- 4 Saxena S, Brody AL, Schwartz JM, *et al*. Neuroimaging and frontal-subcortical circuitry in obsessive-compulsive disorder. *Br J Psychiatry* 1998;35(suppl):26-37.
- 5 Broocks A, Thiel A, Angerstein D, *et al*. Higher prevalence of obsessive-compulsive symptoms in patients with blepharospasm than in patients with hemifacial spasm. *Am J Psychiatry* 1998; 155:555-7.

Cervical spondylotic myelopathy and Kennedy syndrome mimicking amyotrophic lateral sclerosis

A 62 year old male patient presented with a longstanding history of slowly progressive limb weakness, speech, and swallowing difficulties. In 1983 a diagnosis of amyotrophic lateral sclerosis had been made. At that time his physical examination showed tongue atrophy with fibrillations, proximal limb weakness, and brisk lower limb tendon reflexes. Electromyography showed abnormal "spontaneous activity" with fibrillations and positive sharp waves in muscles of all limbs. His further medical and family history was unremarkable.

In January 2000 neurological examination showed mild facial weakness, marked atrophy and fibrillations of the tongue, severe dysarthria and dysphagia, atrophy, and weakness of the shoulder girdle and arm muscles, and an unsteady and broad based gait. Apart from brisk knee jerks, deep tendon reflexes were absent and plantar responses were negative. Sensory testing was normal. General physical examination showed slight gynecomastia. Laboratory testing showed raised creatine kinase (305 U/l) and lactate dehydrogenase (195 U/l) concentrations. Needle EMG demonstrated positive sharp waves and fibrillation potentials and long duration polyphasic motor unit potentials with increased amplitudes in muscles of all limbs. By contrast, motor and sensory nerve conduction studies gave normal results.

As cervical myelopathy is an important differential diagnosis in patients with suspected motor neuron disease, cervical MRI was performed. As shown in figure 1, MRI disclosed marked cervical spondylosis with appreciable narrowing of the spinal canal between C3 and C6. In addition, T2 weighted images showed intramedullary changes with foci of high signal intensity at the level of C5 indicating myelopathy. Although these changes may readily explain the weakness in his upper limbs, the cause of bulbar symptoms and denervation in his lower limbs remained unclear.

The presence of slight bilateral gynecomastia prompted us to look for androgen receptor gene mutations, which cause X linked spinal bulbar muscular atrophy.¹ This disorder, also known as Kennedy syndrome, is caused by an unstable expansion of a CAG repeat in exon 1 of the androgen receptor gene (Xq11-12). The androgen receptor is highly expressed in motor neurons of the brain stem and spinal cord. The CAG repeat expansion is thought to confer a toxic gain of function to the androgen receptor protein resulting in irreversible damage of brain stem and spinal cord motor neurons. In addition, the impaired ability to transactivate androgen sensitive genes of the mutated receptor may account for endocrine features such as gynecomastia or testicular atrophy in spinal bulbar muscular atrophy.²⁻⁴

Genetic analysis in our patient showed one allele carrying an abnormally expanded CAG

Figure 1 Spondylotic ridging at the C3-C6 level causing spinal stenosis is shown in the sagittal T2 weighted MR image. Additionally increased signal intensity is seen within the spinal cord at the level of C5.

repeat (44; normal range 16-33) thus confirming the diagnosis of Kennedy syndrome.

The present patient with coexisting cervical spondylotic myelopathy and Kennedy syndrome highlights the diagnostic value of an intensified investigation including cervical MRI and androgen receptor gene analysis in patients with an unusual clinical presentation of motor neuron disease.

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- 1 La Spada A, Wilson EM, Lubahn DB, *et al*. Androgen receptor gene mutations in X-linked spinal and bulbar muscular atrophy. *Nature* 1991;352:77-9.
- 2 Kennedy WR, Alter M, Sung JH. Progressive proximal spinal and bulbar muscular atrophy of late onset: a sex-linked recessive trait. *Neurology* 1968;18:671-80.
- 3 Sobue G, Hashizume Y, Mukai E, *et al*. X-linked recessive bulbospinal neuronopathy: a clinicopathological study. *Brain* 1989;112:209-32.
- 4 Brooks BP, Fischbeck KH. Spinal and bulbar muscular atrophy: a trinucleotide-repeat expansion neurodegenerative disease. *TINS* 1995;18:459-61.

Posturally evoked vomiting without nystagmus in a patient with Arnold-Chiari malformation

Arnold-Chiari malformation type I (ACM I) is a developmental anomaly of the rhombencephalon characterised by displacement of the cerebellar tonsils into the foramen magnum and elongation of the medulla. It usually presents in adult life with head

motion induced oscillopsia, ataxia, headaches, cervical pain, or Valsalva induced dizziness.¹

Various ocular motor abnormalities have been reported in patients with ACM I. Among these, downbeat nystagmus and periodic alternating nystagmus are the most common. Other often encountered ocular signs, such as gaze evoked nystagmus, rebound nystagmus, and impaired smooth pursuit, reflect cerebellar involvement.^{2,3}

Vertigo of vestibular origin, being peripheral or central, is usually accompanied by nystagmus and nausea, or vomiting, and is often influenced by head position.^{3,4} The entity of central positioning vomiting without, or little, vertigo and nystagmus (posturally evoked vomiting (PEV)) was first reported by Drachman *et al* and later recognised by Brandt and Baloh and Halmagyi.^{3,5}

Posturally evoked vomiting is generally poorly known as a warning symptom of a posterior fossa lesion and is often misinterpreted.⁵ Whereas it has been documented in patients with posterior fossa tumours, it has not been reported in patients with developmental abnormalities. We report on a patient with ACM I where PEV was the most prominent presenting symptom.

This 57 year old woman was seen in our vertigo clinic because of gait unsteadiness and postural vomiting. Her history included an aortic valve replacement for aortic insufficiency, nephrolithiasis, and peptic disease. She was treated with warfarin.

For years, she had dizziness and severe nausea while looking up. During the past months severe nausea and vomiting appeared when she tilted her head to either side and down. Lately, she had become unsteady. She also complained about left high pitched tinnitus and intermittent pain in the left shoulder.

On examination her eyes were properly aligned with a full range of movements. No primary or gaze evoked nystagmus was seen, with and without Frenzel's glasses. The saccadic eye performance was normal, but the smooth pursuit in both the horizontal and the vertical plane was saccadic. The vestibulo-ocular reflex, examined by a doll's eyes movement, head thrust test, and dynamic visual acuity test, was normal. Mild dysmetria on finger-nose testing and finger-finger testing was found bilaterally. The deep tendon reflexes were brisk in the upper and normal in the lower limbs. The plantar toe responses were flexor. Sensation was normal. The gait was atactic and the Romberg test negative.

On testing the eyes in the Dix-Hallpike position to either ear, as well as in the head down position, the patient reported severe nausea, and became pale and perspired. However, no nystagmus was seen either by direct observation, or with Frenzel's glasses.

The symptoms persisted with repetition of the positioning.

An electronystagmogram (ENG) documented saccadic eye tracking in the horizontal plane. The optokinetic nystagmus was asymmetric with little increment after increased speed velocity of the target. When supine and with her head turned to the left, nystagmus of 7°/s, beating to the left, was recorded. No nystagmus was recorded on Hallpike testing. The caloric test was within normal limits.

A behavioural audiogram showed mild bilateral sensorineural hearing loss in the frequencies of 2000 Hz-8000 Hz with normal speech discrimination, interpreted as presbycusis. The brain stem auditory evoked potentials were normal.

Brain T1 and T2 weighted MRI with gadolinium enhancement disclosed low lying cerebellar tonsils with elongation of the medulla and pons, compatible with ACM I. A brain stem or cervical syrinx was not demonstrable (fig 1).

In view of the progressive symptomatology, posterior fossa decompression was considered, but postponed because of the cardiac situation of the patient. Clonazepam was offered to the patient for alleviation of the posturally evoked symptoms, but was refused because of possible sedation.

Central paroxysmal positioning vertigo occurs transiently on changing the head position, as opposed to central positional nystagmus, which persists as long as the head position is sustained.^{3,4} Central paroxysmal positioning vertigo can be differentiated from benign paroxysmal positioning vertigo by its shorter latency, longer duration of the attack, and a direction changing nystagmus which is not attributable to stimulation of a single canal.^{3,4} It usually indicates a lesion around the fourth ventricle, dorsal vermis, or diffuse cerebellar pathology.³ Central positional nystagmus is often bilateral, of low constant frequency, and occurs in association with lower brain stem or vestibulocerebellar lesions.³

In 1977 Drachman *et al* described the entity of PEV.⁵ In their two patients (one with a metastasis and the other with a glioblastoma of the cerebellar vermis) PEV was a prominent clinical sign, whereas vertigo and nystagmus diminished with the progression of the pathological process. Positional vomiting became so severe that the patients had to hold their head in a forced position to prevent vomiting. The symptoms cleared after treatment with vestibular suppressants. Drachman *et al* explained the condition of PEV by dissociation of the vomiting centres from oculomotor and receptive pathways of the complex vestibular syndrome by a brain stem infiltrating lesion.

Arbusow *et al* described a patient with amiodarone induced positional vomiting with mild vertigo, downbeat nystagmus, and limb ataxia, responsive to benzodiazepines. According to their concept, it is in keeping with that of Drachman *et al*, amiodarone disinhibits selectively the connections between the dorsal cerebellar vermis and the vomiting centres in the postremal area and the lateral

reticular formation, sparing relatively the vermis-ocular motor circuitry.⁶

Severe positioning nausea and vomiting with inconsistent dizziness and without nystagmus, was present in this patient with ACM I. Initially, nausea occurred only on head extension, but later on all head movements but anteflexion. An ENG showed only low speed positional—that is, sustained nystagmus in one head direction. On Hallpike positioning testing nystagmus was not recorded. Other ocular motor findings (saccadic pursuit and abnormal optokinetic nystagmus) were compatible with a cerebellar lesion.^{3,4}

In ACM, a normal vestibular input after changes in head position might be misprocessed by transient brain stem compression, leading to dissociation of the vomiting centres from other parts of the vestibular reflex. The relatively mild signs of positional vomiting in our patient might be due to the benign nature of the underlying disease, as opposed to the invasiveness of tumours or acute neurotoxicity, leading to severe PEV.^{5,6}

This patient with ACM seems to be unique in the literature, as PEV has not been reported in this clinical setting.

Further neurophysiological studies are needed to elucidate the pathogenesis of PEV.

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- 1 Stevens JM, Serva WAD, Kendall BE, *et al*. Chiari malformation in adults: relation of morphological aspects to clinical features and operative outcome. *J Neurol Neurosurg Psychiatry* 1993;56:1072-7.
- 2 Bronstein AM, Miller DH, Rudge P, *et al*. Down beating nystagmus: magnetic resonance imaging and neuro-otological findings. *J Neurol Sci* 1987;81:173-84.
- 3 Brandt T. *Vertigo. Its multisensory syndromes*, 2nd ed. London: Springer-Verlag 1999:199-213, 291-8.
- 4 Baloh RW, Halmaghi GM. *Disorders of the vestibular system*. New York: Oxford University Press, 1996:446-53, 488-93.
- 5 Drachman DA, Diamond ER, Hart CW. Posturally evoked vomiting: association with posterior fossa lesions. *Ann Otol Rhinol Laryngol* 1977;86:97-101.
- 6 Arbusow V, Strupp M, Brandt T. Amiodarone induced severe prolonged head positional vertigo and vomiting. *Neurology* 1998;51:917.

Lunchtime headache

Chronic primary unilateral headaches fall into one of five categories: chronic cluster headache, chronic paroxysmal hemicrania, hemicrania continua, cervicogenic headache, and SUNCT syndrome. Overlap between types is recognised. Although the differentiation of these is sometimes difficult, there are important therapeutic implications—for example, indomethacin has a dramatic effect on chronic paroxysmal hemicrania but is less effective in chronic cluster headache. Here, a patient with paroxysmal unilateral headaches, occurring precisely on the same day of the week and at the same time, is described.

A 57 year old man presented with episodic, right sided, moderate to severe headaches of 9 months' duration. He developed these always on a Monday at 1300 hours. The

headaches were sharp and throbbing with the maximum pain behind the right eye. The headache then radiated to the back of the head without crossing the midline. The pain was felt in paroxysms each lasting several minutes. He did not experience any visual symptoms, nausea, nasal congestion, lacrimation, or ptosis. Initially the total duration of the headaches was about 10 hours but it became progressively longer, remaining until Wednesday on some occasions. They were not related to the consumption of food and he did not go out drinking on Sunday nights.

At the age of 20, he had had four episodes of severe headache on the right on four consecutive days after breaking rest during a week of night shifts as a warehouseman. In between the Monday "lunchtime headaches", he remained well. He had no personal or family history of migraine. He was taking propranolol for mild hypertension. His examination was normal. There was mild cervical spondylosis on plain radiology. Brain CT showed minor atrophy.

A diagnosis of chronic paroxysmal hemicrania was made, and he was prescribed indomethacin (50 mg three times on Mondays). With this his headaches were delayed until Tuesday morning and the duration of the headaches was reduced to less than 24 hours. The headaches also became less predictable. A more protracted course of indomethacin rendered him headache free.

Review of this man's headache history suggests that the episode he had when aged 20 was probably an isolated "cluster" headache. The more recent, highly predictable, right sided headaches on Monday afternoons were unusual. The predictability of these was such that he could time it precisely to 1300 hours. Their characteristics were not classic for chronic paroxysmal hemicrania, in which attacks last between 20 and 30 minutes and are accompanied by ipsilateral nasal congestion and lacrimation.¹ Attacks of chronic paroxysmal hemicrania can, however, have clockwork regularity and are abolished by indomethacin.¹ Headaches in this case are also different from chronic cluster headache as the paroxysms occurred many times throughout the day (repetition rate 12-16/day) and the total duration of an "attack" could be up to 72 hours. Recently another variant of chronic paroxysmal hemicrania—hemicrania continua—has been described² and these headaches are non-paroxysmal and continuous. The degree of cervical spondylosis in this patient was mild, and furthermore the nature of the attacks was not that of cervicogenic headache. SUNCT syndrome is a type of paroxysmal unilateral headache consisting of short lasting, unilateral, neuralgiform headaches with conjunctival injection and tearing.^{3,4} Patients have up to 30 paroxysms/hour, occurring once or twice a day.³ This diagnosis is also highly unlikely in our patient, with his clinical presentation and therapeutic response. It was thought that this man's headaches were most probably due to chronic paroxysmal hemicrania evolving from a possible early "cluster" attack, supporting the suggestion that a common pathophysiology underlies these "trigeminal-autonomic cephalalgias".⁴ The precise predictability of the Monday afternoon headaches is fascinating, and unexplained.

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Figure 1 Brain MRI shows low lying cerebellar tonsils and elongation of the pons and medulla, compatible with ACM I.