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Large vestibular aqueduct syndrome and stapes fixation

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Abstract

An abnormally large vestibular aqueduct has a well recognized association with inner ear anomalies and it has been assumed previously to be a variant of a Mondini type of deformity (Shuknecht, 1980; Emmett, 1985). The sole radiological finding in some patients with progressive sensorineural loss has been a large vestibular aqueduct (Valvassori and Clemis, 1978; Valvassori, 1983), which is now accepted as a separate clinical entity, i.e. the large vestibular aqueduct syndrome (LVAS).

A case is presented which is believed to be the first reported with unilateral LVAS and stapes fixation and also the first stapes gusher described in association with LVAS.

Key words: Vestibular aqueduct; Stapes surgery; Hearing loss

Case report

A 21-year-old female presented to another centre with a right-sided hearing loss present since early childhood. On examination, she had a moderate to severe hearing loss on the right side with the Weber tuning fork test lateralizing to that ear. There was no other abnormality. The hearing proved to be of mixed type on pure tone audiometry (Figure 1). Exploration of her right middle ear revealed a fixed stapes, but the attempt to restore hearing had to be abandoned when a perilymphatic gusher developed following stapedectomy. She was referred to our unit where a CT scan of the temporal bone was performed. This demonstrated a large vestibular aqueduct syndrome (LVAS) on the right side (Figure 2) and no further management was advised.

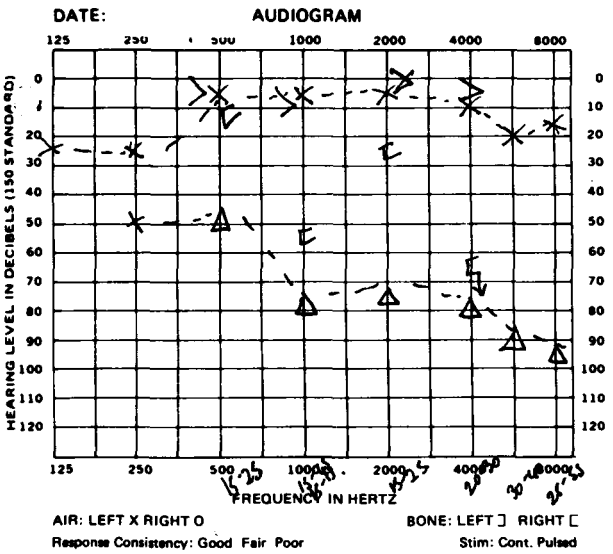

Fig. 1

Pre-operative pure tone audiogram revealing mixed type of hearing loss.

Discussion

Valvassori and Clemis (1978) described a group of 50 patients with an abnormally large vestibular aqueduct (LVA) in a study of 3700 consecutive cases who had tomography of the ear performed. Fifty per cent of these patients were teenagers or younger. Five years later, Valvassori (1983) reported a series of 160 patients with LVA found in 7000 cases referred for tomography, in which a similar proportion were children or teenagers. All but two of these 160 cases had some degree of sensorineural hearing loss. Valvassori (1983) concluded that LVA is the second most common inner ear anomaly after isolated dysplasia or hypoplasia of the lateral semicircular canal.

The vestibular aqueduct is a bony canal in the otic capsule which follows a J-shaped course from the medial wall of the vestibule to the posterior surface of the petrous pyramid. The developing vestibular aqueduct has a straight course parallel to the common crus and the endolymphatic sac overlies the sigmoid sinus in the posterior fossa. The otic labyrinth reaches adult dimensions by mid-term, but development of the posterior fossa

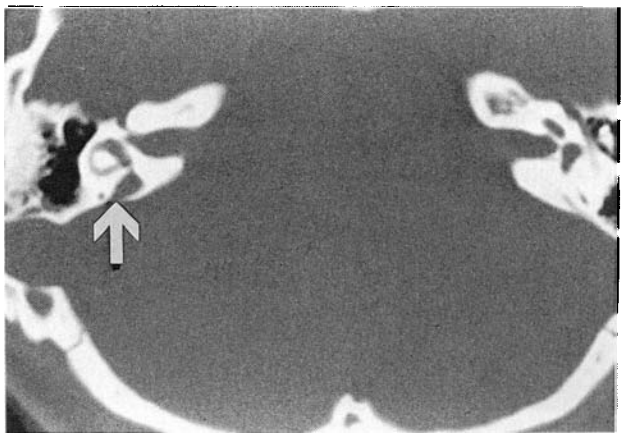

Fig. 2

CT scan of right temporal bone demonstrating LVAS (arrowed).

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structures continue, resulting in downward pull on the distal endolymphatic system, bending the vestibular aqueduct into the normal adult shape of an inverted 'J'. The normal vestibular aqueduct in the adult has a length of 10 mm and its diameter at the external aperture, in the posterior fossa, ranges between 0.5 and 1 mm. Any factor affecting the foetus during the early development of the inner ear may cause the endolymphatic duct to persist in its foetal (large) form (Arenberg, 1980; Valvassori, 1983).

The cause of progressive sensorineural hearing loss in LVAS is not known. The endolymphatic duct and sac, which have very thick and rugose walls in health may be thin walled, cyst-like or lined by flattened epithelium without any rugal or papillary folding in LVAS. This may affect inner ear homeostasis by changing the electrolyte balance (Shuknecht, 1980) or by allowing the accumulation of toxic materials around the hair cells (Gussen, 1985). Our patient did not have any clinical evidence of hydrops which is in keeping with the findings of other authors (Jackler, 1989; Levenson, 1989).

LVAS is responsible for progressive, bilateral sensorineural hearing loss, usually beginning in childhood or adolesence. There is a definite relationship between sudden decrease in hearing and head trauma which may be minor (Hill, 1984). This syndrome is significantly underdiagnosed. Many children with congenital sensorineural loss are not referred for radiological examination and if they are, LVAS can be easily overlooked by radiologists not sufficiently familiar with congenital temporal bone anomalies. The possibility of this syndrome should be considered in children with unexplained progressive sensorineural hearing loss, especially those who develop deafness after trauma to the head, however minor.

There is no known treatment for this syndrome (Valvassori, 1983). Co-existent fistulae have not been reported although an associated round window abnormality has been described and the potential for a perilymph fistula postulated (Belenky *et al.*, 1993). Surgery on the endolymphatic sac is contraindicated (Jackler and Brackmann, 1988).

This case is believed to be the first one reported with unilateral LVAS and stapes fixation. In hindsight, if a patient gives a history of lifelong hearing loss and this is found to be mixed in nature, CT scan prior to surgery may be indicated. Stapes gushers are associated with congenital, progressive and mixed hearing loss (Farrior and Endicott, 1971). The majority of gushers reported in the literature are in conjunction with congenital stapes fixation and an attendant sensorineural component in children (Wiet *et al.*, 1993). The probability that a sigificant sensorineural hearing loss is going to develop eventually, mitigates against performing stapes surgery on patients with LVAS. The pathophysiology of a cerebrospinal fluid (CSF) gusher at stapedectomy is unknown but a widened cochlear aqueduct or a defect in the fundus of the internal auditory canal, resulting in an abnormal connection between the CSF and perilymph, have been proposed (Wiet *et al.*, 1993). This is the first case reported of a stapes gusher occurring with a widened vestibular aqueduct and it is likely that the causal mechanism is similar to that postulated for two other congenital abnormalities of the temporal bone described here. This implies that stapes surgery should not be performed in patients with LVAS due to the possibility of a perilymphatic gusher and subsequent inner ear damage.

Normal active children inevitably suffer minor head injuries. It is our opinion that patients with LVAS will progress to total or near total deafness. Avoidance of contact sports may postpone, but not prevent in the long-term, the deafness that accompanies this condition.

Cochlear implantation should always be considered if the audiological criteria for this procedure have been met (Jackler *et al.*, 1987).

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