

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 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 × 1.5 cm) haemorrhage in the left medial temporal gyrus, adjacent to the superior temporal sulcus (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.

Vestibular 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 lateroflexion and eccentric rotation). No abnormalities were found. Additional testing included the Romberg test, galvanically induced body sway, and the subjective visual 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° rightwards (clockwise) tilt and a reduced accuracy (SD 4.7°). The neurological symptoms and signs gradually disappeared over a few weeks.

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.

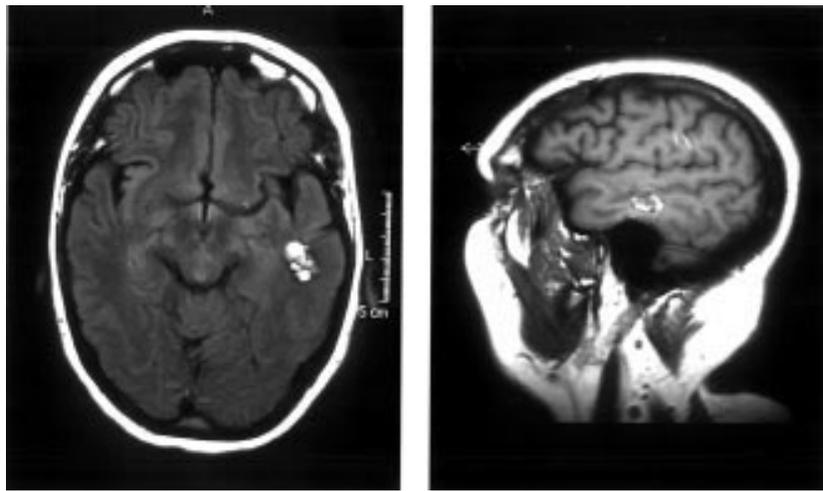


Figure 1 Transverse T2 weighted, fluid attenuated inversion recovery (FLAIR) image (left) and sagittal T1 weighted spin echo image (right). Both show a small popcorn shaped area of increased signal intensity demarcated by a rim of decreased signal intensity (haemosiderin), located in the left medial temporal gyrus. The combination of recent blood products and older haemorrhagic residues is consistent with the diagnosis of a cavernous haemangioma.

Primate studies have shown a well defined vestibular cortical system.¹ In all likelihood, a similar system probably also exists in humans, including, as in primates, several cortical areas.^{1,2} However, one has to be careful in extrapolating results from primates to humans,² so 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.³ 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.⁴⁻⁶ 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.³

We are aware of one other reported patient with rotatory vertigo and a cortical lesion on MRI.⁷ That patient, however, had two cortical lesions: the main lesion was an infarct located in the right posterior insula involving the long insular 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.^{8,9} The results of functional brain studies, the previously reported patient,⁷ 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.

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

Posterior cortical atrophy or the “visual variant” of Alzheimer’s disease is a clinical syndrome with visual agnosia, some or all the components of Balint’s syndrome, transcortical sensory aphasia, and Gerstmann’s syndrome.¹ Although pathologically heterogeneous, several necropsy studies on patients with posterior cortical atrophy have shown Alzheimer’s disease pathology.¹ We report a patient who presented with the features of posterior cortical atrophy who later developed mild parkinsonism, visual hallucinations, and delusions. 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 insidious onset 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 unsuccessfully used 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.

His wife revealed that when he was 61 he was having difficulties working as a mechanic. Also, he could not see other cars and obstacles while driving, and he stopped driving at the age of 63 after being involved in two motor vehicle accidents. He developed progressive difficulties with performing calculations, writing, receptive language, and recent memory. Despite the cognitive difficulties, he retained insight in his disorder.

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 was confrontational. 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 generalised rigidity, postural but not resting tremor, and right limb apraxia. 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) parieto-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, Bielschowsky silver stain, and immunohistochemically with antibodies to tau (Endogen-AT8), amyloid protein, and synuclein (Zimed-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 neurofibrillary tangles were seen in limbic structures. Accentuated neuronal loss and increased neurofibrillary tangle density were noted in the parieto-occipital lobes. The findings satisfied criteria for Alzheimer’s disease by Braak and Braak staging (stage VI/VI)² and by the National Institute on Aging and Reagan Institute working group on diagnostic criteria for the neuropathological assessment of Alzheimer’s disease (high likelihood).² 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 transitional Lewy body disease.³

Comment

The clinical syndrome of posterior cortical atrophy is characterised by prominent dysfunction 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 areas.¹ The predominant features of posterior cortical atrophy are followed by dementia more typical of Alzheimer’s disease.¹ 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.³ Pathologically, the latter is characterised by the presence of Lewy body disease, with limbic or neocortical Lewy bodies.³

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 hallucina-

tions 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.⁴ Although there are reports of prominent visuospatial impairment or Balint’s syndrome in clinically diagnosed and pathologically diagnosed corticobasal degeneration,⁵ visual hallucinations are almost non-existent in corticobasal degeneration.⁶ This suggests that visual hallucinations and parkinsonism in the setting of cognitive impairment reflect underlying Lewy body disease rather than corticobasal degeneration.⁶ 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.

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Reversible callosal 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 alienation and loss of control 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

be sure of its movement. On examination she was alert, fully oriented, and cooperative. Snout and palmomental reflexes were positive. 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 spontaneous 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. Neuropsychological assessment showed fluent speech without dysarthria. Comprehension and reading were intact. Performance in verbal and non-verbal fluency tasks 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 gestures 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 conflict. 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. Furthermore, there was an inability to distinguish 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" sign (or "signe de la main étrangère").¹ Additionally, an inability to cross 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. However, she was correct when asked to point to the location of touch on the face or trunk.

Magnetic resonance imaging showed internal 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 stimulation (TMS) as described previously² and showed a deficient inhibition particularly from left to right (upper panels "left A" in fig 1B). Cerebrospinal fluid (CSF) pressure was normal during lumbar puncture. After removal of 50 cc CSF the alienation of the left arm, the "signe de la main étrangère" and the impaired cross replication of hand postures

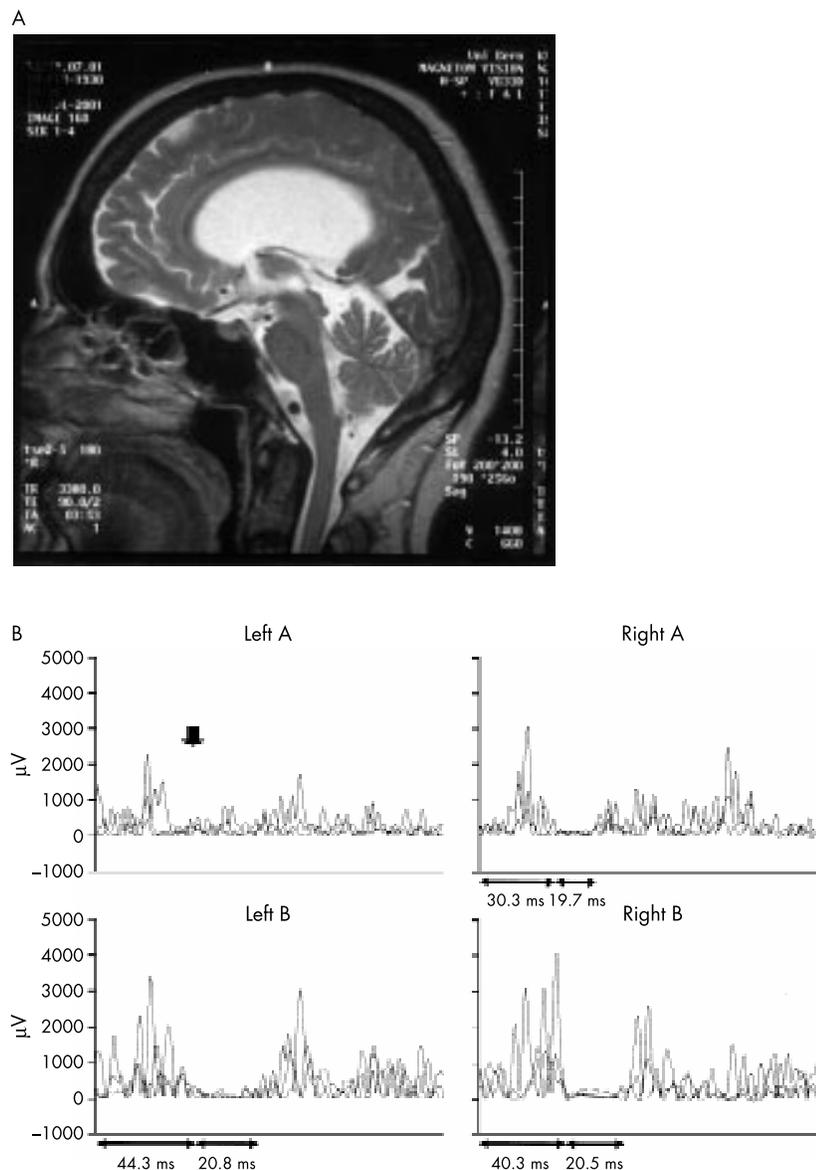


Figure 1 (A) Magnetic resonance image (T2 weighted) showing internal hydrocephalus with thinning of the corpus callosum. (B) Recordings of the rectified tonic electromyographic (EMG) activity of the first dorsal interosseus muscle (ID1) after ipsilateral focal transcranial magnetic stimulation (TMS) 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 A: the findings before lumbar puncture are shown. TI is normal after TMS of the right hemisphere (normal values (mean (SD)): latency 35.8 (7.2); duration 24.8 (5.4)), but missing on the left (arrow). Lower panels B: 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.

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 reappeared. 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,^{3,4} the inability to cross locate touch of the fingers when blindfolded,⁴ and the “*signe de la main étrangère*” (the sign of the foreign hand) describing a failure to recognise self ownership of the left hand when visual cues are removed.¹

We describe a callosal disconnection syndrome with a “*signe de la main étrangère*”, an impaired cross replication of hand postures, and an inability to cross locate touch of the fingers in a patient with internal hydrocephalus that completely disappeared after removing CSF indicating a functional impairment of the corpus callosum. The finding is substantiated by electrophysiological studies: the transcallosal motor inhibition before CSF tapping showed a deficient callosal function that normalised after CSF removal. Before CSF tapping there was also a functional asymmetry in that the transcallosal information transfer from left to right was more deficient than from right to left (fig 1). The origin of this asymmetry is not clear, but may be attributable to an additional involvement of adjacent white matter on one side. Together with the restoration of transcallosal dysfunction after CSF tapping, this asymmetry was largely reversible too.

In summary, the restoration of electrophysiologically assessed transcallosal inhibition in parallel with the recovery of the clinical symptoms and signs of our patient suggest a reversible, partial interhemispheric somatosensory disconnection syndrome attributable to a functional impairment of the corpus callosum.

To our best knowledge this is the first report that underlines that dysfunction of the corpus callosum in internal hydrocephalus may not only cause electrophysiological² but also clinical signs of callosal disconnection.

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Trigeminal autonomic cephalgia-tic-like syndrome associated with a pontine tumour in a one year old girl

The so called trigeminal autonomic cephalgia (TAC) include episodic and chronic paroxysmal hemicrania (CPH), short lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT), and cluster headache (CH).¹ Combinations of cluster headache and chronic paroxysmal hemicrania with trigeminal neuralgia have also been described and have been called cluster-tic syndrome² or CPH-tic syndrome.³ 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 rhinorrhoea on the right side. The attacks usually lasted 12 to 24 hours and occurred weekly, sometimes waking her up at night. During the attacks, the patient wanted to be in a darkened room, but she did not feel nauseous and did not vomit. Initially, neurological examination revealed no abnormalities. Aspirin or other simple analgesics were

not effective. There was no family history of migraine or other severe headaches.

A video recording of several attacks made by her parents showed the child lying still while crying intensely. Her right upper and lower eyelids were swollen and tear production and rhinorrhoea were seen 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 cervical myelum (fig 1A), with a syrinx in the cervical myelum. On the transverse slide, 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 patients' 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

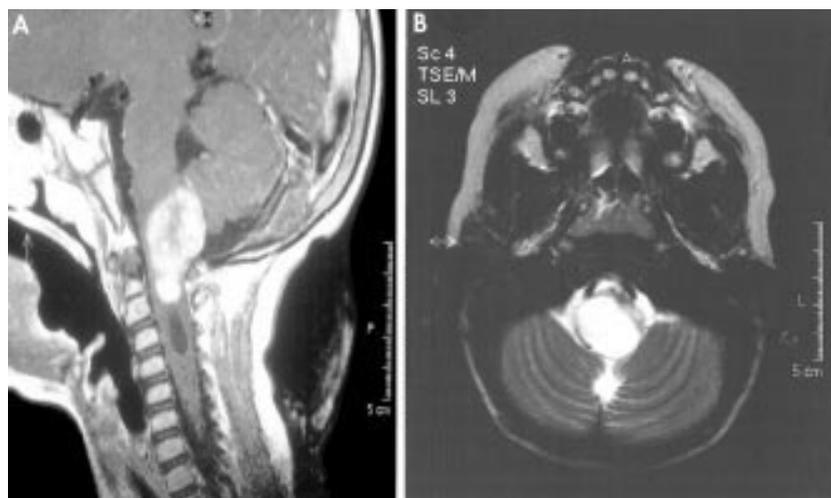


Figure 1 (A) T1 weighted sagittal magnetic resonance image (MRI) scan of the brain after gadolinium contrast, showing a space occupying lesion in the pons, extending to the medulla oblongata. Below the tumour a syrinx is present in the cervical myelum. (B) T2 weighted transverse MRI scan, showing extension of the tumour to the right cerebropontine angle.

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.¹ We 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.^{4,5} A causal relation between the symptoms and the lesions was not easily 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.⁶ Furthermore, cluster-tic syndrome has been reported in association with intracranial pathology.² 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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A video recording of the patient during a headache attack is available on the journal website (www.jnnp.com). The recording shows the patient sitting still with a swollen eyelid on the right side. A few moments later she suddenly starts to cry intensely, while grabbing to her right eye and right ear. During this episode, she moves back and forth, and pronounced lacrymation is seen in her right eye.

Autoimmune myasthenia gravis after cardiac surgery

Autoimmune myasthenia gravis (MG) is a heterogenous 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.¹

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 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.² None had had 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 Scoppetta³), 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.³ 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.⁴

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,⁵ the presence of other autoantibodies or signs of other autoimmune diseases should be sought.

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

Sex, age at the onset of MG	Heart surgery	Time since surgery to onset of MG	Presenting features; (Osserman group at onset)	Clinical diagnosis	AChR antibodies normal value <0.8 nmol	Treatment and response	Follow up and evolution AChR-ab present titre
M, 57	Triple aorto-coronary bypass	3 weeks	Ptosis, double vision; (I)	Response to im neostigmine and oral pyridostigmine	1.9 nmol	Cholinesterase inhibitors: good	6 years, fluctuating ocular MG 3.0 nmol
M, 58	Double aorto-coronary bypass	10 weeks	Dysarthria, dysphagia and arm weakness; (IIb)	Decrement on repetitive stimulation, response to iv edrophonium	8.0 nmol	Cholinesterase inhibitors: mild Prednisone and plasma exchange: good	7 years, fluctuating generalised MG 3.5 nmol
M, 65	Triple aorto-coronary bypass	10 weeks	Double vision, dysphagia and dysarthria; (IIb)	Decrement on repetitive stimulation, response to im neostigmine and oral pyridostigmine	12.0 nmol	Cholinesterase inhibitors: mild prednisone: good	8 months, mild generalised MG 1.1 nmol

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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.¹ Dysphagia occurs in 5–15% of the patients and it may be the only presenting symptom.² Progressive dysphagia caused by Chiari I malformation, mimicking amyotrophic lateral sclerosis, has been reported in this journal in 1996 and 2002.^{2,3} Dysphonia may occur rarely, but it has not been described as an early symptom.² Pain and stiffness in the posterior neck is a common feature, but severe neck extensor weakness 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 increase in the 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 electrophysiological testing was undertaken to investigate the neuromuscular junction. Percutaneous 3 Hz repetitive nerve stimulation of the right accessory nerve along with recordings obtained from surface electrodes over the trapezius muscle did not show significant variations in compound muscle action potential amplitude under baseline conditions, or when the test was repeated three minutes after maximal voluntary effort for 30 seconds. Single fibre electromyography of the right extensor digitorum communis muscle during voluntary activity failed to show any abnormally increased jitter or neuromuscular block, and the mean jitter value was in the normal range. Serum antiacetylcholine receptor antibodies were absent. Standard concentric needle electromyography of proximal and distal muscles in all four limbs was normal. The cervical paraspinal muscles were not investigated. Motor and sensory conduction velocities were normal. A neostigmine test resulted in mild improvement in the dysphonia.

One week later, the patient experienced a worsening of dysphagia and dysphonia and she started to complain of gait disturbances, with instability in walking. Magnetic resonance imaging of the brain and cervical spine showed herniation of the cerebellar tonsils through the foramen magnum, reaching



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 procedure 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. Strangely, 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 of brain stem dysfunction 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.⁴ In our patient, it is possible that slight cervical trauma during anaesthesia for her adenoidectomy may have brought to light the underlying congenital abnormality.

Moorthy *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 instead of or in addition to myasthenia gravis was later found.⁵ 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.⁵ The partial response to anticholinesterase drugs in our case reinforces the view that it is unwise to base the diagnosis of myasthenia gravis purely on a positive pharmacological test.

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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 exceptions to this poor prognosis are cerebral expanding lacunae or, as they are sometimes called, benign intraparenchymal brain cysts.¹ These are intraparenchymal cavities without an epithelial lining, filled with cerebrospinal fluid (CSF), located in the thalamo-mesencephalic arterial territory.^{1,2} Their expanding nature is demonstrated by their progressive clinical course and by the frequent complication of aqueduct stenosis and triventricular hydrocephalus.^{2–4}

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

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 (leg worse than arm), hemirigidity, 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 ponto-mesencephalic 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 incannulated with a 3.9 peel away. 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 neuroglia with few amilaceous 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 very apparent, 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 resumed all her premorbid activities, including dancing.

Comment

A progressive disorder of cognition and hydrocephalus caused by expanding cerebral lacunae in the thalamus and midbrain was first described in 1983.² These lesions consist of multiple grape-like CSF filled cavities, usually located bilaterally in the rostral brain stem. Their incidence is extremely rare (seven cases reported thus far),^{1–5} and apparently not related to any risk factor. Differential diagnosis includes parasitic and neoplastic cystic lesions. However, their pathogenesis remains

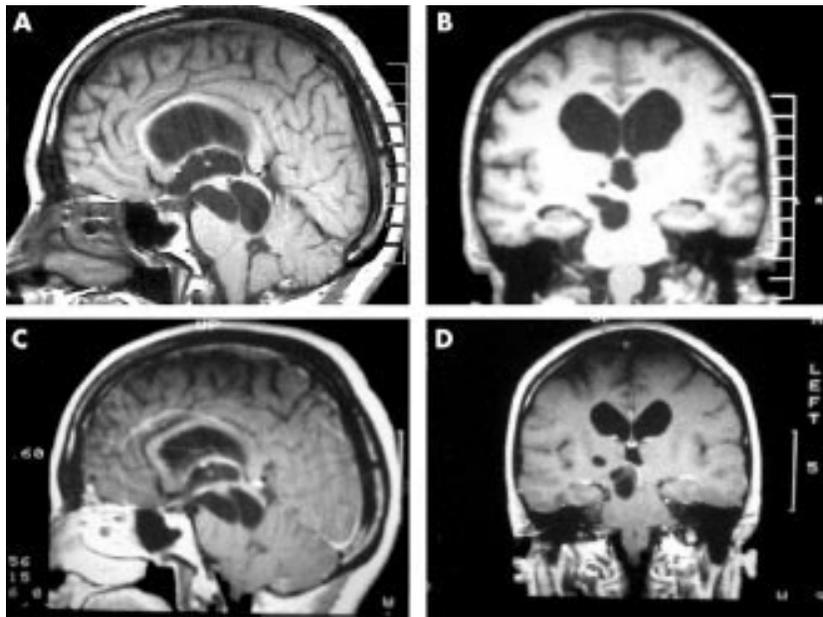


Figure 1 Preoperative sagittal (A) and coronal (B) T1 weighted magnetic resonance (MR) images (gadolinium enhanced). Large multilobulated cystic lesions occupy the right ponto-mesencephalic region, squeezing the aqueduct and causing mild triventricular hydrocephalus. Postoperative sagittal (C) and coronal (D) T1 weighted MR images. There is a slight reduction in volume of the lesions, with partial resolution of the hydrocephalus.

obscure. Clinical presentation is characterised by signs of triventricular hydrocephalus from aqueduct obstruction, and by various extrapyramidal signs, ataxia, and abnormalities of oculomotion. In our patient the tremor disappeared when the motor defects worsened, and reappeared after their resolution. This observation could be explained by the progression of the cystic lesions towards the ventral thalamus causing the functional equivalent of a reversible thalamotomy, the “benefit” of which reversed upon decompression of the cysts.

The expanding nature of the lesions and the progressive clinical worsening justify surgical management. Treatment of the hydrocephalus (shunting, cisternostomy) has seldom been rewarding.^{2–5} Opening and draining the cysts, while carrying a higher morbidity risk, seems to give a better clinical outcome, although the cyst volume is not significantly modified by the procedures.^{1–5} In our case, the use of an endoscopic approach to both the hydrocephalus and the opening of the cysts minimised operative risks and led to an excellent clinical result. The values of endoscopic neurosurgery in expanding cerebral lacunae has been emphasised by others.⁴

While the neuropathology of the lesions and their location supports the interpretation that the cystic spaces are dilated Virchow–Robin spaces, the precise mechanism leading to the dilatation remains unknown. The absence of vasculitis or systemic hypertension in all reported cases reinforces the hypothesis of a localised disturbance in vascular permeability and interstitial fluid reabsorption.⁵

In conclusion we draw attention to this very unusual neuropathological entity. An endoscopic microneurosurgical approach to this

type of lesion has the advantage of a good risk to benefit ratio. As the term “lacuna” is usually associated with a small static vascular lesion, and the term “benign cyst” overlooks the expanding nature of the lesion, we suggest that these lesions should be called “benign expanding cerebral cysts.”

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