Association of Chiari I Malformation and Cerebellar Ectopia With Sensorineural Hearing Loss

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Abstract: We aimed to examine the prevalence of cerebellar tonsil ectopia and Chiari 1 malformation in sensorineural hearing loss (SHL) that has, to the best of our knowledge, not been studied previously. Magnetic resonance imaging records of 166 subjects with SHL and 50 controls without known otologic disturbances were included in the study. A tonsils descent more than 2 mm was assumed as cerebellar ectopia, and a descent equal to or more than 5 mm was assumed as Chiari 1 malformation. A tonsil descent group was also formed by summation of both groups. Transverse diameters of bilateral intracranial vertebral arteries and transverse sinuses were also measured, and all parameters were analyzed using appropriate statistics. A significant difference of frequencies of Chiari 1, ectopia, and tonsil descent was detected between patients and controls. In comparison of cerebellar ectopia and Chiari 1 groups, SHL did not show any significant difference. The left lateral sinus diameter showed positive correlation with tonsil descent. There was no significant correlation for the diameters of other vessels. A powerful correlation was detected between SHL and age. In addition, right and vertebral artery diameters showed positive correlations with age. Chiari 1 malformation and cerebellar ectopia showed an association with SHL. These patients should also be evaluated for otologic disturbances. Further high-resolution magnetic resonance imaging studies to explain the exact cause of this currently unknown association seems required.

Key Words: Chiari 1 malformation, MRI, sensorineural hearing loss

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Radiographically, Chiari 1 malformation (CM 1) is defined as a 5-mm or more caudal descent of cerebellar tonsils below the level of foramen magnum.[1,2] Tonsillar descent less than 2 mm below the foramen magnum is considered unimportant; however, sometimes cerebellar tonsils are noted to be displaced more than 2 mm but less than 5 mm below the foramen magnum. This condition is named *cerebellar ectopia* that is estimated to be present in up to 14% of otherwise healthy persons.[2]

Displaced tonsils may cause dysfunction of brainstem and/or lower cranial nerves resulting in visual symptoms, otologic disturbances, spinal ataxia, dysphagia, sleep apnea, nystagmus, and dysarthria.[2,3] Sensorineural hearing loss (SHL) had been reported accompanying CM 1 in a few previous studies, and a number of case reports, still the direct causation, remains speculative. Milhorat et al[3] reported 16 patients with SHL in 24 who had complete otologic examination because of dizziness and vertigo in their study group with 364 CM 1 patients. In the study of Sperling et al,[4] fluctuating hearing loss was reported by 9 (56%) of 16 CM 1 patients. Simons et al[5] had performed a retrospective evaluation of 113 children with SHL and found 6 patients having Chiari malformation (5.3%) that was much more common than the general population, in which the incidence of CM 1 has been cited to be between 0.56% and 0.77%.[4,5] Besides, there are only a few case reports on the association between hearing loss and CM 1 published in the literature. Although a few aforementioned reports are present on this subject, to the best of our knowledge, ectopia of cerebellar tonsils along with CM 1 in SHL has not been studied previously. In the present study, we aimed to examine the prevalence of cerebellar tonsil ectopia and CM 1 in SHL and discuss a probable correlation between them.

MATERIALS AND METHODS

Magnetic resonance imaging (MRI) records of 166 subjects, 79 men (47.6%) and 87 women (52.4%) (aged between 8 and 85 years; mean [SD], 50.03 [17.12] years) referred to the radiology department between January 2010 and March 2012 for SHL were included in the study. An age- and sex-matched control group was constituted from the brain MRI of 50 patients acquired for different reasons such as headache, psychiatric complaints, and vision problems. The control group has 21 men (42%) and 29 women (58%) (aged between 10 and 88 years; mean, 46.88 [17.83]). The controls had not applied for hearing loss to the otolaryngology department.

Age, sex, hearing loss (as decibels), and tonsil positions of all patients were recorded. No additional procedure was needed for the study except from the routine referral examination. This study conformed to Helsinki Declaration.

Audiometric and demographic information of patients were obtained from the hospital information system. Patients with a history of previous ear surgery or tympanic membrane perforation, chronic otitis media, conductive hearing loss, middle ear pathology, history of noise exposure such as working in a noisy environment, use of ototoxic medicines, or acute infection of upper respiratory system were not included into the study. Pure-tone audiometry had been carried

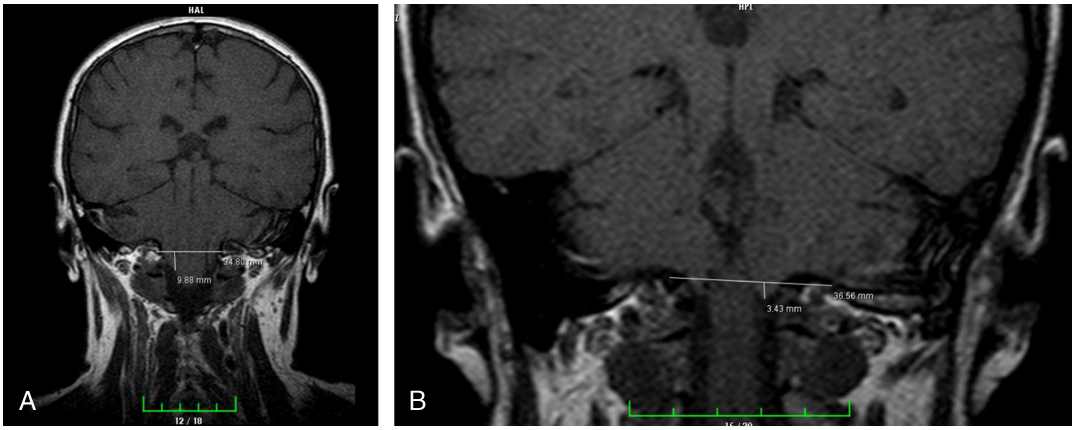

FIGURE 1. Measurement of tonsil descent on coronal T1-weighted MRIs with samples of CM 1 (A) and ectopia (B).

out in all patients. Pure-tone hearing threshold of both air conductions and bone conductions were obtained at 250, 500, 1000, 2000, 3000, 4000, and 6000 Hz using an audiometer (Interacoustics AD 229 audiometer; Interacoustics, Assens, Denmark). All conductive hearing losses were not included in the study group. Presence of hearing loss was accepted when hearing threshold was higher than 20 dB at all frequencies.

All MRIs were performed in a 1.5-Tesla MRI system (Philips Intera; Philips Medical Systems, Best, The Netherlands) in our department. From the routine MRI of patients with SHL, precontrast coronal spin echo T1-weighted images (3-mm thickness and 0.3-mm interslice gap) were used for the evaluation of cerebellar tonsil positions. Contrast-enhanced 3-dimesional gradient echo T1-weighted images (slice thickness, 1 mm with no interslice gap) were used for the evaluation of vertebral arteries and venous sinuses. On coronal images, the center of bilateral lower end of occipital condyles was established for the ectopia measurements (Fig. 1). A descent more than 2 mm was assumed as cerebellar ectopia, and a descent equal or more than 5 mm was assumed as CM 1. A tonsil descent group was also formed by summation of both groups for statistical analysis. Transverse diameters of bilateral intracranial vertebral arteries and transverse sinuses were also measured for any possible correlation (Fig. 2). In statistical evaluation, frequency, χ^2, and correlation analyses were established.

RESULTS

The prevalence of Chiari malformation, ectopia, and tonsil descent in SHL and controls were 7.2% (n = 12) and 0.5% (n = 1); 13.3% (n = 22) and 0.5% (n = 1); and 20.5% (n = 34) and 1% (n = 2), respectively. A significant difference of frequencies of all 3 parameters was detected between patients and controls ($P < 0.05$).

The mean (SD) length of tonsil descent was 4.49 (2.59) mm and 6.72 (7.44) mm for patient and control groups, respectively. Because of the limited number of cases (3) in controls, no statistical comparison could be done between groups for the descent.

The level of SHL as decibels did not show any significant difference between cerebellar ectopia and Chiari groups.

There was no significant correlation for the diameters of vessels with acoustic tests and MRI parameters except the left lateral sinus diameter. It showed a positive correlation with tonsil descent.

A powerful correlation was detected between SHL and age ($P < 0.01$). In addition, right and left vertebral artery diameters showed positive correlations with age ($P < 0.05$).

DISCUSSION

Caudal descent of the cerebellar tonsils more than 2 mm, whether as ectopia (between 2 and 5 mm) or as CM 1 (5 mm or

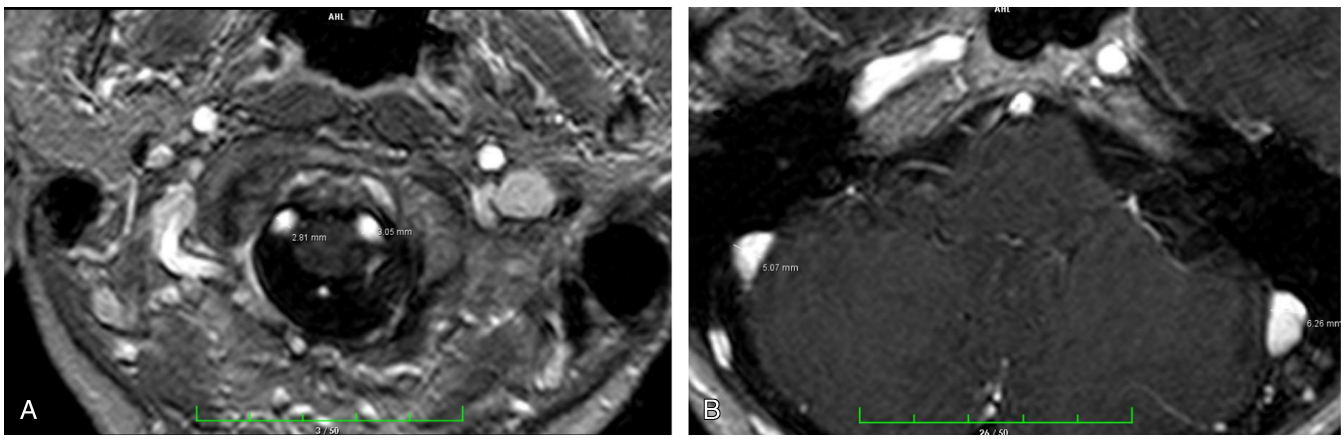

FIGURE 2. Measurement of transverse diameters of bilateral intracranial vertebral arteries (A) and transverse sinuses (B) on contrast-enhanced T1-weighted 3-dimensional MRIs.

higher), associates with SHL. The association of CM 1 with hearing loss has been showed in pediatric age group previously.[5] Aside from that study, there are very limited reported studies and case reports on this topic.[3,4,6–8] Our study seems to be the first to evaluate the association between cerebellar tonsil descent along with CM 1 with SHL.

The clinical presentation of CM 1 malformation is highly variable. Upper extremity weakness and different pain syndromes including neck and arm are the most frequently reported symptoms in the literature followed by occipital headaches, which are exacerbated by Valsalva maneuver or weight lifting. Besides, there are other symptoms that imply otologic impairment such as clumsiness, ataxia, vertigo, hearing loss, and tinnitus.[9] Among them, hearing loss is not a well-known presentation of Chiari patients. Rather, cervical pain and occipital headaches are common signs.[3] Neuro-otologic manifestations of this malformation were documented comprehensively by Sperling et al.[4] The most common aural symptoms reported in their study were tinnitus and aural fullness followed by vertigo and nausea. They reported fluctuating hearing loss in 56% of the patients. In that study, 81% of patients reported their aural symptoms to occur during episodes of neurologic symptoms such as headache. Besides, the authors did not investigate hearing loss whether it is sensorineural or not. The higher prevalence of CM 1, ectopia, and tonsil descent in patients with SHL than in controls in our study suggests that an inverse relationship may also be applicable. Thus, we can propose that Chiari patients should be evaluated for hearing loss and SHL patients with an unknown origin should be examined for a possible tonsil descent.

There have been several assumptions to explain the dysfunction of the vestibulocochlear nerve in Chiari patients including stretching of the nerve due to brainstem herniation, compression of the cochlear nuclei or the eighth cranial nerve by the cerebellar tonsils, ischemic damage to the cochlear and vestibular nuclei that may occur as a result of a microcirculation disorder or due to posterior inferior cerebellar artery compression, and cochlear damage from the transmission of elevated cerebrospinal fluid pressure through an anomalous cochlear aqueduct.[6,8,10] Our results do not contribute to elucidate these relationships. However, there was no significant difference between cerebellar ectopia and Chiari groups for the level of SHL as decibels in our study. This result may undermine the claim of stretching of the nerve or compression of the cochlear nuclei, the eighth cranial nerve by the cerebellar tonsils, or vascular compression-ischemia theories.

There was no significant correlation for the vessel diameters with hearing loss and MRI parameters except the left lateral sinus diameter, which showed a positive correlation with tonsil descent. This may be due to compression of the left lateral sinus by crowding of adjacent extraparenchymal space. We did not evaluate right and left tonsils separately. Such an evaluation could explain a possible cause.

Computed tomography, although having superiority in bony detail, is not preferred because of limited contrast resolution and irradiation risk.[11] Magnetic resonance imaging is a frequently used method for the evaluation of SHL. High-resolution MR sequences using 3-dimensional heavily T2-weighted fast imaging techniques have demonstrated an important role for the evaluation of the fine anatomical structures of the internal auditory canal and inner ear.[12] The superb capabilities of MRI powered with developing technology (3, 4, 7, or higher Tesla machines, advanced soft and hardware, etc) will contribute to further research.

A powerful correlation between SHL and age is an expected result that was found by us as well. In addition, we found a positive correlation of right and left vertebral artery diameters with age. This is an off-the-point result for our study. Nevertheless, increasing vertebral vein diameter with age appears remarkable and may give rise to further researches.

There are some limitations of our study. First, we did not perform any hearing test on the control group. In addition, documentation of the symptoms in the patient group that might be associated with CM 1 would be beneficial. Second, using coronal T1-weighted images with 3-mm thickness for patients and sagittal 5-mm images for controls might have caused inconsistency of measurements. A new control group assessed with identical method will be beneficial. Third, the control group is too small for the comparison; hence, the prevalence of cerebellar tonsil ectopia would be expected more than our results. Besides, evaluation of right and left tonsils separately might provide information why the left lateral sinus diameter showed a positive correlation with the descent.

In conclusion, CM 1 and cerebellar ectopia showed an association with SHL. Chiari 1 patients should be evaluated in collaboration with otologists. Inversely, routine ear MRIs for patients with SHL should include cerebellar tonsils and cranial base. The images must be evaluated for tonsil descent as well. Besides, further research to explain the exact cause of this currently unknown association seems required. This seems to be a topic of interest for high-resolution MRI studies.

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