Otorhinolaryngologic Manifestations in Chiari Malformation

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The Chiari malformation causes herniation of the cerebellar amygdalae through the foramen magnum, resulting in the descent of the brain stem and/or traction on the lower cranial pairs. It is important for otolaryngologists to recognize Chiari malformations as part of the differential diagnosis of balance disorders, because patients may initially exhibit symptoms related to the vestibular system, including ataxia, nystagmus, or vertigo. We report 2 cases. (Am J Otolaryngol 2002;23:99-104. Copyright 2002, Elsevier Science (USA). All rights reserved.)

Chiari malformation is characterized by a herniation of the cerebellar amygdalae through the foramen magnun with a descent of the brain stem and traction of the lower cranial pairs.[1] It is classified into 4 types according to the degree of prolapse[2-4]:

- *Type I* is characterized by herniation of the amygdalae without cerebellar displacement. This is the most frequent type and is diagnosed in mature adults who are symptomatic for years.
- *Type II*, or *Arnold Chiari Syndrome*, consists of a protrusion of the cerebellar vermis, the pons, and the bulb. There may even be an introduction of the fourth ventricle in the cervical spinal column. It is diagnosed in the first months of life as it is associated with myelomeningocele in 90% of cases.[1]
- *Type III* is associated with osteal defects of the occipital bone with complete cerebellar herniation. This lesion is fatal.[5]
- *Type IV* is characterized by hypoplasia of the cerebellum and is also fatal.

Type I Chiari malformation may be asymptomatic for years and later show nonspecific vestibular disturbances, such as instability or nystagmus of insidious appearance and evolutionary character, along with others of a more general character such as migraines. We report 2 clinical cases of patients with Chiari 1 malformation who presented with otorhinolaryngologic symptoms.

CLINICAL CASES

Case 1

A 46-year-old woman with no previous medical history of interest presented twice at the Otorhinolaryngology Service at the Miguel Servet University Hospital, Zaragoza, Spain complaining of a sensation characterized by gyrating objects. The sensation lasted for several hours and was not accompanied by vegetative symptoms or related to the position of the head or any other factor. Later, the patient experienced postcrisis instability that lasted for a few days and was not associated with neurologic or auditory symptoms. She also claimed that she had a history of ronchopathy with morning asthenia and daytime somnolence, although sleep apnea was doubtful.

The otoscopy was normal, showing a horizontal spontaneous nystagmus toward the left and a lower vertical one. In the exploration of equilibrium, the Romberg and Unterberger test results were negative and those of walking, of indices, and of indication were normal. The neurologic examination did not indicate a disease of cranial pairs or cerebellar symptoms. In the complementary tests performed,

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normality was obtained in the tonal audiometry but not in the electronystagmogram (Fig 1). In the saccadic study, a dysmetria due to hypermetria was observed in the gaze to the right and a certain slowness in the vertical saccades. Both findings suggested central alteration. Pendulous ataxic following in both directions (horizontal and vertical) with laddered and sawtooth images suggested a central, probably cerebellar, lesion.

In the exploration of the nystagmus gaze to the right, an ambiopia was found, which was referred to by the patient during the examination, together with a downbeat nystagmus. An evident horizontal spontaneous nystagmus was found to the left. In the study of the positional nystagmus, a fixed direction nystagmus was observed to the left in the supine decubitus position, turned to the right and the left. In the caloric tests, no labyrinth paresia was observed, but there was a moderate left directional prevalence (36.7%).

The brain stem auditory evoked potentials were normal as was the cerebellopontine angle computed tomography (CT). Magnetic resonance imaging (MRI) was performed, and a herniation of the cerebellar amygdalae at the level of the foramen magnum was observed (Fig 2). This finding resulted in the definitive diagnosis of a Chiari I malformation.

To complete the study, we performed a static and dynamic exploration of the air tract in accordance with the protocol of our otorhinolaryngology Service, the Epworth test of daytime somnolence with a value of 13, and polysomnography. The patient was diagnosed with low-degree obstructive sleep apnea, for which hygienic, dietary, and postural treatments were established. Symptomatic pharmacologic treatment was prescribed with betahistine for 2 months and with dihydroergocristine for another 2 months. In 6 months, the instability disappeared and today, 2 months later, a light nystagmus persists in the lateral gaze that can only be seen with a Frenzel lens.

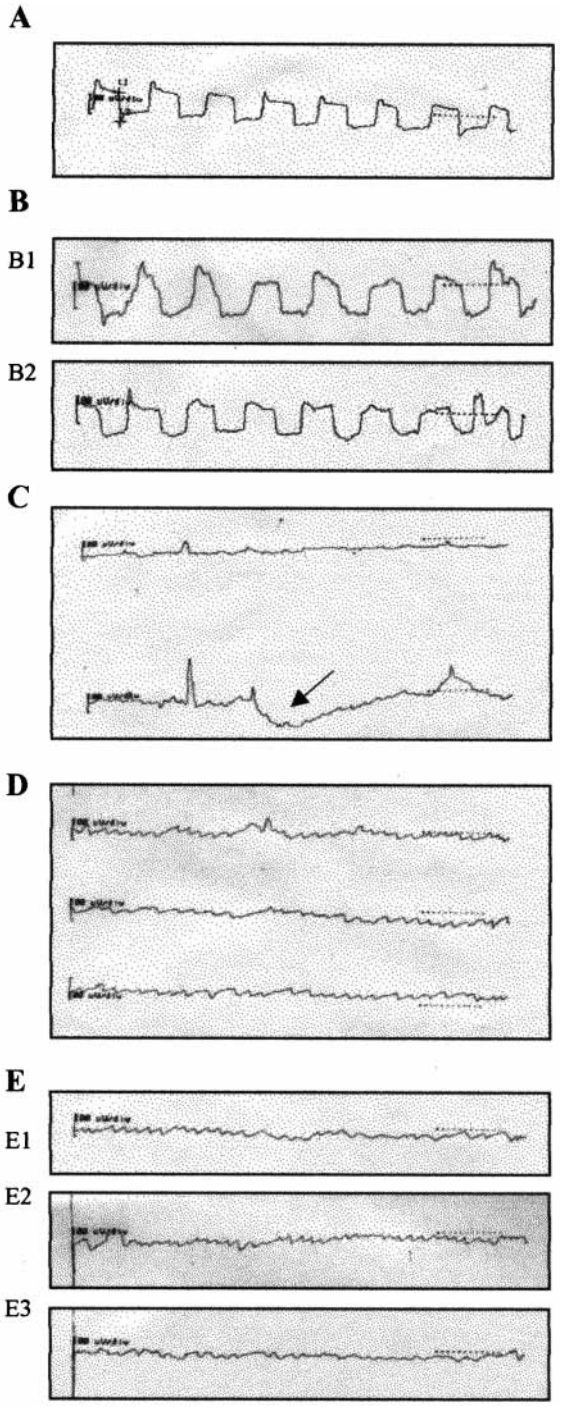

Fig 1. Case 1: Electronystagmographic study. Horizontal saccades (A), horizontal pendular movement (B1) and vertical pendular movement (B2), extreme gaze to the right with downbeat nystagmus (arrow) (C), spontaneous nystagmus (D), and positional nystagmus in supine decubitus (E1), right turn (E2), and left turn (E3).

Case 2

A 48-year-old man with no previous medical history of interest presented with a rocking sensation that had lasted several hours accompanied by vegetative symptoms with clear

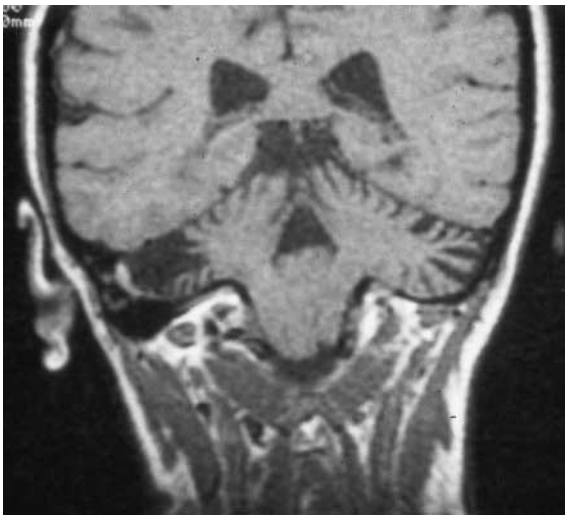

Fig 2. Case 1: MRT in coronal section. A herniation of the cerebellar amygdalae is observed through the foramen magnum.

right lateropulsion. These symptoms had increased in severity in the previous months. The patient also complained of constant tinnitus in both ears and a 2-year progressive bilateral hypoacusis that increased in the periods of crisis. He also manifested ronchopathy for several years with daytime somnolence and possibly sleep apnea. No other neurologic symptoms were present, although he related that his brother also suffered from vertigo.

The otoscopic examination was normal as was the ocular-vestibulum reflex and the neurologic examination. However, in the study of equilibrium, the Romberg, Unterberger, and walking tests showed a remarkable deviation to the right. The test results of indices and indication were normal. In the audiometry, a bilateral neurosensorial hypoacusis was observed that was more acute in the left ear. The most relevant findings in the electronystagmographic study were a pendular movement of saccadic characteristics and a spontaneous horizontal nystagmus to the right. In the evaluation of the positional nystagmus, a nystagmus was observed to the left when the patient hyperextended his head and to the right when he turned to right and left. The electronystagmographic explorations suggested a central type lesion (Fig 3). All of the studies performed in the previous clinical case were carried out in this patient until diagnosis of light obstructive sleep apnea was reached. The brain stem auditory evoked potentials and the CT were normal. In the MRI, a herniation of the cerebellar amygdalae was observed toward the brain stem; thus, the patient was diagnosed with Chiari I malformation (Fig 4).

At present, the patient experiences migraines that are triggered by and increase with certain postures and efforts, the oscillation of objects in his visual field of nonophthalmologic cause, and episodes of falls with loss of consciousness. He has been evaluated by the Neurology Department and remitted to the Neurosurgery Service for surgical treatment of the Chiari I malformation. His vertigo remains the same, continuous instability with more intense episodes, despite symptomatic medical treatment with betahistine, nimodipine,

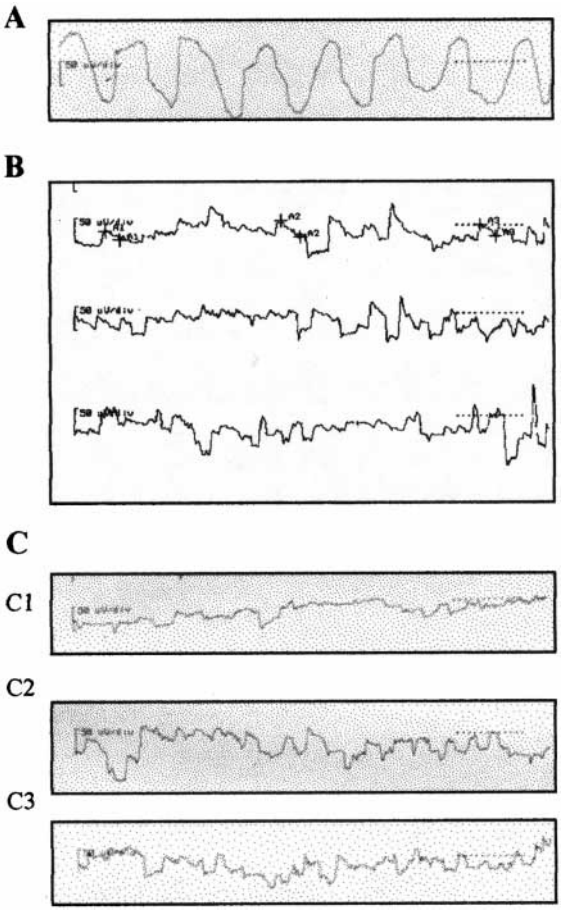

Fig 3. Case 2: Electronystagmographic study. Pendular movement of saccadic characteristics (A), spontaneous nystagmus to the right (B), and positional nystagmus in hyperextension (C1), right turn (C2), and left turn (C3).

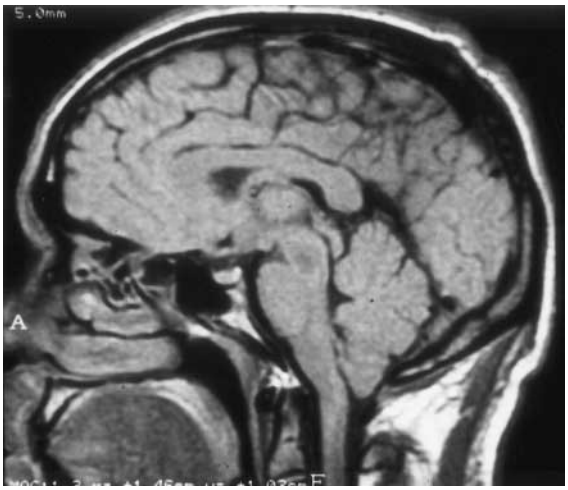

Fig 4. Case 2: MRI in sagittal section. Cerebellar herniation is observed.

and rehabilitation, and the hypoacusis is clearly more severe.

DISCUSSION

There are many etiopathogenic theories postulated to explain Chiari malformation, which is considered by some investigators to be a primary malformation of the brain stem, secondary to the pressure from above caused by hydrocephalia or downward traction of the trunk, and secondary to the caudal fixation of the marrow in the myelomeningocele. However, the most accepted theory is that a primary hypoplasia of the chondrocraneum of the posterior fossa would force the cerebellum to grow towards the brain stem. Extensive studies show the same distribution for both sexes.

Chiari malformation is associated with other malformations, hydrocephalus and the almost constant secondary myelopathy, and syringomyelia, which is the most frequently associated independent malformation.[1,6] This malformation is usually cervical but may also appear in the dorsal or lumbar column or syringobulbia. Arnold Chiari's syndrome (type II) is related to myelomeningocele.

Chiari malformations are commonly found in the context of multiple bone malformations of the cranio-occipital region, the most frequent of which include the basilar groove or odontoid process, platybasia, the partial occipitalization of the atlas, hypoplasia of the atlas, bifidism of the posterior arch of the atlas, occipital fusion with some of the lateral masses, odontoid deviation, occipital vertebralization, persistence of the proatlas, vertebral cervical fusions (Klippel-Feil syndrome or dystrophia brevicollis), and spina bifida or more extended malformations in the rest of the column.

The basilar groove is defined for the ascent or sinking towards the skull of the occipital orifice and neighboring structures, especially the odontoid apophysis of the axis. Usually this apophysis does not extend more than 5 or 6 mm beyond the lines of Chamberlain (it joins the posterior edge of the bone palate with the posterior edge of the occipital orifice) or the lines of McGregor (it joins the posterior border of the bone palate with the lowest portion in the occipital one) in a lateral cranium radiograph. Its penetration above this is indicative of a basilar groove. Practically all of the basilar grooves are platybasic (the clivus is usually shortened and adopts a horizontal direction), but isolated platybasias may exist.

In the Chiari malformation, the clinical presentation is conditioned by 3 factors: firstly, by the degree of herniation of the nerve structures of the posterior fossa; secondly, by the association with syringomyelia; and finally, by the medullary segments affected by the myelopathy. Although it represents a congenital malformation, the classic neurologic affliction is not congenital and may appear with evolutionary characteristics at any age, but typically during youth. Some studies have reported approximately 30% of asymptomatic cases in Chiari I malformation.[7]

The most frequently reported clinical symptom of Chiari malformation is occipital migraine (90%) (Table I). Cerebellar symptoms may also appear, such as ataxia, vertigo, instability, dysmetria, alterations in the diadochokinesia, hypotonia, intentional trembling, or dysarthria, which would lead to a differential diagnosis of a tumor of the posterior fossa. Ocular symptoms are infrequent; ambiopia is the most frequently observed. Bronchoaspirations, syncope, respiratory alterations, and multiple symptoms may appear. The coexistence of unrelated afflictions often leads to a differential diagnosis of this disease with multiple sclerosis.

TABLE 1. Symptoms of Chiari Malformation

General	Cerebellar	Secondary to Syringomyelia	Ocular Alterations
Headache	Hypotonia	Tingling in the extremities	Loss of vision
Fatigue	Trembling	Hypoesthesia in the extremities	Intolerance of bright light
Memory loss	Dysarthria	Burning sensation in the extremities	Diplopia
Pressure on the neck	Ataxia	Thermalgesic anesthesia	
Back pain	Dysmetria	Alteration in muscular reflexes	
Insomnia		Areflexia	
Poor circulation		Alteration of kinesthesia	
Nausea		Motor skill dysfunction	
Menstrual problems			
Sexual alterations			
Hypothermia			
Bronchoaspirations			
Respiratory alterations			
Drop attacks			

Syringomyelia has a thermalgesic anesthetic characteristic of segmental distribution with analgesia of lower cervical segments. Hence, these patients frequently present with burns and traumatisms in the upper extremities. Syringomelia also occurs with amyotrophia and alteration of osteotendinous reflexes due to areflexia, as well as with vesical incontinence.

After several retrospective studies of patients with Chiari malformation, it has been shown that instability is the most frequent symptom in the otorhinolaryngologic field (77%).[8] Vertigo, sickness, nystagmus, and hypoacusis may also occur (Table 2). In some studies, unilateral progressive hypoacusis has been observed.[3] Alterations of the last cranial nerves are characteristic [9] and produce dysphagia, dysphonia, laryngeal stridor, or palatal paralysis.[5] Graham documents a case of bilateral vocal chord paralysis.[10] Facial hypoesthesia may occur because of compression or trigeminal dysesthesia.[11] The cause of the lesion of cranial pairs is not known, but it is thought that it might be due to their elongation.

Sleep apnea is frequent in patients with Chiari malformation.[12] They may have a central component because of the compression of the respiratory center caused by hydrocephalia or traction of the brain stem, as well as an obstructive component, because, in Chiari malformation there are frequently associated malformations of the osteocartilaginous craniofacial skeleton. The latter was the case in our 2 patients in whom the light obstructive sleep apnea was diagnosed. A differential diagnosis will be carried out with other central nervous system diseases, including acoustic neurinomas, multiple sclerosis, and processes of the posterior fossa.

Electronystagmography or videonystagmography will aid in diagnosis, but typical findings of this syndrome being made are not exclusive to this malformation, nor are alterations in pendular movement in the saccades or the downbeat nystagmus, which may also appear in olivopontine cerebellar atrophy,

TABLE 2. Otorhinolaryngologic Manifestations Associated With Chiari Malformation

Vestibular Manifestations	Alterations of Cranial Pairs	Other Symptoms
Imbalance	Dysphagia	Sleep apnea
Swaying	Dysphonia	
Dizziness	Alterations in tongue mobility	
Positional vertigo	Loss of smell	
Spontaneous vertigo	Facial hypoesthesia	
Nystagmus		
Hearing loss		
Tinnitus		

neoplasms, brain stem infarcts, or multiple sclerosis.[13,14] The brain stem auditory evoked potentials are nonspecific. A specific CT of the basilar lamina can be performed, but an MRI will yield a more certain diagnosis. The myelographic study shows an image characteristic of contrast blockage in the cervical region caused by herniated cerebellar tissue.[1]

Surgical treatment consists of a suboccipital craniectomy in adults with Chiari I malformation with debilitating symptoms and in Chiari II patients as an early treatment along with treatment of the myelomeningocele.[15] Surgery will always be accompanied by a marsupialization or drainage of the cavity in cases involving associated syringomyelia. Satisfactory results have been obtained with this treatment, including the resolution of some symptoms but the outcome in patients with syringomyelia is less successful.

We conclude that nonspecific and frequent clinical manifestations such as instability, nystagmus, or tinnitus, in otorhinolaryngology patients may obscure unusual origins like Chiari malformation.

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