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Congenital cerebrospinal fluid fistula through the inner ear and meningitis

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Abstract

Congenital deformities of the labyrinth of the inner ear can be associated with a fistulous communication between the intracranial subarachnoid space and the middle ear cavity. We describe seven such cases, six confirmed by high resolution CT and one by postmortem histological section. The seven patients all presented with meningitis although a cerebrospinal fluid fistula was demonstrated at subsequent surgery or postmortem. The lesions were bilateral in three patients, unilateral in three and probably bilateral in the postmortem case although only one temporal bone was obtained. In every case there was a dilated sac instead of the normal two and a half turn cochlea on the affected side and this was confirmed at surgery. The demonstration of the basal cochlear turn is of paramount importance in any deaf child presenting with meningitis. A true Mondini deformity with a normal basal turn and some hearing is not at risk of developing a fistula.

Key words: Cerebrospinal fluid, fistula; Meningitis; Tomography, X-ray computed

Introduction

Congenital deformities of the petrous temporal bone can give a fistulous communication between the intracranial subarachnoid space and the middle ear cleft. These anomalies result in egress of cerebrospinal fluid (c.s.f.) leading to cerebrospinal fluid otorrhoea or rhinorrhoea. Alternatively ingress of pathogenic organisms causes recurrent attacks of meningitis. As well as these spontaneous fistulae there are others which occur only if there is surgical interference with the stapes. Such a case is referred to as a 'stapes gusher'. Operative intervention in these patients has been undertaken in the past in an attempt to improve the hearing if there is a conductive deafness with a sufficient 'air–bone gap'. The fluid coming from the oval window may be perilymph initially, but if more than a few millilitres must be c.s.f. from a connection with the subarachnoid space. Schuknecht and Reisser (1988) discriminate between a 'gusher' for a pouring jet-like outflow and an 'oozer' for a milder welling type of flow. The route from inner to middle ear is thus almost always via the oval window but the pathway from subarachnoid space to perilymphatic space of the labyrinth is less clearly defined. We report seven cases of cerebrospinal fistula through the inner ear. All the patients presented with meningitis. The malformation of the inner ear and the fistulous pathway were defined by CT and confirmed by operative intervention in six cases (seven ears). One case succumbed to otogenic meningitis without any previous radiological study being done. One temporal bone from this patient was removed at postmortem for histological assessment although the deformity was probably bilateral.

Materials and methods

Seven patients, all of whom presented with meningitis were assessed. Their ages ranged from 2–24 years. Hearing assessment in all cases was made by experienced audiological physicians and pure tone audiometry as well as examination of the brain stem responses made if practical. All patients had thin section high resolution CT examination of the temporal bones; in axial and coronal planes in four cases, axial only in one, coronal only in one. The patient who died from a second attack of meningitis had no imaging investigation but one temporal bone was examined histologically. In the other cases there was surgical exploration of the middle ear on the abnormal side in three, in both sides in two and a tympanotomy is planned for the side thought to be the side of the fistula in one (*Case 2*).

Results

The malformations were entirely unilateral in three patients the opposite ear being audiologically and radiologically normal. Three patients had bilateral but fairly symmetrical deformities of the labyrinth shown by imaging. All the abnormal inner ears showed malformation of the cochlea with no coils of normal calibre present. Generally a widened basal turn communicated with a distal sac and had a wide communication with a dilated vestibule. There was no real evidence of any cochlear function in any of the malformed ears although it was felt that some hearing may be present on one side in the least

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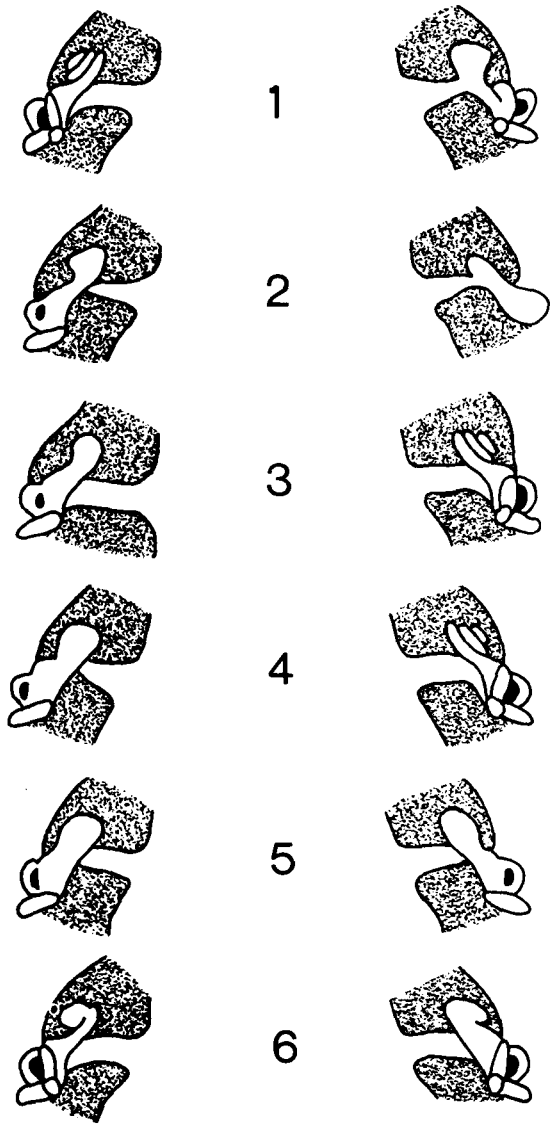

FIG. 1

Six cases of severe cochlear dysplasia as shown by axial CT. Unilateral lesions with normal appearances in the opposite ear are depicted in 1, 3 and 4. The other three are bilateral.

severely malformed bilateral case. Diagrams of the cochleovestibular deformities based on axial CT sections are depicted in Fig. 1.

Case 1

A female (aged 2 years) had her first attack of meningitis aged 13 months followed by another at 18 months. Following the second attack she had left-sided otorrhoea and examination under anaesthetic and myringotomy confirmed that this was cerebrospinal fluid. Imaging of petrous temporal bones by axial CT and coronal and lateral polytomography confirmed severe dysplasia of the left labyrinth. Exploration of the left middle ear showed an oval window in a deep niche and a dehiscence in the stapes footplate. The stapes was removed and the vestibule packed with muscle.

Case 2

A male (aged 2 years) who was profoundly deaf with no brain stem response at 100 dB had two attacks of pneumococcal meningitis. Axial and coronal CT demonstrated severe deformity of the labyrinth on both sides (Fig. 2). The presence of a fistula through the oval window in both ears was confirmed at operation. Removal of stapes on the right side provoked a torrential flow of c.s.f. The vestibule was packed with muscle which subsequently became displaced and had to be repacked. As the deformity in the left ear appeared even more severe than the right the stapes on this side was not removed but temporalis fascia was laid across the incudo-stapedial complex. The operative findings in this case are described in a separate publication.

Case 3

A male (aged 5 years) had three attacks of meningitis before axial CT revealed a severe deformity of the right labyrinth.

Case 4

A female (aged 23 years) had two attacks of meningitis in the space of 3 months the second of which was very severe. She had been deaf in the right ear since birth but axial and coronal CT revealed a severe dysplasia of the right labyrinth which was confirmed at tympanotomy when ballooning of the mucosa around the stapes was seen and a gush of c.s.f. resulted when the stapes was removed.

Case 5

A female (aged 8 years) with severe bilateral sensorineural deafness had had five attacks of meningitis. Computed tomography in the coronal plane showed bilateral labyrinthine dysplasia (Fig. 3). Prior to bilateral tympanotomies fluorescein was injected intrathecally and on both sides this was seen to be extruding around the footplate of the stapes although no actual fistula could be identified. Fascia from the tragus was packed around the stapes on each side and the patient had no further attacks for 18 months but then had two further episodes of pneumococcal meningitis. At two further operations both stapes were removed and the voluminous vestibules packed with fascia.

Case 6

A female (aged 5 years) deaf since birth although it was

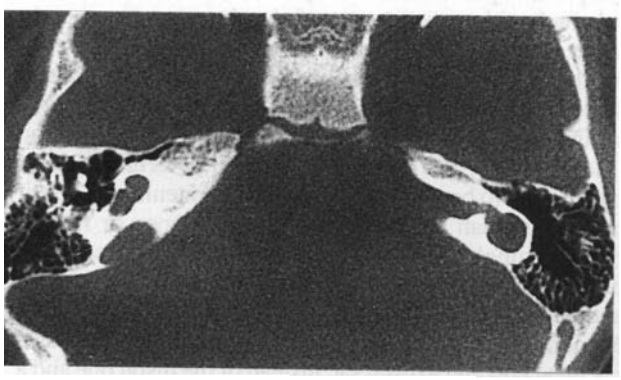

FIG. 2

Axial CT scan of Case 2 showing severe labyrinthine dysplasia on both sides.

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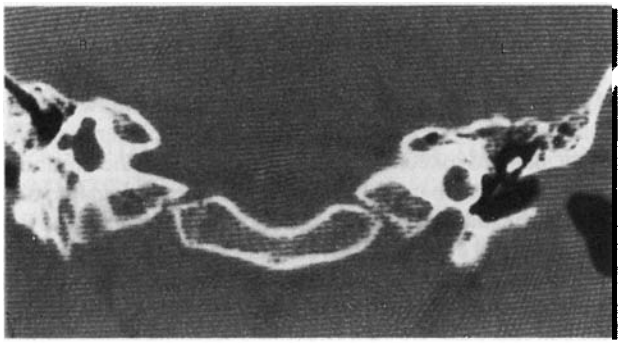

FIG. 3

Coronal CT scan showing bilateral labyrinthine dysplasia. On the right in this section vestibule, superior and lateral semicircular canals are seen to be dilated. On the left an amorphous cochlear sac is shown (Case 5).

thought there might be some cochlea function on the right. She had two attacks of meningitis within a 3-month period. The deformities of the labyrinths were less severe than in the other cases in this series and the basal turn of the right cochlea was thought to be of normal calibre i.e. a true Mondini deformity (Fig. 4). Electrophysiology studies seemed to confirm some response in the right ear only. A tympanotomy is planned for the left ear to see if a perilymph leak can be demonstrated by gently probing the stapes.

Case 7

A female (aged 3 years) had two attacks of meningitis from the second of which she died. She had been thought to be deaf from the age of 8 months but no definite hearing loss had been demonstrated. Eventually however the deafness was thought to be profound although electrophysiology was unsatisfactory. One temporal bone only was incompletely removed at postmortem. There was a single cystic structure in place of the cochlea and vestibule. There was no evidence of an endorgan or spiral ganglion although the VIIIth nerve was present and also the vestibular ganglion. The stapes had a large defect in the footplate (Fig. 5).

Discussion

Congenital malformations of the inner ear are sometimes characterized by a close association of the perilymphatic spaces of the labyrinth and the intracranial subarachnoid space. Arrest of development at an early stage in foetal life between the sixth and eighth weeks results in the anterior part of the labyrinth being represented as an amorphous cavity or primitive cochlea consisting of one dilated coil maybe with a distal empty sac (Jackler *et al.*, 1987). Such congenital deformities seem more likely to be associated with life threatening recurrent meningitis than cerebrospinal fluid otorrhoea or rhinorrhea. Meningitis was the presenting feature in all seven of our cases and proved fatal in one and almost fatal in another. None of these ears could be classed as a true Mondini deformity which consists of a normal basal cochlear coil and a distal sac instead of the distal one and a half turns. This can be confirmed by reading the original description (Phelps, 1986).

We believe that thin section high resolution CT is of the utmost importance in these cases to identify the severe cochlear dysplasia and should be undertaken after just one attack of meningitis in a child known to be deaf in at least one ear. The demonstration of a dysplastic sac instead of the normal two and a half turn cochlea warrants a tympanotomy to see if gently probing the stapes footplate induces a perilymph leak. On the other hand if CT reveals a basal cochlear turn of normal calibre and a distal sac only in place of the distal one and a half turns (a true Mondini deformity) then there is no risk of a spontaneous c.s.f. fistula or recurrent meningitis as no wide communication between subarachnoid and perilymphatic space can exist especially if the presence of some auditory function can be demonstrated. Auditory function would indicate the presence of a spiral ganglion and help to preclude a fistula.

Assessing the width of the basal turn on an axial CT scan can be difficult especially in bilateral cases. This problem is demonstrated well in *Case 6* (Fig. 4) in which there appears to be some hearing on the right side with a normal basal turn. It is hoped that the recurrent meningitis can be cured by surgery to the more deformed left ear and that the right ear can be left alone. An adequate description of the cochlear deformity needs to be given in all such cases especially the state of the basal cochlear turn. Eponyms such as 'Mondini deformity' have been used to describe such a wide range of malformations that they have become nonspecific and should be abandoned.

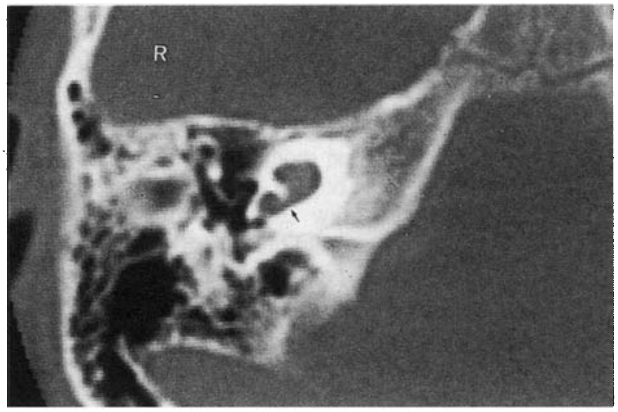

FIG. 4a

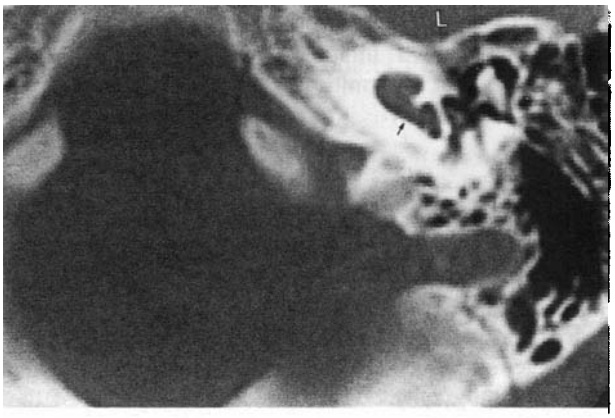

FIG. 4b

FIG. 4

Axial CT scans of Case 6: (a) right; (b) left. A distal sac replaces the distal one and a half turns on both sides but the basal turn of the cochlea (arrow) appears to be of normal calibre on the right although dilated on the left. This would seem to correspond with the audiometry which suggests some hearing on the right side only.

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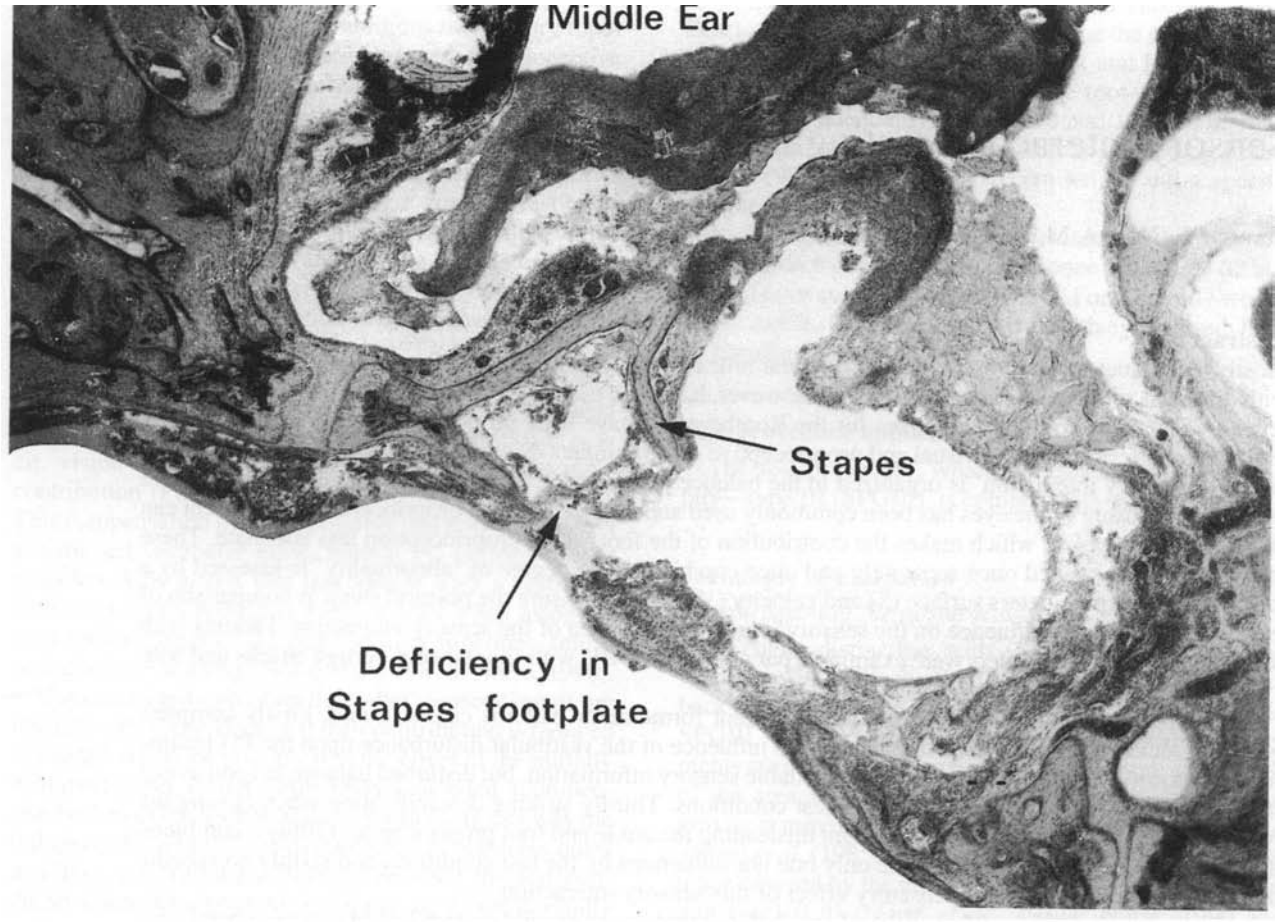

Fig. 5
Axial histological section of one temporal bone of Case 7 showing the hole in the stapes footplate.

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