

## Clinical Records

# Otoneurological manifestations in Chiari-I malformation

F. W. J. ALBERS, K. J. A. O. INGELS, (Ghent, Belgium)

### Abstract

The type I Chiari malformation consists of a caudal displacement of the cerebellar tonsils through the foramen magnum into the cervical spinal canal. The most common presenting symptoms, such as pain, weakness and headache, are frequently preceded by otoneurological symptoms. Sensorineural hearing loss, vertigo, nystagmus, dysequilibrium, tinnitus and other cranial nerve involvement have been reported in Chiari-I malformation. A case report is presented and the clinical features of the disease are discussed with emphasis on the otoneurological aspects.

**Key words:** Chiari-I malformation; Otoneurology.

### Introduction

The Chiari malformation comprises a group of deformities of the posterior fossa in which the cerebellar tonsils and the brainstem are displaced to a varying degree through the foramen magnum. Cleland was the first to describe a case of Chiari malformation in 1883. In 1891 and 1896 Chiari reported four cases of cranio-medullary malformations associated with hydrocephalus. Arnold, in 1894, described a case of spina bifida with caudal displacement of the cerebellum and the brainstem. According to the original classification, the Chiari malformation is divided into four categories:

- Type I: The cerebellar tonsils protrude into the cervical spinal canal without displacement of the brainstem.
- Type II: Extension of the cerebellar vermis into the upper cervical canal with caudal displacement of the lower pons and medulla. (Arnold-Chiari malformation).
- Type III: Herniation of the cerebellum through a bony occipital defect into a high cervical meningocele.
- Type IV: Hypoplasia of the cerebellum.

The type I Chiari malformation is usually asymptomatic until the onset of symptoms in the third to fifth decades (Sclafani *et al.*, 1991). In a review of 71 cases of Chiari-I malformation, Paul *et al.* (1983) described pain, weakness, numbness and unsteadiness as the most prominent symptoms. In reviewing the case histories of 130 patients with the Chiari malformation, Rydell and Pulec (1971) found audiovestibular symptoms in 22 per cent of cases. Otoneurological findings as sensorineural hearing loss, vertigo, nystagmus, dysequilibrium, tinnitus, facial hypesthesia, dysphagia and vocal cord paralysis have been reported in an early phase of the disease and may precede the development of more severe neurological complaints (Sclafani *et al.*, 1991). The importance of a careful otoneurological examination in the diagnosis of Chiari malformation is strongly emphasized by several authors (Rydell and Pulec, 1971; Chait and Barber, 1979; Longridge and Mallinson, 1985; Hendrix *et al.*, 1992).

In this paper the case report of a patient with Chiari-I malformation is described and the clinical features of the disease are discussed from an otoneurological point of view.

### Case report

A 58-year-old man was referred to the hospital complaining of intermittent episodes of vertigo associated with nausea and vomiting for the preceding two years. The patient also described a progressive hearing loss and aural fullness on the left side and periods of occipital headache. The past medical history revealed no abnormalities.

The otoneurological examination was completely normal. Pure-tone audiometry showed a flat sensorineural hearing loss in the left ear and a high frequency sensorineural hearing loss in the right ear (Fig. 1). Further audiological investigation including auditory brainstem response testing did not suggest retro-cochlear pathology. On electronystagmographic examination no spontaneous or positional nystagmus was observed. Caloric testing showed a compensated hypofunction of the left vestibular labyrinth. Routine laboratory studies were normal.

Magnetic resonance imaging (MRI) demonstrated a moderate

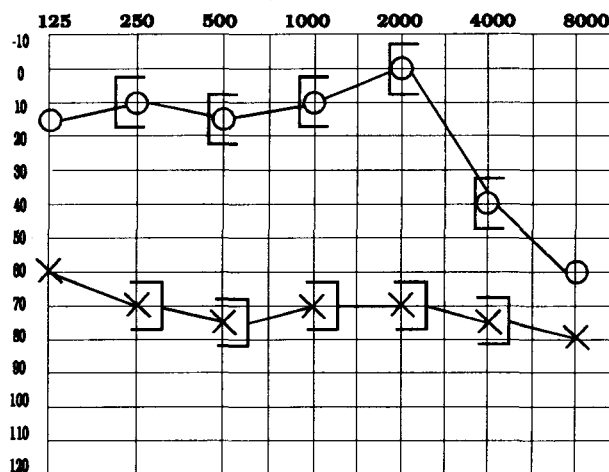


FIG. 1

Pre-operative pure-tone audiometry. O: air conduction right ear; [ : bone conduction right ear. X: air conduction left ear; ] : bone conduction left ear.



FIG. 2

Magnetic resonance imaging (sagittal plane) demonstrating a moderate protrusion of the cerebellar tonsils (arrow) through the foramen magnum.

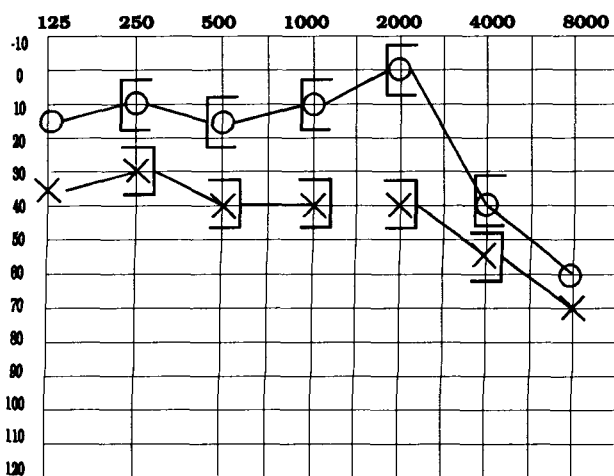


FIG. 3

Post-operative pure-tone audiometry. O: air conduction right ear, I: bone conduction right ear. X: air conduction left ear, J: bone conduction left ear.

herniation of the cerebellar tonsils through the foramen magnum consistent with Chiari-I malformation (Fig. 2). No further abnormalities were observed.

Neurosurgical decompression of the posterior fossa and cervical laminectomy with duraplasty was performed. The cerebellar tonsils were found to protrude in the cervical canal, confirming the diagnosis of Chiari-I malformation. The post-operative recovery was without complications.

In a follow-up period of two years the periodic episodes of vertigo remained unchanged. The patient reported a considerable reduction of the occipital headache and the hearing loss of the left ear. Audiometry after one year showed a moderate improvement of the sensorineural hearing loss on the left side, whereas the right side remained unchanged (Fig. 3). Repeated electronystagmographic examination revealed no further deterioration of the left vestibular function. Follow-up magnetic resonance imaging did not show any abnormalities.

## Discussion

Otoneurological symptoms are frequently observed in type I

Chiari malformation (Rydell and Pulec, 1971; Chait and Barber, 1979; Longridge and Mallinson, 1985; Sclafani *et al.*, 1991; Hendrix *et al.*, 1992).

Rydell and Pulec (1971) reported 13 of 130 patients with Chiari malformation complaining of auditory symptoms. Audiological evaluation revealed usually a bilateral, progressive sensorineural hearing loss. Hendrix *et al.* (1992) described three patients who had as a presenting symptom an asymmetric sensorineural hearing loss with audiological evidence of retrocochlear pathology. In a series of 71 cases with Chiari-I malformation, 10 per cent of the patients complained of tinnitus (Paul *et al.*, 1983).

Unsteadiness and loss of balance have been reported to occur in approximately 40 per cent of patients (Levy *et al.*, 1983; Paul *et al.*, 1983). In large retrospective studies nystagmus is described in 28 to 47 per cent of the patients (Rydell and Pulec, 1971; Levy *et al.*, 1983; Paul *et al.*, 1983). Downward beating vertical nystagmus enhanced by horizontal lateral gaze is reported to be specific for lesions involving the cervicomedullary junction (Cogan, 1968). Bertrand *et al.* (1973) and Chait and Barber (1979) confirmed the presence of downward beating nystagmus on lateral gaze in patients with Chiari malformation. Longridge and Mallinson (1985) presented a case of Chiari malformation with downbeating nystagmus on down gaze and other eye movement abnormalities, suggesting central pathology. In the series of Rydell and Pulec (1971), one patient had a variably-directed nystagmus that changed directions during the recording. On caloric testing usually no abnormalities are observed, although hypoactive responses are reported infrequently (Rydell and Pulec, 1971; Bertrand *et al.*, 1973; Chait and Barber, 1979; Longridge and Mallinson, 1985).

Other cranial nerves are also frequently involved in Chiari-I malformation. The trigeminal nerve is the most commonly affected cranial nerve (Rydell and Pulec, 1971; Sclafani *et al.*, 1991). Paul *et al.* (1983) reported hypesthesia of the Vth cranial nerve in 41 per cent of the cases. Dysfunction of the IXth to XIIth cranial nerves is occasionally associated with Chiari-I malformation. Levy *et al.* (1983) described involvement of the IXth to XIIth cranial nerves in 18 per cent of the patients. Graham (1963) and Ruff *et al.* (1987) reported bilateral vocal cord paralysis in a patient with Chiari-I malformation.

Standard computed tomography was reported to be non-specific in the diagnosis of Chiari-I malformation (Longridge and Mallinson, 1985). Computed tomography with subarachnoid metrizamide contrast can demonstrate Chiari-I malformation in 80 per cent of surgically-proven cases (Paul *et al.*, 1983). Magnetic resonance imaging has great diagnostic value in identifying Chiari-I malformation. The sagittal view on MRI provides an excellent demonstration of the craniocervical junction including the herniation of the cerebellar tonsils (Longridge and Mallinson, 1985; Sclafani *et al.*, 1991).

The most frequently performed treatment of Chiari-I malformation consists of decompressive craniectomy and cervical laminectomy with duraplasty. In a comprehensive review of 18 surgical series including 648 patients with proven Chiari malformation, Levy *et al.* (1983) found symptoms improved in 46 per cent; 32 per cent were unchanged and 20 per cent of the patients were worse post-operatively. Paul *et al.* (1983) reported early post-operative improvement of both symptoms (82 per cent) and signs (70 per cent), followed by late deterioration in 21 per cent of the cases. Rydell and Pulec (1971) observed a partial improvement of audiovestibular symptoms after surgical treatment in two of the four patients. In a case report of Chait and Barber (1979) decompressive craniectomy resulted in a complete relief of the vestibular symptoms and considerable improvement of ataxia and occipital headache.

The diagnosis of Chiari-I malformation must be considered in patients with unilateral or bilateral sensorineural hearing loss, tinnitus, vertigo, dysequilibrium and other cranial nerve involvement. To evaluate the audiovestibular functions in Chiari-I malformation after surgical treatment more detailed pre- and post-operative documentation is needed.

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## Address for correspondence:

F. W. J. Albers, M.D. Ph.D.,  
 Department of Otorhinolaryngology,  
 University Hospital Ghent,  
 De Pintelaan 185  
 B-9000 Ghent,  
 Belgium.