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Clinical Record

A bifid intra-tympanic facial nerve in association with a normal stapes

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Abstract

A rare facial nerve anomaly was incidentally discovered whilst performing a tympanoplasty and ossicular reconstruction on a patient with an acquired unilateral conductive hearing loss. The nerve was seen to bifurcate and straddle a normal stapes superstructure as it ran posteriorly through the middle ear, a unique and as yet unreported combination. This case highlights the importance of vigilance regarding facial nerve anatomical variations encountered during middle-ear surgery thus avoiding inadvertent damage. The purported embryological mechanism responsible for such anomalies of the intra-tympanic facial nerve is discussed.

Key words: Facial Nerve, Abnormalities, Ear, Middle; Oval Window; Otological Surgical Procedures

Introduction

Localized bifurcation of the facial nerve in its intra-temporal course is a rare congenital malformation, an awareness of which is important during middle-ear surgery.[1,2] Splitting of the nerve may occur at any point but is most dramatically demonstrated where the limbs wrap around the stapes footplate and oval window in the medial wall of the tympanic cavity, this precise region being central to ossicular reconstruction for conductive hearing loss. Damage to the nerve at this point is therefore a significant risk, particularly for the unwary. We describe exactly such a case, providing a photographic representation of the anomaly that may confront the otologist.

Case report

A 48-year-old Lawyer presented to the ENT out-patient department with a long history of a non-progressive left-sided hearing loss associated with an intermittent mucous discharge. This unilateral deafness had been present since the age of nine years following a perforated tympanic membrane secondary to an attack of otitis media. Prior to this event the patient's hearing had been normal bilaterally. An attempt at reconstructing the tympanic membrane at the time had failed. The patient had since learnt to live with the disability, which only limited him when exposed to multiple simultaneous conversations; thus in the relative quiet of the courtroom he managed very well.

The patient re-presented due to an increased frequency of left-sided otorrhoea especially after swimming, despite using water precautions. Lawyer denied any associated tinnitus or vertigo and was otherwise well.

On examination Rinne's test was negative on the left with Weber's test also lateralizing to this side. Otoscopy revealed a normal right external auditory canal and tympanic membrane but on the left there was a subtotal perforation through which an eroded distal long process of the incus could be seen. There was no fistula sign or nystagmus and the cranial nerve examination was unremarkable. A pure tone audiogram confirmed a flat 50 dB conductive hearing loss on the left with normal thresholds on the right.

A per meatal tympanoplasty was performed. There was no active mucosal disease. The long process of the incus was confirmed as being eroded but the malleus and stapes appeared normal and were intact and mobile. Interestingly, a dehiscent and bifurcated facial nerve was identified which straddled the stapes as it coursed posteriorly (see Figure 1). Both limbs contained active motor fibres as confirmed by gentle nerve stimulation. The chorda tympani was identified running a normal anatomical course. The defect in the ossicular chain was bridged with Serenocement^{TM} (Corinzhian Surgical Ltd, Mansfield, Nottinghamshire, UK) and a myringoplasty was performed using an underlay tragal perichondrial/cartilage composite graft.

Post-operative recovery was uneventful. The patient reported a subjective improvement in hearing at follow up two weeks later. Subsequent follow up at six weeks confirmed a healed tympanic membrane with a closure of the air–bone gap to less than 10 dB.

Discussion

The facial nerve has the longest intra-osseous course of any cranial nerve and is thus more prone to injury than any other. Moreover its anatomy is subject to variations and anomalies which may be masked or exacerbated by disease. It is therefore important that otologists embark on middle-ear procedures with an awareness of these irregularities in order to prevent inadvertent damage to this vital structure.

Clinically relevant anomalies of the tympanic part of the facial nerve can be characterized by a congenital bony dehiscence with or without an irregular course. The latter

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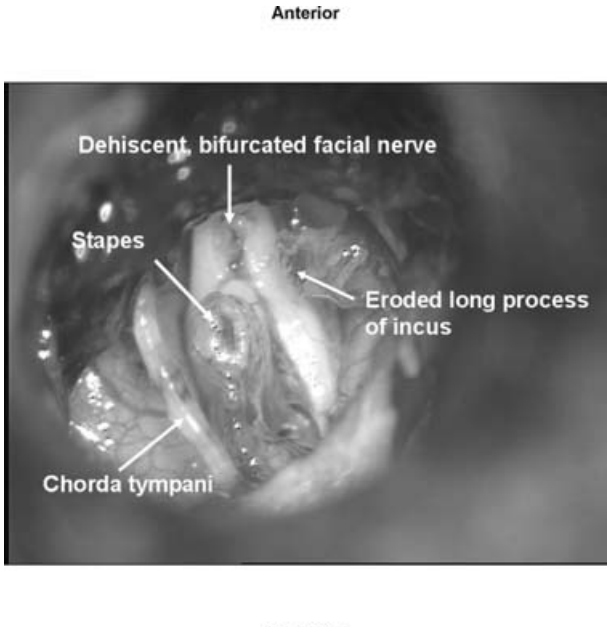

FIG. 1

A view into the left tympanic cavity through the operating microscope. The two limbs of the bifurcated horizontal part of the facial nerve can be seen straddling the stapes footplate as it courses posteriorly. Note the eroded long process of the incus.

may be subdivided according to whether the nerve runs a posterior course (1) superior to the lateral semicircular canal; (2) directly over the oval window with either a proximal bifurcation (with the limbs straddling the stapes footplate distally) or as a single trunk passing directly under the stapedial arch; or (3) infero-anterior to the oval window.[1,3]

The reported prevalence of dehiscence varies considerably depending on whether intra-operative, anatomical or pathological observations were performed, the latter yielding the highest incidence. Authors have quoted figures of between 15 and 74 per cent with the tympanic region being the most frequently affected area.[4] The incidence of associated facial nerve anomalies is much rarer, the literature being limited to case reports and small series. In addition these anomalies appear to be almost exclusively associated with congenital hearing loss.[1,3,5–7]

The tympanic facial canal originates in early fetal life as a sulcus on the medially placed primordial otic capsule and is later completed by a contribution from Reichert's cartilage (second branchial arch cartilage) which also gives rise to the stapes, the long process of the incus and the styloid process.[8] Embryologically the facial nerve (of the second branchial arch) initially runs posterior to its cartilage before turning anteriorly. This is manifest postnatally as the facial nerve lying posterior to the stapes and that part of the canal derived from Reichert's cartilage in the tympanum, eventually coursing behind the styloid before wrapping around it to continue anteriorly. Dehiscence can be explained by failure of fusion and/or ossification of Reichert's cartilage with the otic capsule. Likewise failure or delayed union of the stapes footplate and the otic capsule enables forward progression of the facial nerve to lie completely anterior to the oval window or traverse the stapes as in bifurcation. It is not surprising, therefore, that an anomalous course of the facial nerve is usually associated with congenital ossicular and other developmental abnormalities of the branchial arches.

Our case is unique in that there was no history of a congenital hearing deficit and no associated clinically overt branchial arch malformation. We found a normal stapes footplate and superstructure. The absent long process of the incus could be attributed to erosion from the initial childhood or subsequent recurrent episodes of inflammatory middle-ear disease; indeed the initial operation may also have contributed. The facial nerve anomaly was, however, unlikely to have been acquired.

- **This report describes an incidental focally dehiscent facial nerve with a bifurcation straddling the stapes footplate, discovered during surgical ossicular reconstruction**
- **This combination of a rare facial nerve anomaly in association with a normal stapes footplate and superstructure has hitherto been unreported**
- **The case highlights the importance of vigilance regarding facial nerve anatomical variation when embarking on middle-ear surgery**

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Dr J Ahmed takes responsibility for the integrity of the content of the paper.
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