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Large Endolymphatic Duct and Sac Syndrome – A Case Report

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ABSTRACT

A 17-year-old girl with a history of hearing loss, presented with recurrent episodes of vomiting. CT scan revealed bilateral enlarged vestibular aqueducts and MR scanning confirmed the diagnosis of large endolymphatic duct and sac syndrome. This article looks into the anatomy and physiology of the endolymphatic duct and sac as well as possible explanations for the hearing loss associated with this syndrome.

Keywords: vestibular aqueduct, sensorineural hearing loss, inner ear, trauma, cerebrospinal fluid

INTRODUCTION

The enlarged vestibular aqueduct was first demonstrated radiologically in 1978[1] although its relationship with sensorineural hearing loss (SNHL) was already known before that time. This condition was first called the large vestibular aqueduct syndrome and was initially thought to be a variant of the Mondini type of inner ear deformity. It was later recognised as a separate clinical entity[2]. Computed tomography (CT) scan is currently the investigation of choice for this condition[3]. The enlarged endolymphatic sac component was only recently being demonstrated radiologically by magnetic resonance imaging (MRI) and the condition has since been renamed as the large endolymphatic sac and duct syndrome[4]. This is a case report of a patient with the above condition and is believed to be the first reported case in a local journal. The purpose of this case report is to raise the awareness of this condition amongst local physicians.

CASE REPORT

The patient is a 17-year-old girl with a history of deafness since childhood. This was presumed to be the result of neonatal kernicterus. She started to complain of recurrent vertigo and vomiting since 1989 and was admitted to hospital once.

Physical examination was essentially normal except for transient nystagmus towards the left side at the time of presentation. Audiometry showed bilateral severe to profound SNHL. There was no middle ear dysfunction. High resolution CT scan of the inner ear showed bilateral dilated vestibular aqueducts (Fig 1). The enlarged endolymphatic ducts and sacs were well demonstrated on MRI (Fig 2).

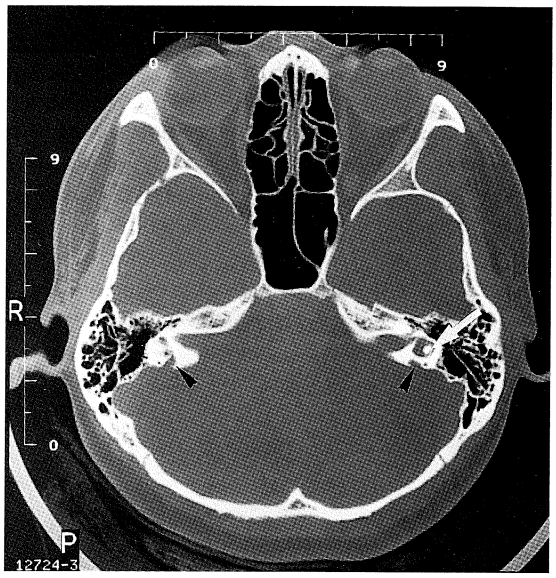

Fig 1 – High resolution CT scan in the axial plane showing the enlarged vestibular aqueducts (arrowheads) on both sides. The white arrow points to the lateral semicircular canal.

DISCUSSION

It has long been recognised that there is an association between SNHL and enlarged vestibular aqueducts demonstrated on histological studies. In 1978, Valvassori and Clemis demonstrated the enlarged aqueducts radiologically for the first time on inner ear tomography[1]. They coined the term "large vestibular aqueduct syndrome" to describe the condition. The vestibular aqueduct is defined to be enlarged if it had a diameter greater than 1.5 mm. The CT appearance of this condition was described by Swartz et al in 1985[3].

Originally, the large vestibular aqueduct syndrome was thought to be a temporal bone dysplasia possibly of the Mondini-type due to the frequent association of the enlarged vestibular aqueduct with inner ear anomalies[1]. It was redefined as a distinct clinical entity in children in 1989 following the work by Levenson MJ and colleagues[2]. They also noted that this condition was relatively common in children with SNHL and was probably significantly underdiagnosed.

MR imaging of this condition was described in 1995 by Harnsberger et al who demonstrated the direct visualisation of the endolymphatic duct using high-resolution fast spin echo (FSE) sequence[4]. The enlarged endolymphatic sac component, which prior to this could only be seen during surgery, was

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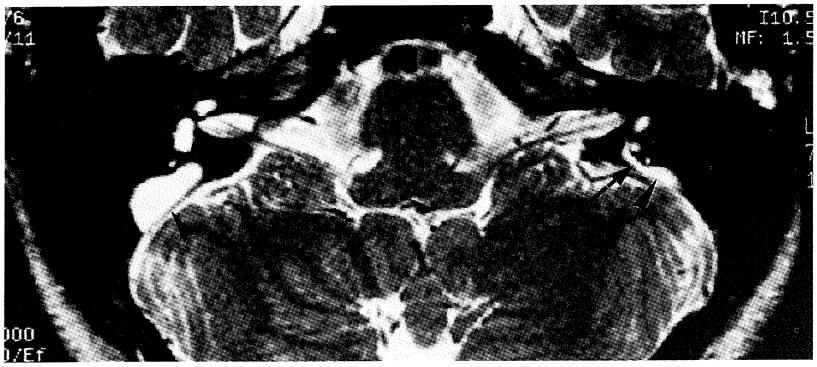

Fig 2 – High resolution FSE MR of the same patient at the level of the internal auditory canals showing the bilateral endolymphatic sacs (arrowheads) and the dilated left endolymphatic duct (arrow).

demonstrated radiologically for the first time. This condition was then renamed the "large endolymphatic duct and sac syndrome".

The large endolymphatic duct and sac syndrome is defined as the conbination of the clinical presentation of unilateral or bilateral SNHL in an infant or child and the radiological identification of the large endolymphatic duct and sac[4]. Bilateral involvement is twice as common as unilateral involvement. There is a female predominance[1]. The hearing loss is acquired rather than congenital and typically progress as a stepwise decrement often triggered by minor head trauma[5].

Anatomy and embryology

The bony vestibular aqueduct originates from the medial wall of the vestibule and extends dorsally for about 1 cm, opening on the dorsal surface of the petrous temporal bone within the posterior cranial fossa.

The endolymphatic duct courses within the bony vestibular aqueduct and connects the endolymphatic sac with the vestibular labyrinth and by means of the ductus reuniens with the cochlear duct (Fig 3). During the fourth week of gestation, three buds are formed from the primordial otocyst. Two of the buds developed into the cochlea and semicircular canals respectively. The third bud develops into the endolymphatic duct system. Early in embryogenesis the duct is short, straight, and proportionally much broader than at maturity (which is usually attained by 4 years of age). Subsequently, the duct narrows, elongates, and assumes the typical J-shaped form of adult life. It is believed that a teratogenic insult to the inner ear during the early stages of development caused the endolymphatic duct to persist in its large fetal form[5]. The same insult could theoretically affect the development of the other two buds and thus explains the frequent coexistence of vestibular aqueduct enlargement with other inner ear anomalies.

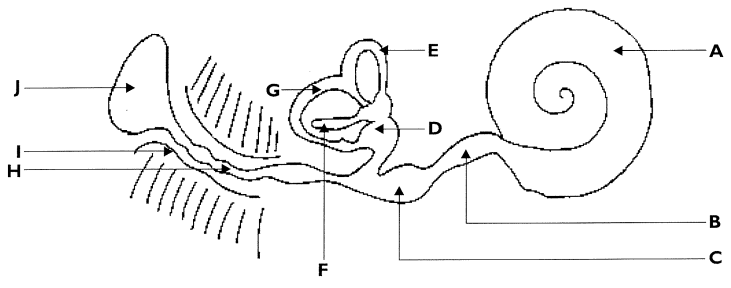

Fig 3 – Line diagram of the labyrinth with the endolymphatic duct and sac. A-cochlea, B-ductus reuniens, C-saccule, D-utricle, E-superior semicircular canal, F-lateral semicircular canal, G-posterior semicircular canal, H-endolymphatic duct, I-vestibular aqueduct, J-endolymphatic sac.

Physiology and pathogenesis

The exact role that the endolymphatic sac plays is still uncertain at present. It could be the site for active ionic exchange of endolymph with the cerebrospinal fluid[2]. Alternatively, it could be serving as a reservoir for endolymph and, due to its capacity for water absorption, may have a pressure-regulating role. It was noted that the endolymphatic sac has a surprisingly high protein content, making it markedly hyperosmolar in relation to the osmolarity of the endolymph contained within the remainder of the membranous labyrinth[2].

It is postulated that the enlarged patent endolymphatic duct placed the endolymphatic circulation at risk of reflux from the hyperosmolar endolymphatic sac content. Sudden fluctuation in cerebrospinal fluid pressure, eg. during minor trauma to the head, could have compressed the dural envelope surrounding the lymphatic sac and thereby forcibly pushing the hyperosmolar fluid within the endolymphatic sac through the dilated duct and into the endolymphatic circulation[2]. The hyperosmolar content would then damage the neuroepithelium of the cochlea, leading to hearing loss.

The abnormal transmission of cerebrospinal fluid pressure fluctuations to the inner ear via the wide vestibular aqueduct could also be a contributor to hearing loss[5]. Normally, the inner ear is buffered from any rapid intracranial pressure changes by the narrowness of the vestibular and cochlear aqueducts. When the vestibular aqueduct is enlarged while the cochlear aqueduct is normal in size, any rapid fluctuation in cerebrospinal fluid pressure, as in the case of head trauma, might create transient force imbalances across the cochlear partition. As a result, shearing forces could be created causing damage to the membranous labyrinth.

CT scan is currently the imaging modality of choice but high resolution FSE MR may become superior to CT in the assessment of this condition[4]. Currently, endolymphatic surgery has not been proven to be useful and could even be detrimental to hearing[5]. However, upon diagnosis of the condition, the parents of the child should be advised on the following points:

1) Fitting of hearing aids as soon as possible if hearing loss is present.
2) Speech/language intervention by therapist during important language learning years so that the child can develop good oral speech and language skills.
3) Head protection. As it is known that even minor head trauma could lead to irreversible drop in hearing, the child should avoid contact sports like soccer, hockey etc. Using protective head gear for sports like cycling should be encouraged.

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Corrigendum

The Editor of the SMJ wishes to apologise for the oversight in the following article:

Multiple Organ Failure and Septic Shock in Disseminated Tuberculosis (Singapore Med J 1999; 40:176-8).

The second author should in fact be C K Liam instead of K L Chong.