Large Vestibular Aqueduct Syndrome

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Large vestibular aqueduct syndrome (LVAS) may cause early-onset and progressive fluctuating sensorineural hearing loss (SNHL), and often leads to delayed speech in children. Previous head trauma, baro-trauma, air travel or an upper respiratory infection may lead to loss of hearing function which might recover spontaneously or develop into profound hearing loss. Early diagnosis and hearing aids are helpful to prevent speech disturbance. We present a 5-year-old boy who had had impaired hearing and speech disturbance since childhood. Fluctuating hearing function was demonstrated at another hospital when he was 2 years of age. CT scan of temporal bone revealed enlarged bilateral vestibular aqueducts. He had been admitted to hospital several times since 2 years of age. On each previous admission, he presented with vertigo and deafness; symptomatic treatment included mannitol and low dose prednisolone. He recovered within 3 days; however, it is unclear whether his recovery was due to the medication or whether it was spontaneous. We present this case to discuss the diagnosis, treatment, and probable pathophysiology of this syndrome. **(Mid Taiwan J Med 2003;8:327-31)**

Key words

large vestibular aqueduct syndrome, sensorineural hearing loss, vertigo

INTRODUCTION

The earliest description of large vestibular aqueduct syndrome (LVAS) was in 1978 by Valvassori and Clemis [1]. The enlarged aqueduct is the most common abnormality of the inner ear in adults and children with SNHL [2], and is the factor which contributes most to SNHL of children [3]. Patients with this disorder often have slight hearing impairment in the prelingual period, which gradually worsens to profound hearing impairment by adulthood [4]. Patients with hearing impairment in the prelingual period suffer from speech disturbance [5]. Differential diagnosis in children with SNHL is important; therefore, we present this case to discuss the diagnosis, treatment, and possible pathophysiology of LVAS.

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CASE REPORT

A 5-year-old boy with no systemic disease, hereditary disease or special problems during the prenatal period or labor initially presented to us with intact drums, but different levels of SNHL according to a test at another hospital when he was two years old. He had been wearing hearing aids and undergoing speech therapy since that time. His medical history included a prior head injury and an upper respiratory infection. In February 2002, pure tone audiometry (PTA) demonstrated moderate hearing loss in his left ear and profound hearing loss in the right. We performed a systemic survey beginning in 2002 but the laboratory data were normal; however, CT scan of the temporal bone showed enlarged bilateral vestibular aqueducts (Fig. 1). LVAS was the preliminary diagnosis. During a one-year period (2002 to 2003), regular follow-up revealed fluctuating bilateral hearing function (Figs. 2, 3), but no vertigo or vomiting occurred until January 2003. At that time he developed acute vertigo and

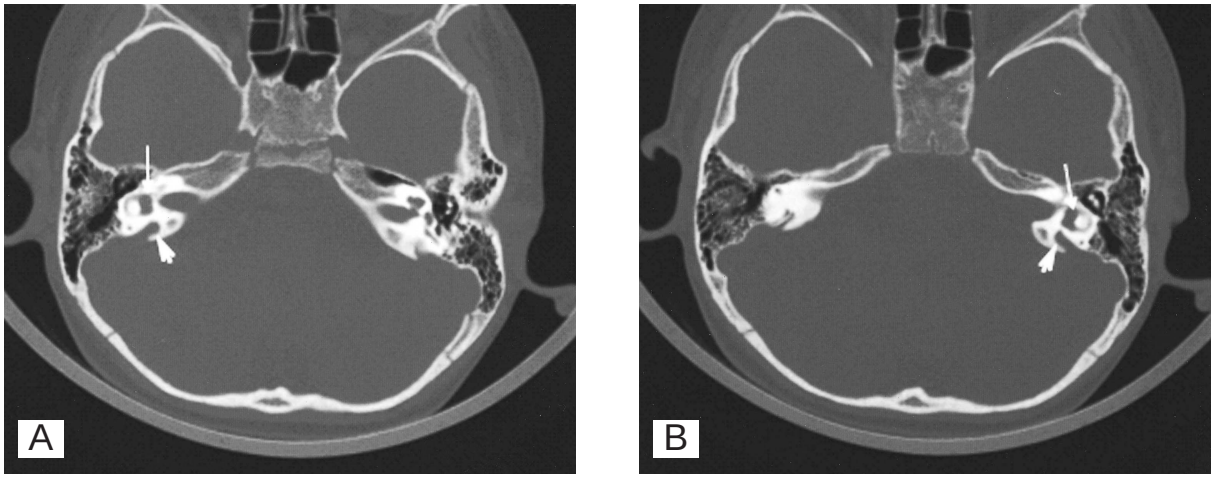

Fig. 1. Mastoid CT (axial view). Enlarged vestibular aqueduct (short arrow) and widening vestibule (long arrow). A: Right. B: Left.

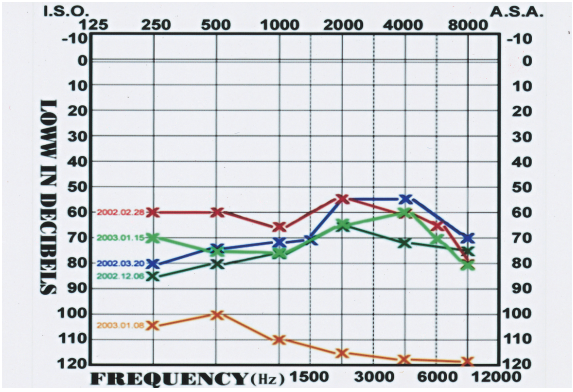

Fig. 2. Fluctuating hearing impairment of left ear.

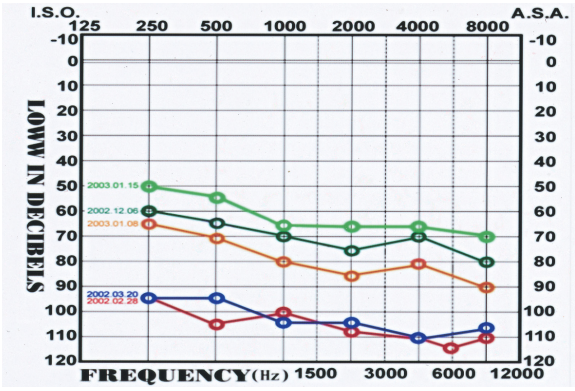

Fig. 3. Fluctuating hearing impairment of right ear.

complained of worsening deafness in both ears so was admitted to our hospital. Intact ear drums with no obvious nystagmus or neurological deficits were noted on physical examination, but profound hearing loss in the left ear and moderate hearing loss in the right were noted by PTA. Electronystagogram and a caloric test showed left canal paresis indicative of LVAS. We treated him with low dose mannitol and prednisolone. His hearing function recovered partially and became more stable.

DISCUSSION

The vestibular aqueduct originates from the medial wall of the inner ear vestibule and extends its orifice to the cerebellar aspect of the petrous pyramid. It has an inverted "J" shape and forms a bony canal which connects the endolymphatic sac and labyrinth of the vestibule. The vestibular aqueduct contains vessels and the endolymphatic duct, which connects the endolymphatic sac and the ductus reunions of the cochlear duct. The endolymphatic sac has tall ruga epithelial cells which regulate endolymphatic pressure and endolymph concentration [1,6,7].

The earliest description of LVAS was reported by Valvassori and Clemis. They reported that the incidence of enlarged vestibular aqueduct among 3700 patients with SNHL was 1.5% [1]. It is also the most common abnormal image finding in children with SNHL [3,8]; statistics from abroad show a female predominance [1,6,9], but statistics in Taiwan show a male predominance [10]. SNHL typically occurs in both ears and over

half of the patients with this syndrome have other inner ear abnormalities, including enlarged vestibules and abnormal semicircular canals. Some patients have maldevelopment of the inner ear which is considered a variant of Mondini dysplasia [4,8].

Impaired hearing in the prelingual period is the main symptom of LVAS. Initially, bilateral unsymmetrical slight to moderate SNHL may worsen at a speed of 4 dB per year. The bilateral hearing function may be unsymmetrical to 30 dB. Eventually, profound hearing impairment in both ears is usually noted in adulthood [4]. A history of minor head trauma is also common [6].

Embryologically, the otocyst appears during the late 4th week of gestation, and the endolymphatic duct appears from the diverticulum located behind the otocyst. These structures are initially wide and short, but enlongate and are restricted by development of the embryo. The semicircular canals and vestibular system appear in the 5th week. The membranous system of the inner ear is already developed by the 8th week. The final size of the inner ear labyrinth is fully developed by the middle trimester. It is hypothesized that failure of the endolymphatic ducts to narrow in the 5th week of gestation causes LVAS [1,9,11]. However Pyle [3] believes that hypoplasia of the vestibular aqueduct from the neonatal period to the age of 4 years is a contributing factor of SNHL around the perilingual period.

Another theory was proposed by Gussen [12] who stated that the change in intra-endolymphatic duct pressure causes the destruction of cochlear cells and the walls of the vestibular aqueduct; Levenson et al [6] indicated that the enlarged vestibular aqueduct can not prevent regurgitation from endolymphatic sacs which contain a high concentration of fluid. The regurgitation destroys the basal portion of the cochlea (inner hear cells) and causes high tone SNHL; it even leads to the rupture of the Reissner's membrane and basal membrane which damages the hearing-vestibular system, contributing to hearing impairment and acute vertigo, as in our patient. Okamoto et al [7] reported that a high CT density over the endolymphatic sac and duct areas compared with CSF predicts a high concentration of fluid in the endolymphatic sac which might hurt inner hear cells in the cochlea by regurgitation, causing sudden deafness. He suggested that medication which lowers brain pressure, such as mannitol, subsequently decreases the pressure in the endolymphatic sac, thereby improving inner ear injuries. Belenky et al [13] consider fragile cochlea and abnormal oval window to be responsible for perilymphatic fistula caused by head injury or increased intracranial pressure, which contribute to hearing impairment and acute vertigo [6,10].

It is still unclear whether LVAS is a congenital or hereditary disease. LVAS may be an autosomal recessive, autosomal dominant, or multifactorious syndrome, and may be related to thyroid dysfunction. The gene for LVAS overlaps the PDS (Pendred syndrome) gene (7q31); thus, patients with PDS often present with enlarged vestibular aqueducts. Cemers et al reported that all patients with the PDS gene in his study showed enlarged vestibular aqueducts [14].

We should be very alert to this syndrome when evaluating children with SNHL and fluctuating hearing impairment. The differential diagnoses of LVAS include sudden deafness, Meniere's syndrome, and other congenital hereditary hearing impairments; Mafong et al [15] reported that imaging studies are more valuable than laboratory studies for diagnosing SNHL in children. Although Valvassori and Clemis [1] reported that the lateral view of polytomographic images can diagnose this syndrome, polytomography of temporal bone is never as clear as CT scan. Moreover, patients who undergo polytomography receive higher dosages of radiation than those who undergo CT scan of temporal bone. The diagnostic criterion is a mid-aqueduct larger than 1.5 mm in diameter.

There is no standard LVAS treatment. Surgical methods do not improve hearing impairment, and often contribute to further hearing loss. Endolymphatic sac occlusion which blocks the regurgitation of endolymphatic fluid

has a poor outcome; Emmett [8] made a shunt between the endolymphatic duct and the subarachnoid space to release intra-endolymphatic duct pressure, but no improvement in hearing was found. Furthermore, deafness has been reported after surgical intervention [9]. Even though there are no effective treatments for recovering hearing function, early diagnosis, prevention of head trauma and activity restriction can slow down hearing impairment [3,10]. Hearing rehabilitation such as hearing aids and cochlear implants is very helpful around the perilingual period and long term follow-up of patients with LVAS is necessary.

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大前庭導水管症候群

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大前庭導水管症候群是早發性幼兒感音性聽力障礙的可能原因之一，常因而構成兒童語言發展之障礙，其聽力障礙程度常在頭部外傷或上呼吸道感染後惡化，隨後會逐漸緩解，但可能又復發。早期診斷出此疾病並輔以聽能治療如配戴助聽器，可避免兒童的語言發展受到影響。本部曾經歷一例五歲男童，自幼即有聽力及語言障礙並接受語言治療，追蹤聽力呈波動性變化，經高解析度顳骨電腦斷層診斷爲兩側大前庭導水管症候群；患者並曾因反覆發作之頭暈嘔吐及突發性耳聾而住院治療，患童頭暈及聽力急性惡化現象，經藥物治療後，眩暈症狀明顯改善且惡化之聽力也得到部份恢復，因本例之罕見及其在小兒感音性聽障中重要之鑑別診斷角色，提出本病例報告，並對其診斷、治療及可能的病理生理機轉作文獻回顧。（中台灣醫誌 **2003;8:327-31**）

關鍵詞

大前庭導水管症候群，感音性聽力障礙，眩暈

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