig. 2
Axial section showing anomaly with accessory ossicle.

From the Department of Otolaryngology, Queen Alexandra Hospital, Portsmouth and the Departments of Otolaryngology* and Radiology†, Poole Hospital, Poole, UK.
Accepted for publication: 17 March 2003.

tomogram of the defect was the best way to discern the nature of the abnormality and to see if it was associated with the facial nerve palsy on the patient's left side.

The defects were clearly demonstrated to be mucosal cavities on the floor of each external ear canal. They measured 3 mm in diameter and were just lateral to the bony part of the external canal (Figure 1). Each cavity contained a 3 mm accessory ossicle (Figures 1 and 2). The scan was otherwise normal. The course of the facial nerve was not involved in this congenital abnormality.

Conservative management was adopted, the facial nerve recovered completely and the patient remains well on telephone enquiry a year later.

Discussion

First branchial cleft abnormalities result in abnormalities of the external ear canals, which are categorized into aplasia, atresia, stenosis and duplication. Aplasia means absent external canals with fistulous openings connecting the middle-ear cavity to the skin (Figure 3). Atresia implies failure of canalization, which may result in the external canal being obstructed by fibrous or bony tissue. Stenosis refers to a narrowing of the canal. It is frequently seen in children with Down's syndrome. The middle ear may be abnormal in all of these situations.[2]

First cleft anomalies of the duplication type are rare. The authors note that, although there is a reported incidence of 1 in 10 000–20 000 cases of external canal atresia or stenosis,[4] the total number of duplication anomalies reported since 1963 is only 53.[3,5–8]

Duplication anomalies are of two forms. Type 1 anomalies are duplicated cartilaginous external canals that occur medial to the concha and may extend to the postauricular crease (Figure 4(a)). They pass anterior and deep to the ear lobe, superior to the facial nerve, and end at a bony plate at the level of the mesotympanum. The lesions have a squamous epithelial lining and skin adnexae indicating ectodermal origin.[3,5] They tend to present with a periauricular fistula, sinus or cyst, which may enlarge or discharge.[4,6]

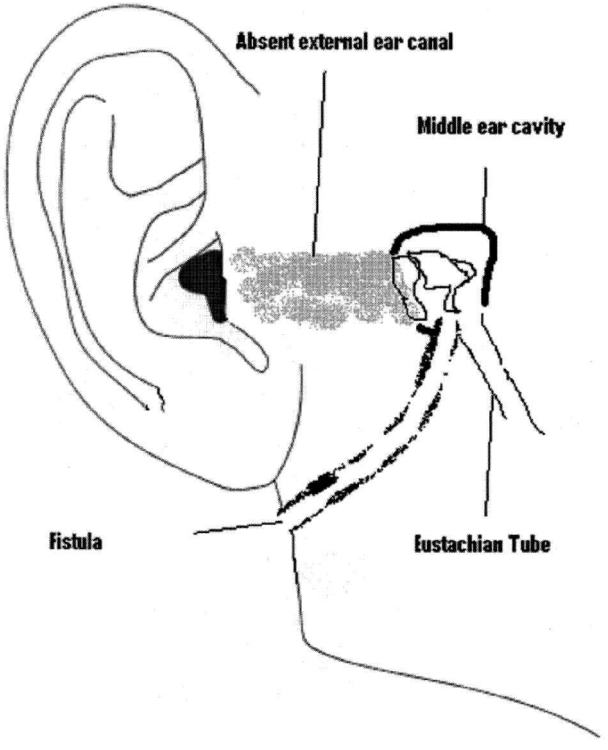

FIG. 3
Aplasia, with fistula to external skin.

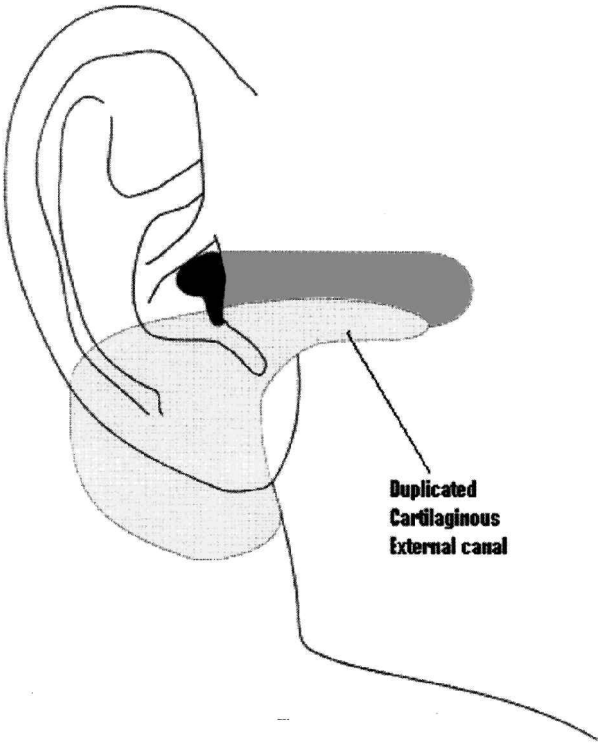

FIG. 4(a)
Type 1 duplication anomalies – duplicated external canals.

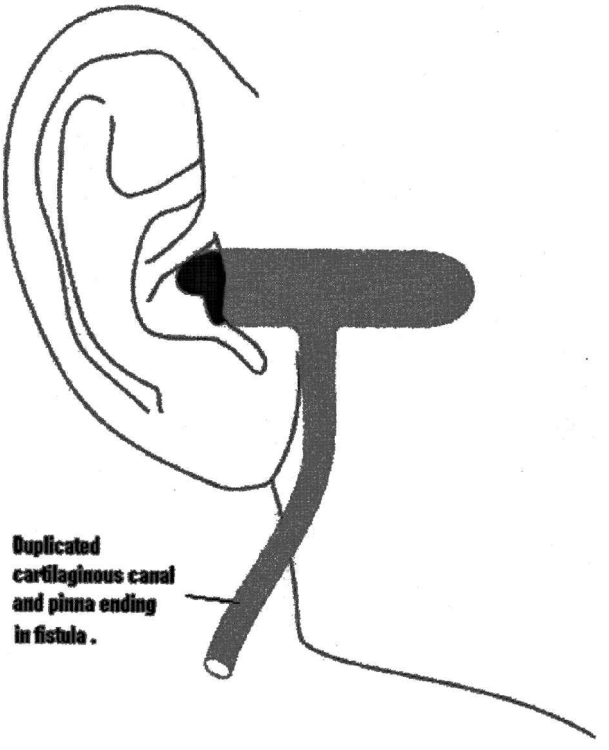

FIG. 4(b)
Type 2 duplication anomaly – may also end in fistula with skin externally.

https://doi.org/10.1258/002221503321892424 Published online by Cambridge University Press

Type 2 anomalies are considered to be a duplication of the cartilaginous external canal and pinna (Figure 4(b)). The lining of the tract has both skin and cartilage components microscopically and hence is said to contain ectodermal and mesodermal components.[5]

- **This paper presents a variant of a Type 2 branchial cleft anomaly with accessory ossicles that has not been previously reported**
- **The patient presented in middle age with a facial palsy that was coincidental**
- **The authors emphasize that CT scanning is the best way to characterize such congenital abnormalities**

In this patient, other than the sinus seen on the floor of the cartilaginous external canal, the rest of the external canal and the tympanic membrane were normal. The patient's middle-ear function was also normal, indicating this anomaly probably involved only the first branchial cleft. As the patient had bilateral pits, and the anomaly had given rise to almost identical otoscopic appearances, this anomaly was presumed to be of congenital aetiology.

Accessory ossicles are noted in this case. This contradicts the view of Work and others who have stated that type 2 duplication anomalies involve only skin and cartilage.[3,9] This patient's anomaly presumably has resulted from mesodermal condensations of either the first or second arches, or both, within the confines of a type 2 duplication anomaly.

Ossicles are normally located in the middle ear and are a derivative of the first branchial pouch. Thus, alternatively this anomaly may represent a variant of aplasia whose canal does not end in a fistula on the skin but ends, instead, in the external canal. Indeed it is possible this is a duplication of the first branchial pouch (tubotympanic recess). However, both these latter anomalies would usually require communication, or a remnant of communication, to the nasopharynx – and this is certainly not evident in this case.

Hence the variant in this case is hitherto unreported. It is probably a variant on the Type 2 duplication anomaly (? a type 2(a)) (Figure 5).

A final point to note from this case is that the abnormality in this asymptomatic case was made on scanning with computer tomography (CT). This has been used to delineate the size and extent of first branchial arch anomalies.[4] It was used specifically here to show that the facial nerve was not involved with the abnormality and therefore a conservative approach in this case could be undertaken confidently.

In conclusion, a new variant on type 2 duplication anomaly is presented, the diagnosis of which was only made possible through the use of modern imaging.

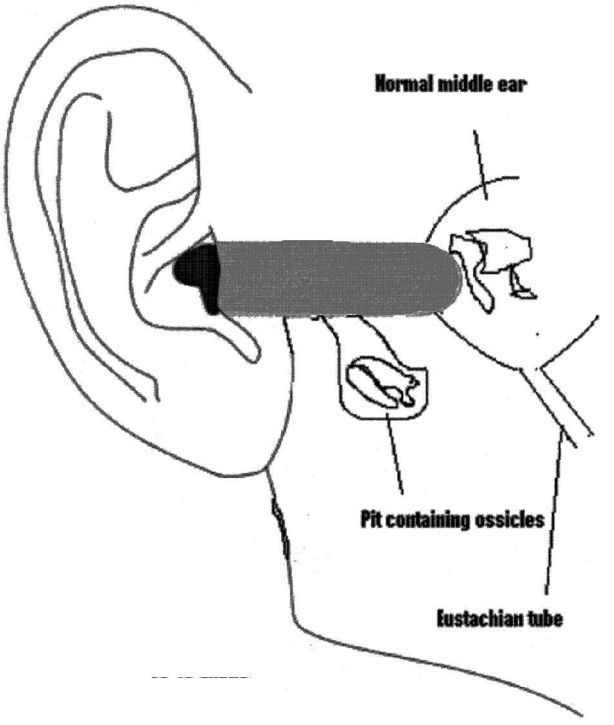

FIG. 5
The Type 2 (variant) anomaly in this case.

References

1 Sadler TW. *Langman's Medical Embryology*, 8th edn. Baltimore: Lippincott Williams and Wilkins, 2000
2 Karmody CS. A classification of the anomalies of the first branchial groove. *Otolaryngol Head Neck Surg* 1979;**87**:334–8
3 Work WP. Newer concepts of first branchial arch defects. *Laryngoscope* 1972;**132**:1581–93
4 Benton C, Bellet PS. Imaging of congenital abnormalities of the temporal bone. *Neuroimaging Clin N Am* 2000;**10**:35–53
5 Ford GR, Balakrishnan A, Evans JNG, Bailey CM. Branchial cleft and pouch anomalies. *J Laryngol Otol* 1992;**106**:137–43
6 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**:291–5
7 Stulner C, Chambers PA, Telfer MR, Corrihgan AM. Management of first branchial cleft anomalies: report of two cases. *Br J Oral Maxillofac Surg* 2001;**39**:30–3
8 Ohishi M, Torigoe Y, Nakazato K. First branchial cleft anomaly and congenital aural sinus – a case report. *Int J Pediatr Otorhinolaryngol* 1983;**5**:99–106
9 Aronsohn RS, Batsakis JG, Rice DH, Work WP. Anomalies of the first branchial cleft. *Arch Otolaryngol* 1976;**102**:737–40

Address for correspondence:
Mr Jason Gabriel Rockey,
c/o ENT Department,
Queen Alexandra Hospital,
Cosham,
Portsmouth PO6 3LY, UK.

E-mail: jason_rockey@hotmail.com

Mr J. G. Rockey takes responsibility for the integrity of the content of the paper.
Competing interests: None declared