**LETTERS**

**Acute rotatory vertigo caused by a small haemorrhage of the vestibular cortex**

Central rotatory vertigo is in most cases caused by a lesion of the cerebellum or brain stem. We describe a patient with acute rotatory vertigo following a small haemorrhage in the left medial temporal gyrus, which probably injured the vestibular cortex.

**Case history**

A 53 year old woman suddenly experienced leftwards directed rotatory vertigo in the yaw plane and nausea without vomiting. She felt unsteady and had short lasting slurring of her speech. She had no hearing loss or tinnitus. On examination, she could stand unaided but tended to fall after a short while, without a directional preponderance. Gait was severely unsteady and she could not walk unaided. The rotatory vertigo was worse when she was sitting upright than when lying down in bed. Vertigo was also increased by head movements.

Examination of the cranial nerves showed no abnormalities; specifically there was no nystagmus or hearing loss and the eye movements were normal. Neurological examination showed a minimal asymmetry of the limbs (motor and sensory function, coordination, and reflexes) was normal. Electroencephalography showed no abnormalities, supporting a non-epileptic cause of the vertigo.

Magnetic resonance imaging (MRI) on sagittal T1 weighted and transverse T2 weighted spin echo and FLAIR images showed a small (2.0 × 1.3 cm) haemorrhage in the left medial temporal gyrus, adjacent to the superior temporal gyrus (fig 1). There were no lesions of the brain stem or cerebellum. The appearance of the temporal lesion was consistent with haemorrhage from a small cavernous haemangioma.

Vertebrobasilar function was evaluated by electronystagmography (gaze, saccade, smooth pursuit, optokinetic, torsion swing, velocity step, and caloric tests; search for spontaneous nystagmus), and by video-oculography (ocular counter rolling induced by lateralolateral and eccentric rotation). No abnormalities were found. Additional testing included the Romberg test, galvanically induced body sway, and the subjective vertical. On Romberg testing there was abnormally increased body sway (especially with the eyes closed). The patient could stand long enough with the eyes closed to measure the galvanically induced body sway, which had normal excitability. The subjective visual vertical showed a 6° right-body sway, which had normal excitability. The patient could stand long enough with the eyes testing there was abnormally increased body sway, and the subjective visual vertical. On Romberg testing, there was no nystagmus, and by video-oculography (oculopursuit, optokinetic, torsion swing, velocity step) there was no nystagmus or hearing loss and the eye movements were normal.

Functional brain studies have shown a well defined vestibular cortical system. In all likelihood, a similar system probably also exists in humans, including, as in primates, several cortical areas. However, one has to be careful in extrapolating results from primates to humans, since human studies are important to further elucidate the existence and location of the human vestibular cortical system.

The vestibular cortical system seems to be distributed among several multisensory areas in the parietal and temporal cortex, and is integrated in a larger network for spatial attention and sensory-motor control. The parieto-insular cortex is postulated to be the core region within the vestibular cortical system; representation is bilateral, with a right hemispheric dominance. Recent research seems to indicate that there might be no specific vestibular cortex, contrary to the visual and auditory systems. Electrophysiological recordings of vestibular cortical neurones, positron emission tomography, and fMRI brain activation studies during caloric and galvanic stimulation all confirm the multisensory character of cortical areas that receive a substantial vestibular input. One can understand this when one realises that during motion not only the labyrinths but also the visual and proprioceptive systems will be stimulated. This could make a unimodal vestibular cortex unnecessary.

We are aware of one other reported patient with rotatory vertigo and a cortical lesion on MRI. That patient, however, had two cortical lesions: the main lesion was an infarct located in the right posterior insula involving the long insula and transverse temporal gyrus; the other lesion was in the right parietal cortex. We believe that our patient is the first reported case of rotatory vertigo resulting from a lesion (haemorrhage) of the medial temporal gyrus, adjacent to the superior temporal sulcus. Functional brain studies have shown that the human vestibular cortical system may be located in the superior temporal region posterior to the auditory area, probably in the superior temporal gyrus. The results of functional brain studies, the previously reported patient, and our own patient indicate that the human vestibular cortical system is located in several adjacent cortical areas: the superior temporal gyrus, the long insular and transverse temporal gyrus, and the medial temporal gyrus.

**Comment**

Our patient is of interest for two reasons. First, she demonstrates that acute rotatory vertigo may be caused by a lesion of the cerebral cortex, supporting the existence of a cortical area in humans with a substantial vestibular input. Second, she could be considered as an “experiment of nature”: a small lesion confined to a particular brain structure, enabling precise localisation of an area in the cortex that seems to be very much engaged with the vestibular system.

The exact location of the vestibular cortex in humans has not yet been established.

**References**

Coexistent Lewy body disease in a case of “variant visual of Alzheimer’s disease”

Posterior cortical atrophy or the “variant visual” of Alzheimer’s disease is a clinical syndrome with visual agnosia, some or all of the characteristic features of Balint’s syndrome, transient cortical sensory aphasia, and Gerstmann’s syndrome. Although pathologically heterogeneous, several necropsy studies on patients with posterior cortical atrophy have shown Alzheimer’s disease pathology. We report a patient who presented with the features of posterior cortical atrophy who later developed mild parkinsonism, visual hallucinations, and dementia. Neuropathological evaluation revealed combined Alzheimer’s disease and Lewy body disease.

Case report
A right handed retired diesel mechanic, with 12 years of formal education, was referred for evaluation of an “unusual dementia.” His difficulties started at the age of 58 with the initial symptoms of visuospatial dysfunction. Initially he was not able to fill out bank deposit slips or write numbers correctly. He had been an avid reader but had to re-read material in order to comprehend it, and he had to use a card to keep his eyes focused when reading. He was not able to locate the refrigerator door handle until he groped over the surface to find it.

When he was 62, his wife noted that he moved in a stiff manner, did not swing his left arm, and acted “like a little old man.” At age 67, he developed well formed visual hallucinations (he would see bugs, spiders, and people) and paranoid delusions (he expressed concern that people were tearing away his home). He developed personality changes and at times became disorientational. He became entirely dependent on his wife for all of his activities of daily living. No features of REM sleep behaviour disorder were ever noted by the family.

The initial neurological evaluation at the Mayo Clinic when he was 67 revealed a complete Balint’s syndrome, a partial Gerstmann’s syndrome, and impairment on visuospatial tasks and recall. On language examination he had paraphasic errors and neologisms. He also showed bradykinesia, a slow wide based gait with reduced arm swing bilaterally, mild generalized rigidity, postural but not resting tremor, and rigidity of the left arm. He had limited upgaze but preserved downward and horizontal gaze. Visual acuity was 20/80 and 20/100 in the right and left eye, respectively. There was no alien limb phenomenon, dystonia, or myoclonus.

Neuropsychological testing showed impairment in verbal skills and verbal memory and the inability to complete the visual tasks. Magnetic resonance imaging and single photon emission computed tomography of the brain showed, respectively, marked asymmetrical (left more than right) parietal-occipital cortical atrophy and hypoperfusion.

Towards the end of his life, he became wheelchair bound and was transferred to a chronic care facility. He developed more behavioural problems, declining vision, and persistent visual hallucinations and delusions. He was unable to recognise family members by sight or sound. He died at 71 years of age.

His past medical history was only significant for a total thyroidectomy for cancer, for which he was on thyroid replacement. There was no family history of any neurodegenerative disorder.

At necropsy examination, standard brain fixation and dissection was undertaken. Tissue sections were cut and stained with haematoxylin and eosin, Bielchowsky silver stain, and immunohistochemically with anti-bodies to tau (Endogen-AT8), amyloid protein, and synuclein (Zined-LB509). The brain weighed 1136 g. Focal, asymmetrical (left greater than right) parieto-occipital cortical atrophy and mild pallor of the substantia nigra were observed. The basal ganglia, thalamus, and cerebellum appeared normal. Microscopically, moderate to frequent diffuse and neuritic plaques and frequent neurofibrillar tangles were seen in limbic structures. Accentuated neuronal loss and increased neurofibrillary tangle density were noted in the parietal and occipital lobes. The findings satisfied criteria for Alzheimer’s disease by Braak and Braak staging (stage V/VI) and by the National Institute on Aging and Reagan Institute working group diagnostic criteria for the neuropathological assessment of Alzheimer’s disease (high likelihood). In addition, synuclein positive Lewy bodies, pale bodies, and Lewy neurites were found in the substantia nigra, amygdala, entorhinal cortex, and cingulate gyrus; however, the substantia nigra was less affected than the limbic structures, where synuclein pathology was severe. These findings are consistent with a diagnosis of Alzheimer’s disease.

Comment
The clinical syndrome of posterior cortical atrophy is characterised by prominent dys-function of the neuronal networks in the biparietal and occipital cortices and does not imply an underlying pathology. Neuropathological examination in most cases shows neurofibrillary tangles and neuritic plaques characteristic of Alzheimer’s disease, but with a higher concentration of the pathology located in the primary visual cortex and visual association cortices. The predominant features of posterior cortical atrophy are followed by dementia more typical of Alzheimer’s disease. Visual hallucinations and parkinsonism are distinctly uncommon but are recognised features, in addition to fluctuations in cognition that are considered characteristic of dementia with Lewy bodies. Pathologically, the latter is characterised by the presence of Lewy body disease, with limbic or neocortical Lewy bodies.

Our case presented with the typical features of posterior cortical atrophy, and findings of Alzheimer’s disease and Lewy body disease were revealed on neuropathological examination. To our knowledge, there have been no previous pathological reports of cases of posterior cortical atrophy with coexisting Lewy body disease. The visual hallucinations and parkinsonism in our patient were consistent with dementia with Lewy bodies, and there was evidence of transitional Lewy body disease at necropsy. The other major features of posterior cortical atrophy would be consistent with the prominent Alzheimer’s pathology in occipito-parietal cortices.

Interestingly, few patients with posterior cortical atrophy experience visual hallucinations as an initial symptom despite the marked pathology affecting the primary visual and visual association cortices. Our patient also had clinical features suggesting corticobasal degeneration. Although there are reports of prominent visuospatial impairment or Balint’s syndrome in clinically diagnosed and pathologically diagnosed cortico-basal degeneration, visual hallucinations are almost non-existent in corticobasal degeneration. This suggests that visual hallucinations and parkinsonism in the setting of cognitive impairment reflect underlying Lewy body disease rather than corticobasal degeneration. Our case supports this contention. We suggest that underlying Lewy body disease should be considered in any case of posterior cortical atrophy associated with parkinsonism and particularly visual hallucinations.

Acknowledgements
Supported by grants AG 16574 and AG 07216 from the National Institute on Aging. We extend our appreciation to the family for participating in research on aging and dementia.

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Competing interests: none declared

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Reversible collateral disconnection syndrome in internal hydrocephalus
A 74 year old woman was referred to the neurological department for evaluation of progressive gait disturbance. On admission she complained of a fall and loss of use of her left arm for six months. There were no spontaneous movements without the patient’s intention, but she had always to rely on visual cues. For example, when she was cooking, eating, or doing exercises with her home trainer she had to watch her left arm to

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be sure of its movement. On examination she
was alert, fully oriented, and cooperative.
Snout and palomental reflexes were posi-
tive. There was no visual, somatosensory,
or auditory extinction. Motor examination
revealed a mild left sided facial droop and a left
sided pronator drift but strength was full and
symmetric and there was no lack of spontane-
ous movement in the left upper limb. The
plantar response was extensor on the left. Gait
was slow, unsteady, and wide based. The steps
were short with reduced step height. Neuro-
psychological assessment showed fluent
speech without dysarthria. Performance in
verbal and non-verbal material was diminished,
and colour-word interference was slightly increased. Long term memory
was slightly deficient for verbal and non-
verbal material. Visuo-constructive abilities
were normal and there was no spatial neglect.
There was no apraxia of the left hand for ges-
tures neither on command nor for imitation.
Also, there was no agraphia or tactile anomia
of the left hand. She could perform bimanual
tasks without evidence of intermanual con-
flict. She did not exhibit grasp reflex in either
upper limb, and there was no compulsive
manipulation of objects.

There was, however, an inability of one
hand to imitate the posture of the opposite
hand when visual cues were removed. Fur-
thermore, there was an inability to distin-
guish the left hand from an examiner’s hand
when these were placed in the patient’s right
hand behind the back, which is known as
“strange hand” (or “signe de la main
étrangère”). Additionally, an inability to
accurately locate touch of the fingers was found: the
patient was blindfolded and touched by the
examiner on one finger either of the left or the
right hand. Then she was asked to point to the
location of touch with the contralateral hand.
The accuracy was impaired for both directions
but especially for right to left pointing. How-
ever, she was correct when asked to point
to the location of touch on the face or trunk.

Magnetic resonance imaging showed inter-
nal hydrocephalus (fig 1A) and an old lacunar
ischaemic lesion in the right anterior limb of
the internal capsule. Transcallosal inhibition
was assessed by transcranial magnetic stimu-
lation (TMS) as described previously and
showed a deficient inhibition particularly
from left to right (upper panel “left A” in fig
1B). Cerebrospinal fluid (CSF) pressure was
normal during lumbar puncture. After re-
moval of 50 cc CSF the alienation of the left
arm, the “signe de la main étrangère” and the
impaired cross replication of hand postures

Figure 1  [A] Magnetic resonance image (T2 weighted) showing internal hydrocephalus with thinning of the corpus callosum. [B] Recordings
of the rectified tonic electromyographic (EMG) activity of the first dorsal interosseous muscle (ID1) after ipsilateral focal transcranial magnetic
stimulation (TMS) are shown. Traces of three trials are superimposed over each other. The recording and stimulus sides as well as the latencies
and durations of the transcallosal inhibitory responses (TI) are indicated. Upper panels: the findings before lumbar puncture are shown. TI is
normal after TMS of the right hemisphere (normal values [mean (SD)]: latency 35.6 (7.2); duration 24.8 (5.4)), but missing on the left (arrow).
Lower panels: results after withdrawal of 50 cc cerebrospinal fluid (CSF). TI could be revealed on both sides. The latencies after the lumbar
puncture differed clearly and were prolonged after TMS of the left hemisphere.

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completely disappeared. There was also gait improvement, and for a walking distance of 20 m the number of required steps decreased by five and the time needed by 10 seconds. In accord with the clinical finding there was a neurophysiological restoration of the deficient transcallosal function, which now showed a normal inhibition of the tonic EMG activity (lower panels in fig 1B).

Within three weeks the alienation of the left hand and the gait disturbance was subsided. When readmitted to our department identical callosal dysfunction could be ascertained. After shunt implantation there was permanent recovery of callosal dysfunction and gait disturbance.

Various callosal disconnection signs may follow a structural lesion of the corpus callosum. Among others, there is the sign of impaired cross replication of hand postures when visual cues are removed, the inability to cross locate touch of the fingers when blindfolded, and the “signe de la main étrangère” (the sign of the foreign hand) describing a failure to recognize self ownership of the left hand while visual cues are removed.

We describe a callosal disconnection syndrome with a “signe de la main étrangère”, an impaired unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT), and cluster headache (CH). Combinations of cluster headache and chronic paroxysmal hemicrania with trigeminal neuralgia have also been described and have been called cluster-tic syndrome or CPH-tic syndrome. In order to diagnose TACs, it is essential to record the case history carefully. Only rarely have intracranial lesions such as aneurysms or tumours been observed in association with TACs. In the majority of cases, no brain abnormalities are found using conventional imaging.

We describe a three year old girl who suffered attacks of severe right sided temporal pain and autonomic disturbances and in addition neuralgic shooting pains associated with a pilocytic astrocytoma in the pons and medulla oblongata, extending to the upper cervical cord. The attacks disappeared once the tumour had been debulked.

Case report
A three year old girl presented with a history of extremely painful right sided temporal headache attacks since the age of one year. During these attacks she would grab her right ear and cry intensely. Her eyelids were slightly swollen, with rhinorrhea on the right side. It seemed that she was in continuous pain, with superimposed paroxysms of very intense pain, lasting seconds, during which she rocked back and forth (the video recording is available at the JNNP website: www.jnnp.com). These attacks occurred spontaneously at any time of day or night with no particular regularity or trigger points. Magnetic resonance imaging (MRI) of the brain revealed a tumour in the pons, extending to the medulla oblongata and cerebral myelum (fig 1A), with a synrix in the cerebral myelum. On the transverse slice, the tumour extended to the cerebropontine cistern on the right side (fig 1B). Repeated physical examinations after six weeks showed hyperreflexia of the right arm and leg and positive Babinski reflexes on both sides. After debulking the tumour, the attacks resolved completely and neurological examination normalised. Pathological examination of the tumour revealed a pilocytic astrocytoma. The patient has remained headache-free following neurosurgery during two years of follow up. Neurological examination has remained normal during this period.

Comment
The diagnosis of primary headaches associated with autonomic symptoms, such as cluster headache or chronic paroxysmal hemicrania, is based on the patient’s history, because diagnostic tests are not available. As shown here, a video recording of the attacks may be very helpful when patients are unable to describe the attacks in detail themselves. In this case the autonomic symptoms during the headache and the sudden additional shooting pains were recognised by the parents and the physician after studying these video recordings. Although the attacks lasted 12 to 24 hours, the combined headaches best resembled a combination of a TAC-like syndrome in association with trigeminal neuralgia or idiopathic stabbing headache (ISH). It is difficult to distinguish trigeminal neuralgia from ISH in this case, as both headaches only last seconds, may occur many times per day, and...
may be located in the first division of the trigeminal nerve. ISH is often described in association with other primary headaches and usually responds to indomethacin, in contrast to trigeminal neuralgia. It cannot conclude with certainty that the stabs in this case are ISH, because indomethacin was not used.

The attacks were found to be associated with a pilocytic astrocytoma in the pons that extended to the medulla oblongata on the same side as the attacks. Similar cases have suggested involvement of vascular or neoplastic intracranial lesions in TACs. A causal relation between the symptoms and the lesions was not established in all cases, but involvement of the intracranial lesions with trigeminal structures was suggested. One patient with an upper cervical meningioma had cluster headache attacks, possibly caused by direct compression of the C1–C3 rootlets or the trigeminal nucleus. Furthermore, cluster tic syndrome has been reported in association with intracranial pathologies. In this case, direct compression of the trigeminal root by the basilar artery entering deep into the cerebellopontine cistern was suggested as a cause of the pain. It is not clear how the lesion in our case caused paroxysmal symptoms, but disappearance of the symptoms after debulking suggests a causal relation.

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Competing interests: none declared

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Autoimmune myasthenia gravis after cardiac surgery
Autoimmune myasthenia gravis (MG) is a heterogeneous disorder. In young women, the thymus gland is often hyperplastic, and the patients respond well to thymectomy. However, in the increasing number of patients over the age of 40 years, predominantly men, thymic hyperplasia is uncommon, and there are no clear aetiological clues.

We diagnosed MG in three male patients who had undergone cardiac surgery between three and ten weeks before developing symptoms. Table 1 summarises their main features. The patients presented with ocular, bulbar, and mild generalised weakness (Osserman classification grade I to IIb). None of them was taking antiarrhythmic agents or other drugs impairing neuromuscular transmission and none of them had any thyroid or other autoimmune disorder. Two showed typical decremental responses to repetitive stimulation on EMG, and all three had positive levels of antibodies to acetylcholine receptors, which are diagnostic for MG. None had either postoperative recovery difficulties that might have suggested pre-existing MG. All three patients were treated with cholinesterase inhibitors, two also required corticosteroids (for the plan of treatment, see Sanders and Scoppetta1). and one required plasma exchange. The patients have now been followed up for up to seven years and all still require some medication and have positive AChR antibodies.

The development of MG within a few weeks of cardiac surgery is intriguing, and has not been reported before. The presentation of MG in these subjects may be purely coincidental but, because of the short delay between surgery and presentation, it is difficult to escape the conclusion that the cardiac surgery precipitated the condition or exacerbated subclinical disease. During cardiac surgery there is damage to the atrophic thymic remnants that are present in the anterior mediastinal fat. The thymus contains muscle-like cells, called myoid cells, which express whole AChR molecules. Release of thymic AChR could increase an existing subclinical antibody response, or allow AChR to be presented de novo to the immune system; the lack of postoperative difficulties, which are commonly encountered in undiagnosed MG, suggests that the second hypothesis is most probable. The AChR is very immunogenic; for instance, in mice, intraperitoneal injection of purified murine AChR without adjuvants can result in the typical antibodies and clinical expression of the disease.

Further studies should investigate the presence of muscle weakness and positive titres of acetylcholine receptor antibodies after cardiac surgery, and could compare with similar measurements after other forms of major surgery in this age group. Equally, as other autoantigens are also expressed in the thymus, the presence of other autoantibodies or signs of other autoimmune diseases should be sought.

Competing interests: none declared.

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References

Table 1: Clinical and laboratory features of three patients with autoimmune myasthenia

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<tr>
<th>Sex, age at the onset of MG</th>
<th>Heart surgery</th>
<th>Time since surgery to onset of MG</th>
<th>Presenting features; (Osserman group at onset)</th>
<th>Clinical diagnosis</th>
<th>AChR antibodies normal value &lt;0.8 nmol</th>
<th>Treatment and response</th>
<th>Follow up and evolution AChR-ab present titre</th>
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</thead>
<tbody>
<tr>
<td>M, 57 Triple aorta-coronaric bypass</td>
<td>3 weeks</td>
<td>Pharynx, double vision; (l)</td>
<td>Response to im neostigmine and oral pyridostigmine</td>
<td>1.9 nmol</td>
<td>Cholinesterase inhibitors; good</td>
<td>6 years, fluctuating ocular MG</td>
<td></td>
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<tr>
<td>M, 58 Double aorta-coronaric bypass</td>
<td>10 weeks</td>
<td>Dysthria, dysphagia and arm weakness; (llb)</td>
<td>Decrease on repetitive stimulation, response to iv edrophonium</td>
<td>8.0 nmol</td>
<td>Cholinesterase inhibitors; mild</td>
<td>7 years, fluctuating generalised MG</td>
<td></td>
</tr>
<tr>
<td>M, 65 Triple aorta-coronaric bypass</td>
<td>10 weeks</td>
<td>Double vision, dysphagia and dysphoria; (llb)</td>
<td>Decrease on repetitive stimulation, response to im neostigmine and oral pyridostigmine</td>
<td>12.0 nmol</td>
<td>Cholinesterase inhibitors; mild</td>
<td>8 months, mild generalised MG</td>
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Chiari I malformation mimicking myasthenia gravis

Chiari I malformation is accompanied by a variety of symptoms and signs suggesting brain stem, cerebellar, or cervical spinal cord lesions. The most common symptoms include headache, neck pain, sensory loss, and ataxia. Dysphagia occurs in 5–15% of the patients and it may be the only presenting symptom. Progressive dysphagia caused by Chiari I malformation, mimicking myasthenia gravis, has been reported in this journal in 1996 and 2002. Dysphonia may occur rarely, but it has not been described as an early symptom. Pain and stiffness in the posterior neck is a common feature, but severe neck pain with worsening leading to dropped head syndrome has not so far been reported in Chiari I malformation.

Case report

A 13 year old girl was admitted to our department of neurology four weeks after adenoidectomy under general anaesthesia, because of progressive difficulty in lifting her chin off her chest, together with dysphagia and dysphonia. There was no pain or stiffness in the posterior neck. Computed tomography of the head was reported as normal. There was mild fluctuation of the dysphagia and dysphonia during the day, with worsening of the dysphonia after prolonged conversation or after reading in a loud voice. There was no sleep disturbance.

Neurological examination revealed an increased deep tendon reflexes in all four limbs. Routine serum biochemistry and blood count were unremarkable.

On the basis of the history and clinical data, myasthenia gravis was suspected, so electromyography of proximal and distal muscles in all four limbs was normal. The cerebellar tonsils through the foramen magnum revealed herniation of the cerebellar tonsils.

Figure 1

Brain and cervical spine magnetic resonance imaging: herniation of the cerebellar tonsils through the foramen magnum.

C2–C3 vertebral level (fig 1). No syrinx or hydrocephalus was demonstrated. A diagnosis of Chiari I malformation was made.

A posterior fossa craniectomy was undertaken. The anaesthesia and operative procedures were uncomplicated. Two days after the operation, the dysphagia and dysphonia improved and one month later there had been a remarkable improvement in the neck extensor muscle weakness.

Comment

Dropped head may be a part of a generalised neuromuscular disorder, such as myasthenia, polymyositis, amyotrophic lateral sclerosis, adult onset nemaline myopathy, or chronic inflammatory demyelinating polyneuropathy. Our patient had a dropped head “plus” syndrome secondary to Chiari I malformation. The neck pain and stiffness were not referred. This case report suggests that one should suspect Chiari I malformation in patients with neck extensor muscle weakness, especially if this is associated with lower cranial nerve impairment. We postulate that the symptomatology in this girl may have been the result brain stem impairment secondary to the compression caused by the malformation. Dysfunction of the lower cranial nerves and the higher cervicospinal roots by a retrograde effect of the compression may be the pathogenic explanatory mechanism. The rapid disappearance of the symptoms after posterior fossa decompression supports this hypothesis. Fluctuations of dysphonia and dysphagia may, on the other hand, reflect variations in intracranial pressure. Recently, a presentation of a previously asymptomatic Chiari I malformation was reported following a flexion injury to the neck by a trivial car accident. In our patient, it is possible that slight cervical trauma during anaesthesia for her adenoidectomy may have brought to light the underlying congenital abnormality. Mootry et al have reported eight cases, initially diagnosed as ocular myasthenia on the basis of clinical features and response of anticholinesterase agents, in which an intracranial mass lesion was active. In addition to myasthenia gravis was later found. Patients with dropped head and lower cranial nerve involvement, although presenting with a clinical history strongly suggestive of myasthenia, should be carefully evaluated and the diagnosis of Chiari I malformation considered. A response to anticholinesterase agents observed clinically or recorded electrically has been reported in a variety of disorders, including Eaton–Lambert syndrome, botulism, and transverse myelitis, and even in patients with intracranial mass lesions. The partial response to anticholinesterase drugs in our case reinforces the view that it is unlikely to base the diagnosis of myasthenia gravis purely on a positive pharmacological test.

Competing interests: none declared.

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References


Expanding cerebral cysts (lacunae): a treatable cause of progressive midbrain syndrome

A progressive motor defect presenting in adulthood is an ominous sign, being often associated with either neoplasia or neurodegenerative diseases. Notable if very rare is the expansion of this poorly prognosis are cerebral expanding lacunae or, as they are sometimes called, benign intraparenchymal brain cysts. These are intraparenchymal cavities without an epithelial lining, filled with cerebrospinal fluid (CSF), located in the thalamo-mesencephalic arterial territory. They expand their nature is demonstrated by their progressive clinical course and by the frequent complication of aqueduct stenosis and tinecricentric hydrocephalus. We present a case of progressive midbrain syndrome associated with expanding cysts, which was successfully treated by neuroendoscopy.

Case report

A 43 year old woman with an unremarkable clinical history presented in 1996 with progressive resting tremor and weakness of the left arm. The tremor persisted during posture maintenance and action. Within a year the motor problems extended to the left leg. Brain magnetic resonance imaging (MRI) showed large (10 to 20 mm) well defined lesions with signal intensities identical to CSF occupying most of the right thalamo-mesencephalic region. There was no contrast enhancement either in the lesions or in the surrounding tissue. The ventricular spaces were only mildly

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enlarged. A search for cystic lesions elsewhere in the body was negative.

In the following year disturbance in posture and diplopia in the right lateral gaze became apparent. The tremor resolved spontaneously, but the left hemiparesis worsened. She was referred for neurosurgical evaluation.

At admission, objective findings were a left hemiparesis (less than arm), hemirigidly, severely reduced automatic movements and left bradykinesia, brisk tendon jerks on the left, diplopia on rightward gaze, and Parinaud syndrome. The patient was alert and oriented, with intact gross cognition.

A preoperative brain MRI showed multiple large cystic lesions occupying the right paramedian pontomesencephalic region and smaller lesions in the right thalamus (fig 1, panels A and B). CSF flow sequences revealed aqueductal stenosis and slight triventricular hydrocephalus.

Surgical procedure

The patient underwent a surgical endoscopic procedure. A flexible 2.5 mm neuroendoscope (Storz) was inserted through a burr hole, and the third ventricle was cannulated with a 3.9 mm sheath. The floor of the ventricle posterior to the mammillary bodies appeared severely deformed by a large cystic mass that did not allow access to the aqueduct. The cystic mass was coagulated and opened into the third ventricle. A fragment of the cyst wall was taken for pathology, which showed normal neuropil with few amilolocytic bodies, no epithelial lining, and no signs of old or recent haemorrhage.

Once opened, the inside of the cyst revealed a multilobular structure. The flux of fluid towards the ventricle was obstructed, indicating multiple intercommunicating lesions under moderate pressure. The last surgical procedure was a third ventriculocisternostomy 3.5 mm anterior to the mammillary bodies.

After the operation there was a transitory disturbance of convergence and limitation of lateral eye deviation, which resolved spontaneously on day 3.

Follow up

At three months the patient showed a remarkable improvement in motor performance but there was reappearance of a modest resting tremor in the left hand. MRI documented a mild reduction in cyst volume and moderate reduction in ventricular size (fig 1, panels C and D). At 18 months the patient was neurologically normal except for the mild resting tremor of the left hand. She had lateral eye deviation, which resolved spontaneously, and diplopia in the right lateral gaze became apparent. The tremor resolved spontaneously, but the left hemiparesis worsened. She was referred for neurosurgical evaluation.

A progressive disorder of cognition and hydrocephalus. Postoperative sagittal (C) and coronal (D) T1 weighted MR images. There is a slight reduction in volume of the lesions, with partial resolution of the hydrocephalus.

References