Incidentally depicted herniation of posteroinferior cerebellar lobe

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Abstract: Background and objective- the mild herniation of the posteroinferior cerebellar lobe used to be considered as a normal variant or irrelevant. The current study aims to determine the symptoms associated with incidentally discovered mild herniation of the posteroinferior cerebellar lobe through the foramen magnum. **Methods-** thirty incidentally depicted cases of mild herniation of the posteroinferior cerebellar lobe through foramen magnum (less than 5 mm) with no associated cervical cord syringomyelia or cerebral disorders associated with Chiari malformations, seen between December 2007 & March 2009. Those cases were subjected to complete neurological examination to depict the symptoms related to the herniation of the posteroinferior cerebellar lobe. The diagnosis of the herniation of the posteroinferior cerebellar lobe is based on mid-sagittal T1 or T2 FSE MRI. **Results-** the symptomatic cases mainly presented with chronic intractable occipital headache, vertigo, vomiting & disequilibrium. All patients included in the study showed normal brain except for herniation of the posteroinferior cerebellar lobewhich has been historically thought of no clinical relevance. 12 symptomatic cases subjected to further neurological examinations to elicit the etiological relationship between the posteroinferior cerebellar lobe herniation and such symptoms.

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Introduction

Chiari Type I malformation is a well-known disease involving caudal descent of the cerebellar tonsils and is occasionally associated with syringomyelia. 1–9 In 1891 and 1895, Chiari1 2 precisely described this malformation and related ones, which he designated type II, type III1, and type IV2 malformation. He speculated that this condition, which is a herniation of the posterior fossa contents below the level of the foramen magnum, is caused by hydrocephalus.1 2 Recently, many authors have used the term "Chiari type I malformation" to describe tonsillar herniation beyond 12 mm with and without hydrocephalus, resulting in confusion regarding the meaning of this term.3 4 7 9 10 it was postulated that only herniation beyond 12 mm is symptomatic, herniation between 5-10 mm is asymptomatic & herniation below 5 mm is a normal finding. Several cases of descent of the cerebellar tonsils of less than 5 mm below the foramen magnum identified through neurological examinations (tonsillarectopia) in which the patients experienced medically intractable dull pain in the occipital area and cerebellovestibular dysfunction symptoms. Some of these patients underwent posterior fossa decompression surgery, following which their clinical symptoms improved. In this study, we depict the implications of incidentally discovered herniation of the posterior inferior cerebellar lobe (cerebellar tonsil) including chronic intractable occipital dull pain and cerebellovestibular dysfunction.

2. Methods

From December 2011through March 2014, thirty cases of incidentally discovered mild herniation of the posteroinferior cerebellar lobe through foramen magnum of less than 5 mm measured from the tip of the posteroinferior cerebellar lobe to a line connecting the posterior margin of foramen magnum to the distal end of the clivus on mid-sagittal T1 or T2 FSE MR sequences. The cases had no other cerebral, cerebellar or cervical spine or cord abnormality. The 30 cases subjected to complete neurological examination for detection of the symptomatic cases & monitor such symptoms to elicit the relation of the herniation and such symptoms.

The subjects were 6 males & 24 females between 14 and 65 years old. The patients underwent MRI examination of the brain & cervical spine and the posteroinferior cerebellar lobe (cerebellar tonsil) herniation measured in the mid-sagittal plane.

All patients underwent complete neurological examination to elicit the symptoms related to the herniation of the posteroinferior cerebellar lobe.

MRI and data analysis:

All patients underwent MR examination of the brain and cervical spine which was performed.
The extent of herniation was measured in the mid-sagittal plane. The bottom of the foramen magnum was determined by drawing a line from the lowest cortical bone (visualized as a signal void) at the inferior tip of the clivus (basion) to the lowest cortical bone at the inferior border of the posterior lip of the foramen magnum (opisthion). Data are expressed as means (SD).

Neurological examination:

Neurological examinationwas performed for all symptomatic patients by one of us to assess the function of the cerebellovestibular system. The possibility of conductive or sensorineural hearing loss was ruled out from the start.

Brain stem auditory evoked potentials (BAEPs) were recorded (90 dB, 2.0 kHz click stimulation). The neurological examination consisted of two parts, the first was speech audiometry, subdivided into a speech discrimination test and a monaural speech integration test. The second part, equilibrium was examined using spontaneous nystagmus evaluation, eye tracking test (ETT; 0.33 Hz, 10°/s), optokineticnystagmus test (OKN test; acceleration 4°/S2) including vertical upward and downward OKN and horizontal optokinetic pursuit (OKP) test, and visual suppression test.

Eye tracking test (ETT) patterns were classified according to the system described by Benitez,19 as follows: pattern I, smooth sinusoidal tracing; pattern II, a few intermittent non-nystagmic movements every few seconds, superimposed on a sinusoidal curve; pattern III, fast saccadic movements superimposed on an otherwise sinusoidal curve; pattern IV, loss of the sinusoidal curvedue to an eye movement imbalance. Patterns I and II are both considered normal.

The visual suppression is a visual fixation test in which the maximal slow phase velocity of caloric nystagmus during 10 seconds in the dark (a) is compared with the maximal slow phase velocity during 10 seconds in the light with the eyes open (b). The degree of the visual suppression is quantified using the following formula:

Visual suppression (%) =a-b/a×100

Visual suppression of the slow phase velocity of caloric nystagmus is 66 (11) % in normal subjects and is considered abnormal when it is less than 40%.

3. Results

Of 30 cases of incidentally depicted herniation of the posteroinferior cerebellar lobe, 12 cases had a variety of symptoms. The mean age of the 18 subjects with asymptomatic herniation of the posteroinferior cerebellar lobe was 50 and that of the 12 patients with symptomatic herniation at the time of diagnosis was 43 years. This is higher than the reported mean age of patients with Chiari type I malformation.

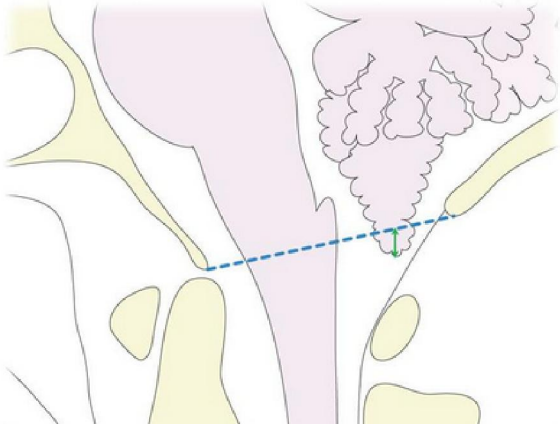

Fig. 1: schematic drawing of the method of measurement of the degree of descent of the posteroinferior cerebellar lobe through the foramen magnum. The horizontal line indicates the basion-opisthion line. The vertical line indicates the degree of herniation & measured from the tip of the posteroinferior cerebellar lobe to the basion-opisthionline.

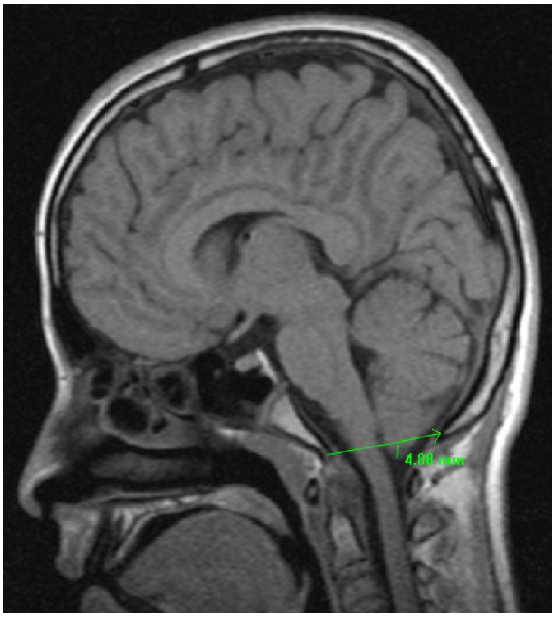

Fig.2: T1 weighted midsagittal MRI of a 14 years old patient. The horizontal line indicates thebasion-opisthionline. The posteroinferior cerebellar lobe position is 4.8 mm below the foramen magnum.

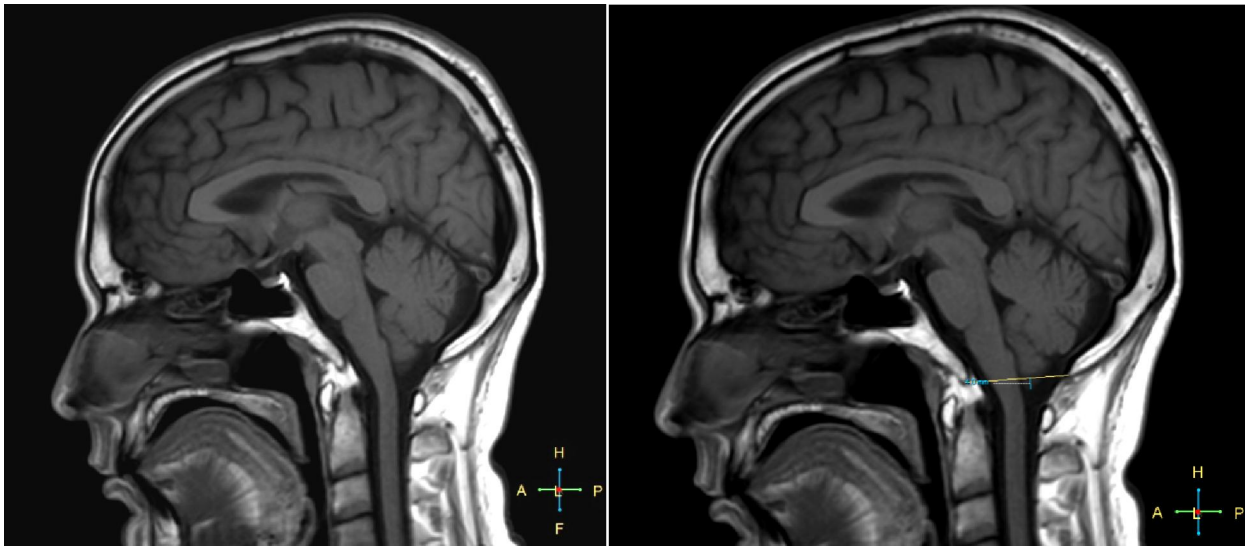

Fig. 3: T1 weighted midsagittal MRI of a 35 years old patient before and after measurement of the degree of descent of the posteroinferior cerebellar lobe. The horizontal line indicates the basion-opisthionline. The position of the tip of the posteroinferior cerebellar lobe is 4 mm below the foramen magnum. The posteroinferior cerebellar lobe is oval shaped rather than having a peg-like configuration.

The duration of symptoms in these 12 patients varied from 3 months to 10 years, with a mean of 5 years. The most common symptom was pain in the occipital area or the posterior cervical region, similar to findings in other studies.5 6 all of the patients with symptomatic posteroinferior cerebellar lobe herniation had this symptom. The next most common symptom was vertigo, followed by unstable gait, numbness of the limbs andinfrequent nausea and vomiting.

Table 1: Clinicalcharacteristic of patients with symptomatic posteroinferior cerebellar lobe herniation.

Patient number	Age/sex	Occipital /posterior cervical pain	vertigo	Ataxic unstable gait	Nausea/ vomiting	Numbness	Duration of symptoms	Position of posteroinferior cerebellar lobe
1	14/M	+	+	_	+	_	8 months	3 mm
2	32/M	+	_	+	_	+	2 years	4 mm
3	44/F	+	_	+	_	+	1.5 years	4.5 mm
4	22/F	+	_	_	+	_	3 years	3 mm
5	27/F	+	+	_	+	_	4 years	3.5 mm
6	53/F	+	+	+	_	_	7 months	4 mm
7	37/M	+	_	+	_	_	1 year	4 mm
8	57/F	+	+	+	_	+	2 years	4.5 mm
9	35/F	+	+	_	_	+	14 months	3 mm
10	65/F	+	+	+	_	+	5 years	4 mm
11	35/F	+	+	+	_	+	2 years	4.5 mm
12	41/F	+	+	_	+	+	3 years	4.5 mm

+ = present; − = absent.

Posteroinferior cerebellar lobe displacement below the foramen magnum did not exceed 5 mm in any of the symptomatic patients. None of the symptomatic patients showed syringomyelia, cerebral or cerebellar lesion responsible for the symptoms.

Table 2 shows the neurological examination findings. Five of the twelve patients had vibratory sensory disturbance. Superficial sensation was normal in all patients. Patients who complained of numbness did not always have a vibratory sensory disturbance. Three of the twelve patients had truncal ataxia, three had limb ataxia, and two had motor weakness. Three of the patients were neurologically normal. Because the symptoms and neurological findings were not specific to tonsillarectopia, we performed neurological examinations to elucidate the posterior fossa pathology.

Table 2: Symptomatic cases neurological clinical findings

Patient No	Vibratory sensory disturbance	Truncal ataxia	Limb ataxia	Motor weakness
1	+	_	_	_
2	_	+	+	_
3	+	_	+	+
4	_	_	_	_
5	+	_	-	_
6	+	+	+	_
7	_	_	_	+
8	_	-	_	_
9	_	_	_	_
10	+	_	_	+
11	+	+	_	_
12	-	_	_	_

+ = present; − = absent.

Table 3: Symptomatic cases preoperative neurological findings

Patient No	Pure tone audiometry	Speech discrimination test	Monoaural speech integration test	Spontaneous nystagmus	BAER	ETT	OKN
1	Ab	N	Ab	+	N	I	N
2	Ab	N	Ab	_	N	II	N
3	N	N	Ab	_	Ab	I	Ab
4	N	Ab	Ab	_	Ab	III	Ab
5	N	Ab	Ab	_	N	IV	Ab
6	Ab	N	Ab	+	Ab	II	Ab
7	Ab	N	Ab	+	N	III	Ab
8	N	N	Ab	_	Ab	I	Ab
9	N	N	Ab	+	Ab	I	Ab
10	N	Ab	Ab	+	N	I	Ab
11	Ab	N	Ab	+	Ab	I	Ab
12	Ab	N	Ab	_	N	II	N

BAEP = Brain stem auditory evoked potential; ETT = eye tracking test; OKN = optokinetic nystagmus test.

The 12 patients underwent posterior fossa decompression surgery, including a suboccipitalcraniectomy with duraplasty. C1 laminectomy not performed because the descent of the cerebellar tonsils was slight. The arachnoid membrane was not penetrated. Table 4 shows the results of the neurological assessment performed two months after surgery.

Nine of the patients showed improved hearing levels in the monaural speech integration tests. Spontaneous nystagmus disappeared in four of the patients. ETT and OKN disclosed an improved pattern in all patients who had shown an abnormal pattern preoperatively. The visual suppression test results were improved in four of the six patients in which it was applicable and returned to a normal pattern in four of the patients.

Table 4: Neurological findings at two months after surgery

Patient No	Pure Tone Audiometry	Speech discrimination test	Monoaural speech integration test	Spontaneous nystagmus	BAEP	ETT	OKN	Visual suppression on calories
1	Normalized	NA	Abnormal	-	NA	I	NA	Normalized
2	Normalized	NA	Improved	NA	NA	II	NA	Improved
3	NA	NA	Improved	NA	Improved	I	Improved	NA
4	NA	Normalized	Improved	NA	Improved	II	Improved	Normalized
5	NA	Normalized	Improved	NA	NA	II	Improved	Na
6	Abnormal	NA	Improved	-	Improved	I	Improved	Normalized
7	Normalized	NA	Improved	-	NA	I	Improved	Improved
8	NA	NA	Abnormal	-	Improved	I	Improved	Improved
9	NA	NA	Improved	NA	Improved	I	Improved	Normalized
10	NA	Normalized	Improved	+	NA	II	Improved	NA
11	Normalized	NA	improved	NA	Improved	I	Improved	Improved
12	Normalized	NA	Improved	NA	NA	II	NA	NA

NA = not applicable. For the other abbreviations see table 3.

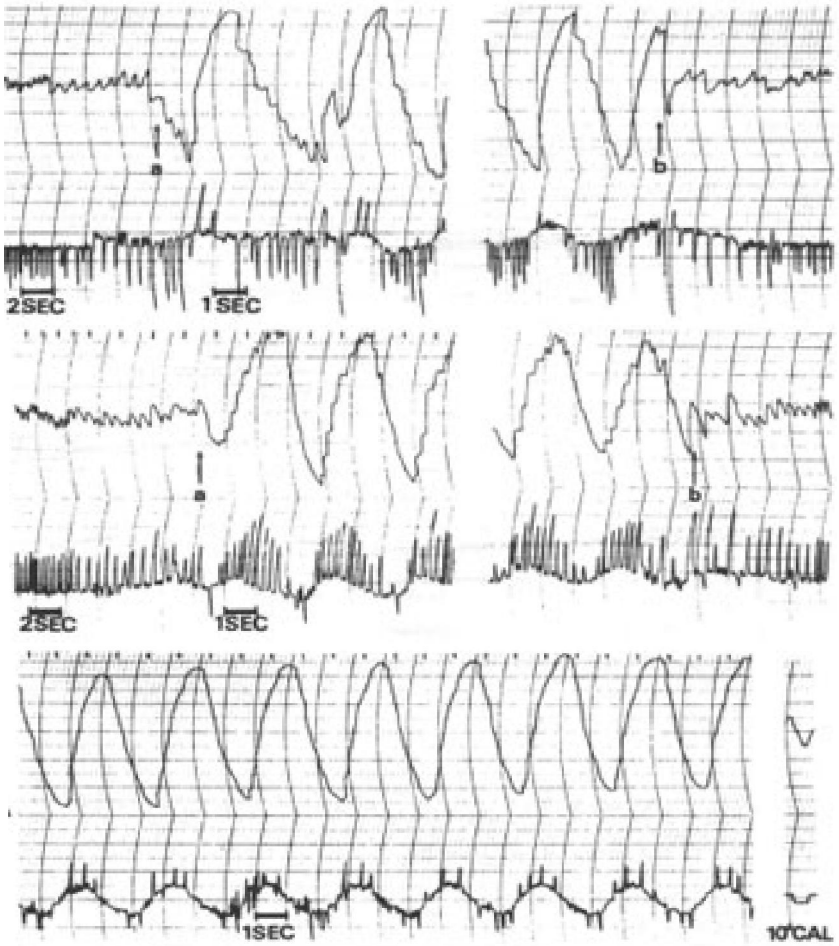

Figure 4: ETT before & after surgery for patient 3 (*An eye tracking test (ETT) shows an abnormal response (pattern III on Benitez' s classification).*

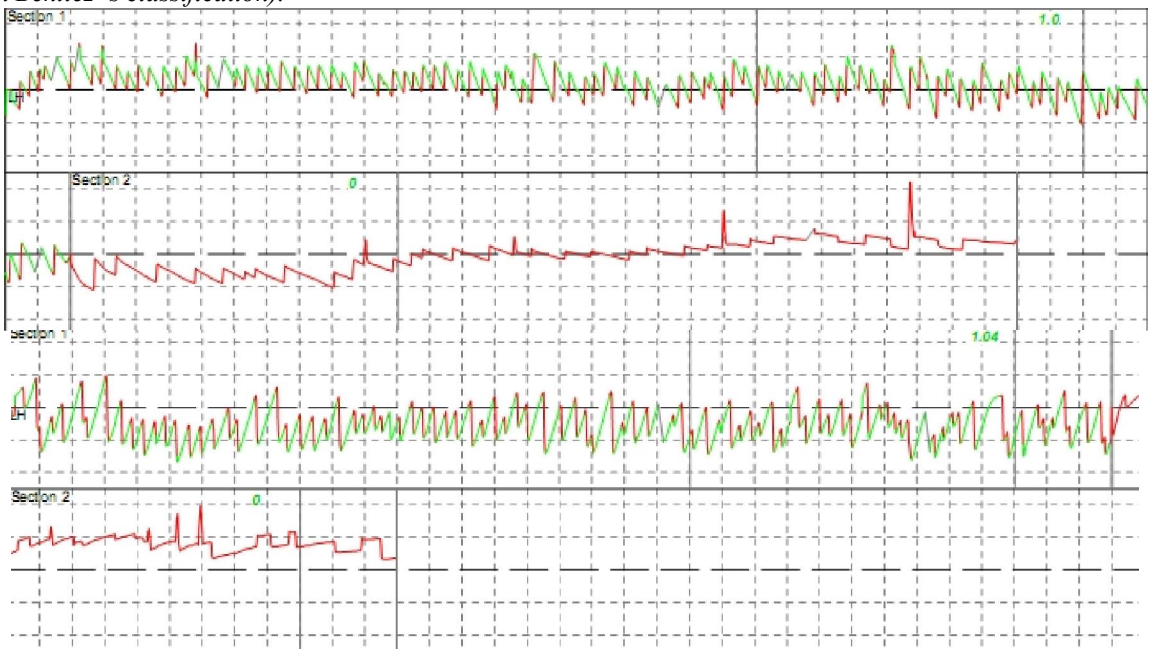

Figure 5: OKN for patient 3 before & after decompression surgery

4. Discussion

The current study showed that of 30 patients of incidentally depicted herniation of the posteroinferior cerebellar lobe through foramen magnum of less than 5 mm as depicted by mid-sagittal MRI, 12 patients had variety of symptoms.

Although several authors have measured the degree of herniation of the posteroinferior cerebellar lobe and determined that a mild degree of herniation is often of no clinical importance.(1)

Cervical spine or brain mid-sagittal plane MR provides an excellent non-invasive way of studying the posteroinferior cerebellar lobe and its relation to the foramen magnum.(2)

Aboulez et al reported that on T1 or T2 weighted images in the mid-sagittal plane, the average distance from the tonsillar tips to the foramen magnum ranged from 2.9 mm above to 3.4mm below.25 They indicated that extension of the tonsils to up to 3 mm below the foramen magnum can be considered normal and that extension is clearly pathological only when it exceeds 5 mm.(3)

Barkovich et al measured the position of the cerebellar tonsils in 200 randomly selected patients and in 25 patients with a firm diagnosis of Chiari malformation. They stated that MRI demonstration of less than 2 mm of tonsillarectopia was probably of no clinical relevance in the absence of syringomyelia.(4)

According to our findings, mild herniation of the posteroinferior cerebellar lobe less than 5 mm should not be considered a normal variant and it could cause symptoms. It is uncertain at present how tonsillarectopia could cause cerebellovestibular dysfunction. However, some neurologists have postulated that in cases of Chiari type I malformation, cerebellovestibular dysfunction is caused by compression of the vestibular and cochlear nuclei by the herniated tonsils, stretching of the cochleovestibular nerve over the porusacousticus, and ischemia caused by distortion of the posterior inferior cerebellarartery or one of its branches.5

We performed neurological examinations to elicit the etiological relation between posteroinferior cerebellar lobe herniation and the depicted symptoms. Several authors strongly emphasized the importance of careful neurological examination in the diagnosis of Chiari malformation,26–32 and that although not usually considered typical presenting complaints, neurological findings were often present and might even have preceded the development of the more severe and obvious neurological complaints.(6)

Usually patients with CNS lesions have hearing loss in the speech range, although they show no impairment of hearing of pure tones.32 Under ordinary conditions, the redundancy of the elements of information contained in an ordinary speech wave insures the intelligibility of a message, even though only one of the factors of the equation varies.(7)

Under pathological conditions, monaural speech integration ability assessed through addition of frequency distortion or interrupted distortion is impaired because compensationby the redundancy principle is impaired, even though ordinary speech discrimination abilityis normal.(8)

Based on these findings, impairment of monaural speech integration ability indicates the presence of a CNS lesion. The afferent routes in the vestibular nervemake synaptic contact with the hair cells of the macules and cristae and transmit signals to theinterneurons which control the activity of the extraocular muscles (the vestibulo-ocularreflex).(9)

Disturbance in this neuronal circuit leads to nystagmus which represents a loss of reflex activity. ETTs involve recording of eye movements in response to a peduncularvisualstimulus which results in a sinusoidal curve tracing.(10)

In Benitez's study, 65% (34 of 52) of the patients showing pattern III and 97% (36 of 37) of those showing pattern IV cameinto the CNS lesion category.19 Benitez's study also disclosed that a single test was insufficient to localize a CNS lesion, but a combination of two or three ENG findings might help to localize the lesion.(11)

Optokinetic tests, as part of a visual test battery, are very useful for baseline assessment of part of the vestibulo-ocular reflex arc.(12)

Both reflex arcs share the final oculomotor pathways from the oculomotor nuclei to the eye muscles with the fixation reflex arc, which is usually evaluated for the fixation effect on spontaneous or induced vestibularnystagmus.(13)

Horizontal visual field testing is routine in cases suggestive of a CNS lesion, although vertical visual field testing is at times also very important. A bidirectionally reduced OKN response, either symmetric or asymmetric, indicates the presence of CNS disease, usually in the brainstem or the cerebellum.(14)

In the present study, all but one of the symptomatic patients showed a bidirectional reduced OKN response. Visual fixation either suppresses or abolishes caloric nystagmus in normal subjects and patients with peripheral vestibular disorders. However, in patients with metabolic encephalopathy, or brainstem or cerebellar disease, visual fixation fails to suppress or may even enhance caloric nystagmus. This sign has been termed "failure of fixation suppression".(15)

Alpert stated that failure of fixation suppression was highly correlated with posterior fossapathology, provided that there had been no recent drug intake.22 Six of the twelve patients in the present study scored

below 40% in the visual suppression test. There has been much speculation regarding the anatomical site of visual-vestibular interaction.(16)

Yules et al stressed the importance of the pontine and midbrain reticular formation in mediating the vestibulo-ocular reflex arc.(16)

However, **Lorente** reported that in the vestibularsystem, there is no "localisation" of reflexes in anatomical nuclei, and that the whole system is a functional unit.(17)

The symptomatic posteroinferior cerebellar lobe herniation is associated with no single pattern of neurological findings; rather, a combination of findings suggesting the presence of a CNS lesion contributes to the diagnosis.

The differential diagnosis of symptomatic posteroinferior cerebellar lobe herniation must include cervical headache. Stimulation of parts of the vertebral column, the cervical muscles and their attachments to bone, the cervical nerve roots, and the vertebral arteries under such conditions as arthritis, trauma, infection, tumor, dissection, or atherothrombotic occlusion of the vertebral arteries clearly gives rise to pain.(18)

Brain MRI may be useful for distinguishing posteroinferior cerebellar lobe herniation from these conditions. The differential diagnosis must also include vertebrobasilar insufficiency.

MRI disclosed no brainstem or cerebellar infarct in the present cases. Angiography of the vertebrobasilar system is not always necessary to rule out vertebrobasilar insufficiency.

Herniation of the posteroinferior cerebellar lobe through the foramen magnum less than 5 mm may cause symptoms. Carful neurological examination is necessary to verify the cerebello-vestibular dysfunction.

Further research on symptomatic herniation of the posteroinferior cerebellar lobe will contribute to elucidate its etiological significance.

Based on the radiological & neurological findings, we conclude that herniation of the posteroinferior cerebellar lobe less than 5 mm could cause symptoms.

Conclusion

Subtle herniation of posteroinferior cerebellar lobe less than 5 mm in symptomatic patients should be submitted to neurological evaluation to elucidate the relationship of the herniation and such symptoms. Some of symptomatic cases might worth surgical intervention.

Neurological assessment found necessary to verify the etiological relation between the herniation and various symptoms.

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