Sudden hearing loss as a manifestation in a case of Arnold-Chiari malformation

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Arnold-Chiari malformation (ACM) is categorized into 4 types on the basis of the severity of hindbrain herniation and cerebellar dysplasia. Type I malformation, also known as adult ACM, there is caudal descent of the cerebellar tonsils only and can be a cause of unexplained headaches, dizziness, blurred vision, vomiting, vertigo, and drop attacks. In this unique case report, a 52-year-old man who suffered a fresh sudden hearing loss (SHL) of the left ear, accompanied with dizziness, tinnitus and difficulty to fall asleep. His history also included a first attack of SHL and tinnitus in the right side 15 months ago. A pure-tone hearing test showed sensorineural hearing loss on the bilateral sides (slight loss on left and moderate loss on right). On vestibular evaluation, a spontaneous down beating nystagmus was seen. Then a head MRI demonstrated the existing of ACM type I. Two months after the neurosurgical decompression, the patient’s nystagmus diminished thoroughly, whilst mild improvement of hearing. It is suggested that the finding of abnormal central vestibular nystagmus or abnormal vestibular-visual interaction is a useful sign to help determine whether a ACM is underlying some otolaryngological symptoms such as hearing loss, dizziness or tinnitus. MRI is the first choice to diagnose the ACM type I.

Key words: Arnold-Chiari malformation, sudden hearing loss, nystagmus, MRI

Introduction

Arnold-Chiari malformation (ACM) consists of a group of cranial-cervical junction abnormalities that involve the cerebellum, medulla, and stellate anatomy of the posterior cranial fossa. In 1883, John Cleland first described the congenital brain stem anomaly and proposed the mechanism of its formation. In 1892, Chiari described three different types of brain stem malformation. Several years late Arnold discussed the similar hindbrain findings as the Chiari malformation type II. Then, Schalbe and Gre-dig added Arnold’s name to Chiari I and II malformation in 1907. For convenient, today all of the Chiari malformations are often referred to Arnold-Chiari malformation.

ACM is categorized into 4 types on the basis of the severity of hindbrain herniation and cerebellar dysplasia (Table 1). Type I is characterized by displacement of cerebellar tonsils into foramen magnum and elongation of medulla. Type II involves displacement of cerebellum, low pons and medulla, and is
Table 1 Summary of the classification of Arnold-Chiari malformation

<table>
<thead>
<tr>
<th>Type</th>
<th>Imaging findings</th>
<th>Presentation</th>
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<tbody>
<tr>
<td>I</td>
<td>Caudal displacement of cerebellar tonsils through foramen magnum into the upper cervical spine.</td>
<td>Insidious onset. Often asymptomatic until adulthood. Common age of diagnosis in the forth to sixth decade.</td>
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<tr>
<td>II</td>
<td>Caudal displacement of inferior cerebellar vermis through foramen magnum. Thin elongated pons and medulla oblongata. Thin ogen slit like IVth ventricle extending into the spinal cord.</td>
<td>Often present at birth with meningocele, or meningomyelocele. Commonly associated with hydrocephalus.</td>
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<tr>
<td>III</td>
<td>Bony occipital defect with complete cerebellar descent into the cervical spine.</td>
<td>Presents at birth with occipital meningoencephalocele.</td>
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<tr>
<td>IV</td>
<td>Cerebellar hypoplasia.</td>
<td>This is not a syndrome involving herniation.</td>
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usually diagnosed in childhood. In type III, cerebellar and brain stem are displaced into a high cervical meningocele. Meanwhile, type IV is characterized by cerebellar hypoplasia and also considered by some authors as an unrelated malformation to ACM.5

Type I malformation, also known as adult ACM, typically occurs in person during early adulthood and can be a cause of unexplained headaches, dizziness, blurred vision, vomiting, vertigo, and drop attacks. The intorduce of magnetic resonance imaging (MRI) scanning makes possible the accurate and noninvasive diagnosis of apparently symptomatic ACM type I. On the other hand, the diagnosis of type I malformation in adults is often delayed because of the slowly improved or underlain symptoms. In this paper, we report a patient with ACM type I where hearing loss was the most prominent presenting symptom.

Case Report

This 52-year-old man was referred to Miyoshi ENT clinic because of a fresh sudden hearing loss (SHL) of the left ear, accompanied by dizziness, tinnitus and difficulty to fall asleep. His history also included a first attack of SHL and tinnitus in the right side, stuffy feeling in the left ear (without hearing loss), and balance disorder 15 months ago. He was diagnosed as right SHL and bilateral SHL respectively at another clinic. Just before he came to our clinic, a head computer tomograph (CT) was carried and no abnormality was found according that.

The patient was alert and oriented during our physical examination. Corneal reflexes, facial sensation, and facial movements were normal. The palate elevated in the midline and the gag reflex was present bilaterally. Movement in the tongue and motorsystem revealed normal strength. The tympanic membranes were also normal.

In a standard soundproof booth, the pure-tone hearing threshold (moderate sensorineural hearing loss) of the right ear was almost the same as that of 15 months ago. But a new slight sensorineural hearing loss on the left side was detected in the audiogram (Fig. 1). On vestibular examination, a spontaneous nystagmus was seen under Frenzel grass. Whenever up-gaze, down-gaze or lateral-gaze, the patient always had a down beating nystagmus of both eyes on sitting position (Fig. 2).

By the light of nystagmus test, both an angiography and an MRI scanning were performed although the negative demonstration of the head CT scanning. The angiography indicated downward displacement of posteriorinferior cerebellar arteries to the upper cervical spinal canal (Fig. 3). Eventually, MRI demonstrated a ACM type I with
Figure 1  Pure-tone audiogram revealed a fresh sudden hearing loss on left side whilst the threshold of the opposite keeping no change comparing with 15 months ago.

Figure 2 The finding of spontaneous nystagmus under Frenzel glass and positional nystagmus in position changing test.
   a) Spontaneous nystagmus under Frenzel glass on sitting position
   b) Positional nystagmus on head hanging position to dorsal decubitus

abnormal cerebellar tonsils descending to the upper cervical spinal canal, which compressed the cord and lower brain stem ventrally (Fig. 4). The patient was referred further
to the Department of Neurosurgery, National Sendai Hospital and underwent posterior fossa decompression, which included of craniotomy of the posterior fossa, removal of the arch of C-1, opening of the dura and the arachnoid. One month after the operation, the nystagmus stabilized. Two months later, the nystagmus diminished thoroughly with a slight improvement of hearing on the left side (Fig. 5).
Discussion

ACM consists of a group of congenital malformations, involving brain stem, cerebellum, upper spinal cord and surrounding bony structure. Type I malformation, the mildest of the four types of ACM, consists of an elongation of the cerebellar tonsils and medial parts of inferior lobed of the cerebellum as a tongue like process enveloping the medulla and displaced into the upper cervical vertebral canal.

Common clinical manifestations of ACM type I include nystagmus, headache, dizziness, blurred vision, vomiting, and drop attacks. Various types of nystagmus have been described in this disease but a downbeat type of nystagmus has been traditionally considered to associate with ACM, although it may also be produced in many other brain disorders. In our case, just by the prompting of downbeat nystagmus, we recommended a MRI scanning to the patient and finally discovered this type I malformation. Gaze-evoked nystagmus and horizontal spontaneous nystagmus are also commonly seen in ACM type I. Other rare patterns, such as upbeat nystagmus, skew nystagmus, rebound nystagmus, periodic alternating nystagmus and so on, have been sparsely reported.

ACM type I typically presents with varying combinations due to either a foramen magnum compression syndrome or a central cord syndrome. However, several asymptomatic cases, such as initially manifested as vocal cord paralysis, airway abnormality, posturally evoked vomiting, were reported respectively in recent years. Rarely, as in the case described here, hearing loss also can be the presenting manifestation. The first known cases, which hearing loss not as the original complaint, have been reported by Malis et al in 1951. Eight from forty cases had deafness as a component of their symptoms. After two decades, Rydell and Pulec reviewed the charts of 130 patients with previously diagnosed ACM and noted 29 patients with symptom of hearing loss. No character about those hearing impairment can be acquired. And they did not note any patients with complaint of sudden deafness as their initial symptom. In this unique report case, under the first attack only the right ear showed a moderate sensorineural hearing loss in all frequencies. While at the second attack, the threshold of right ear nearly kept no change but he opposite showed a slight hearing loss, especially among the low-frequency.

The pathophysiological causes underlying these abnormal vestibular finding are unknown, but are probably based on abnormalities of the elongated brain stem in which some cranial nerve roots exit. The most common cranial nerve involvement is with the Vth nerve in the forth of neuralgia,
followed by the 11th to XII th cranial nerves with symptoms including dysphagia, vocal fold palsy,4) and central apnea. Because the signs of brain stem dysfunction often evolve slowly over many years, the diagnosis of ACM in adults is often delayed.

Not only the symptomatic ACM type I, there also have lots of asymptomatic cases been reported. In 1984, Miyoshi18) (one author of this paper) first described a 55-year-old man who at beginning was suspected as olivopontocerebellar atrophy because of central balance disorder and an anxiety disorder, and then ACM type I was eventually diagnosed. Just one year late, Iwabuchi et al19) described another case of anxiety disorder in association with ACM. In 1993, Chisholm et al20) reported a patient who suffered concurrently from panic disorder with agoraphobia and ACM. Surgical correction of the neuroanatomical anomaly altered that patient's symptom pattern, enabling a more clear delineation of his anxiety disorder. The writer considered that the symptoms of ACM have acted as a trigger for episodes of anxiety.

Conversely, some situation such as positional changes,17) hip replacement surgery,10) pregnancy,21) may also make a role to the attack of type I ACM. Elster and Chen22) noted approximately 30% of patients with cerebellar tonsils herniating 5 to 10 mm below the foramen magnum were free of symptoms. Thus, with more widespread introduction of MRI to screen a broad segment of the population, it is likely a higher prevalence of asymptomatic ACM will be discovered. And otolaryngologists will be increasingly called on to evaluate patients with these incidental ACM.

Today, the diagnosis of ACM is most often made by MRI. Conventional CT scans can show the bony abnormalities of the skull and cervical vertebrae, but they do not demonstrate the brain tissue anomalies well. Just as in our case, CT scanning is often read as normal in patients with type I malformation because of the lack of hydrocephalus, the normal fourth ventricle, and the poorly visualized region of the foramen magnum.23) Vertebro angiography is often helpful for it can disclose the displacement of the posterior inferior cerebellar arteries.24) Compared with CT scanning, MRI can clearly show the displacement of the cerebellar tonsils into the upper cervical spine. It is also sensitive to detect the cerebellar tonsils below the level of the foramen magnum. By MRI, a diagnosis of ACM type I can be obtained easily. Furthermore, MRI can be performed periodically to follow the disease progression.

The treatment for ACM type I is posterior fossa decompression. This procedure consists of craniotomy of the posterior fossa, removal of the arch of C–1 and C–2 if needed, opening of the dura and the arachnoid, and resection of any adhesions. Patients with symptoms for longer than 2 years fared much more poorly than those with symptoms for less than 2 years.25) Early surgical decompression is recommended on symptomatic patients to obtain a better outcome. Surgical procedures appear to be successful at stabilizing and sometimes improving symptoms of brain stem dysfunction. As to other forms of vestibulopathy that may be occurring concurrently in patients with asymptomatic ACM type I, a conservative therapy, such as vestibular rehabilitation can be recommended and it is also considered to be effective.8)

In conclusion, it is important for otolaryngologists to recognize ACM type I as part of the differential diagnosis of SHL. ACM should be considered in patients in whom clinical evaluation or eye movement re-
cordings demonstrate central vestibular nystagmus. Therefore a head MRI should be recommended strongly.

References

突発性難聴を発症し、判明したアーノルド・キアリ奇形の1例
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アーノルド・キアリ奇形は小脳扁桃が大後頭孔に嵌入するという診断基準をもとに4つのタイプに分類される。うちタイプ1奇形は、成人型アーノルド・キアリとも呼ばれているが、小脳扁桃が尾側に下降し、説明不可能な頭痛、立ちくらみ、視野の欠損、嘔吐、めまい、失神などを引き起こす。今回報告する症例は、めまい、耳鳴り、不眠をともなう左側実発性難聴の52歳の男性で、15カ月前に右側実発性難聴と耳鳴りの最初の発作があった。標準純音聴力検査では両側に感音性難聴が認められた（左は軽度、右は中等度難聴）。眼振検査では、下眼瞼向きの眼振がみられた。頭部MRIよってアーノルド・キアリ奇形タイプ1の存在が明らかとなった。脳外科に手術が行なわれ、その2カ月後、聴力は、徐々に回復し眼振も完全に消失した。難聴、めまい、耳鳴りなどの耳鼻科的症狀のもとに隠れているアーノルド・キアリ奇形の診断をするに当たり、垂直性眼振の発見が重要であり、またMRIはアーノルド・キアリ奇形タイプ1を診断するにあたって、必要不可欠といえる。