autosomal dominant late onset cerebellar ataxia with myoclonus, peripheral neuropathy and sensorineural deafness. Necropsy in one patient showed loss of cells in the dentate nuclei, a reduced amount of cerebellar white matter, and pallor of the gracile tracts in the spinal cord. We report another case with this syndrome.

A 56 year old male was referred to us with an intention tremor involving both arms. In his twenties he noticed bilateral hand tremor and decreased hearing in both ears. In his thirties he had bilateral deafness, increasingataxia and tremor in both arms and an increasing gait imbalance. During the past three years he experienced jerking of his arm when engaged in purposeful movements and also progressive thinning of his legs and hands. One of his two daughters had pes cavus and one (maternal) uncle was deaf. His mother had pes cavus and tremor and his maternal grandmother had tremor and deafness. Positive findings on examination included bilateral deafness, mild dysthria, marked ataxia, a slight postural and antverted intention tremor and intention myoclonus in his arms. He had decreased reflexes in the upper limbs, abolished reflexes in the lower limbs, wasting of the interossei, thenar and hypothenar eminences and distal legs with pes cavus, slight distal loss of vibration sense in the lower limbs and a mild gait ataxia. Mental status, optic fundi and ocular movements were normal, and there was no nystagmus or pyramidal signs.

Normal laboratory values included cholesterol, triglycerides, ceruloplasmin, lactate, pyruvate, lead (urine and serum), δ-aminolevulinic acid and hexosaminidase A. Audiometry confirmed bilateral severe hearing loss of more than 100db for both high and low tones (from 250 to 8000 Hz). EMG showed signs of chronic partial denervation and reinnervation, both on the upper and lower limbs, with reduction in the number of motor units and polyphasic, large, high-amplitude motor unit potentials. Motor nerve conduction velocity was reduced in ulnar (31.7 m/s) and peroneal nerves 15.4 m/s). Sensory nerve conduction velocity was slowed on the ulnar (27.3 m/s). No sensory potential was obtained from stimulation of the sural nerve at the ankle. Visual evoked potentials were normal. Somatosensory evoked potentials showed delayed lumbar responses on tibial stimulation and delayed Erb's point potentials after stimulation of the median. Latencies between next potentials were normal. No high-amplitude cortical waves were recorded. The EEG showed a few paroxysms of spikes and sharp waves on both temporal leads. Simultaneous EMG recording demonstrated intention myoclonus, without any temporal relationships to the EEG paroxysms. CT showed mild atrophy of the cerebellar vermis. Skin biopsy was normal, with no PAS-positive inclusions on cells of eccrine sweat ducts. Muscle biopsy (deltoit) showed that the muscle was composed of type I fibres only. No ragged-red fibres were seen.

The following drugs were used to try to control tremor and myoclonus: bcaprin (15 mg/daily), piracetan (4800mg/daily), baclofen (45 mg/daily), propranolol (60 mg/daily) and isoniazid (900 mg/daily) had no effect. Lisuride 0.1 mg (IV) produced no improvement but caused nausea, vomiting and drowsiness. Valproate (1500 mg/daily) slightly improved the myoclonus and had minimal benefit on the tremor. Clonazepam (up to 10 mg) clearly reduced myoclonus, but produced intolerable sedation.

Classification of ADCA of late onset is controversial. Harding grouped the ADCA on four types, based on clinical features only. Our patient fits the characteristics of ADCA type IV (with myoclonus and deafness). Furthermore he also had tremor, myathrophy, absent tendon reflexes in the lower limbs and electroneurovascular evidence of a sensori-motor polynuropathy. ADCA type IV is rare. Only two families have to our knowledge, been described. The family described by May and White had cerebellar ataxia, myoclonus and deafness, but no muscular atrophy or evidence of polyneuropathy. Like our patient, the family observed by Baraitser presented with polyneuropathy and absent tendon reflexes in the lower limbs, but none of them had marked myathrophy, a clinical feature not previously reported in ADCA type IV. As usually seen in other types of ADCA in this family expressivity was variable, both on the severity and diversity of the clinical picture, ataxia, deafness and amyotrophy being either an isolated finding or combined with each other. Late onset ataxic disorders with additional features such as dementia, myoclonus, deafness or peripheral neuropathy have been described in association with defects of the mitochondrial respiratory chain. Andersmann et al. suggested that the hearing loss in the May-White syndrome indicated that this disorder is also due to mitochondrial dysfunction. Previous cases have not had muscle histology with the modified Gomori Trichrome method, or reveal ragged-red fibres, the morphological hallmark of mitochondrial myopathy.

Mitochondrial diseases can be transmitted by either mendelian or maternal inheritance, and usually show a wide variability in the phenotypes of affected individuals, as in this family. However the pedigree is incompatible with maternal inheritance, as the deafness of the proband was also affected. Furthermore our patient had no ragged red fibres on muscle biopsy and his serum pyruvate and lactate levels were normal.

TERESA P MELO,
JOSÉ M FERRO
Department of Neurology.
Hospital de Santa Maria.
Lisbon, Portugal.

We thank Drs M Alves, R Serrão, F Morgado and Dr Baraitser for their help with this study.

References


Persistent primitive trigeminal artery: a possible cause of trigeminal and abducens nerve palsy

Sir: The persistence of a primitive trigeminal artery (PTA) is usually found coincidentally on angiography. Several authors have documented its possible association with
other vascular anomalies such as aneurysms, arteriovenous malformation. It appears, however, that the incidence of cranial nerve dysfunction induced by PTA is quite rare.

A 62 year old woman noticed the onset of diplopia when she woke up, which was more marked on looking toward the left and associated with left orbital dull pain. The next day she developed a headache in her left temporo-occipital region. The pain gradually improved within one month, but double vision on left lateral gaze persisted. There were no bruits on auscultation of the head, orbits, or carotids. Neurologically, mild hypesthesia in the first division of the left trigeminal nerve and incomplete left abducens nerve palsy were noted. Other cranial nerves were intact and there were no pulsatile exophthalmos or chemosis of the bulbar conjunctiva suggestive of a carotid-cavernous fistula.

Angiography showed that the anastomotic artery arose from the superolateral wall of the left internal carotid artery at the C4-5 junction with an infundibular-like aneurysmal dilatation at its origin, coursed postomedially, and then joined the rostral basilar artery just below the origin of the superior cerebellar artery, filling the BA and the right SCA partially (fig). This artery was opacified on vertebral angiography only in the early arterial phase. The calibre of the BA was small and the left posterior cerebral artery was opacified through the fetal type posterior communicating artery. Using a thin section with high resolution CT cisternography, which was performed to rule out the presence of any tumour as another causative lesion, also disclosed this artery in the posterior fossa just medial to the left trigeminal nerve. The abducens nerve palsy improved gradually with conservative therapy and she was neurologically free eight weeks later. Anatomically and radiologically, we assumed that the aneurysmal dilatation at the origin of the PTA induced the abducens nerve palsy and the hypesthesia in the first division of the trigeminal nerve.

Anatomical studies suggest that trigeminal neuralgia and abducens nerve palsy might result from anomalous vessels. Among 17 PTAs which were considered as a responsible lesion for several cranial nerve dysfunctions, three cases had unruptured aneurysms or aneurysmal dilatation at the origin of PTA. Sugar suggested that the origin of the trigeminal artery, where the PTA normally become obliterated in the embryo, could be a weakened point and would be predisposed to rupture. It is, therefore, possible to consider that unruptured aneurysm or aneurysmal dilatation, or even the influence of paroxysmal change in the blood flow on the PTA, could exert pressure on cranial nerves resulting in transitory or chronic dysfunction of cranial nerves.

Persistent primitive arteries must be kept in mind as a causative agent of certain cranial nerve dysfunctions. Spontaneous recovery of cranial nerve disorder due to the vascular compression has occasionally been encountered. In such cases, cranial nerve dysfunction could be caused by the transient increase of the blood flow or the dilatation of the vessels, but the abducens nerve could tolerate the mechanical compression and recover in several weeks. There have been several reports of surgery for aneurysms of the PTA by opening the cavernous sinus or by ligation of the carotid artery. However, there was a risk of injury to the adjacent intact cranial nerves or, especially, of sacrificing blood supply not only to anterior but also to posterior circulation.

Kiyonobu Ikezaki
Kiyotaka Fujii
Takashi KishiKawa
Department of Neurosurgery and Radiology, Saga Medical School, Nishinomia, Saga, Japan

Correspondence to: Dr Kiyonobu Ikezaki, Division of Neurosurgery, UCLA School of Medicine, Los Angeles, California 90024, USA.

References

Norpethidine induced myoclonus in a patient with renal failure

Sir: Pethidine (meperidine) is biotransformed by the liver to norpethidine. Accumulation of norpethidine in uraemia may produce an encephalopathy similar to uraemic encephalopathy. We report a case of norpethidine induced generalised myoclonic status in a hemodialysis patient, resulting in severe hyperkalaemia, and successfully treated with clonazepam.

A 33 year old man with a six year history of renal failure secondary to mesangio-proliferative glomerulonephritis was admitted to the Royal Perth Hospital in August 1988. The uraemia was managed with three weekly home haemodialysis. Renal colic that occurred during dialysis, approximately 48 hours before admission, caused him to stop dialysing prematurely. Pethidine, 100 mg, was administered at a regional hospital and a further 200 mg was administered during the following eight hours. Morphine and prochlorperazine were used thereafter, with total doses of 60 mg and 62.5 mg respectively. Approximately 36 hours after the last dose of pethidine, he showed increasing confusion, agitation and myoclonic jerks. He was then transferred to the Royal Perth Hospital.

Clinical examination revealed a sallow, disorientated man who obeyed verbal commands inconsistently. His temperature was 36.7°C, pulse was regular at 100/minute and blood pressure 230/120. There were continuous, spontaneous, arrhythmic, asyn-
Persistent primitive trigeminal artery: a possible cause of trigeminal and abducens nerve palsy.
K Ikezaki, K Fujii and T Kishikawa

*J Neurol Neurosurg Psychiatry* 1989 52: 1449-1450
doi: 10.1136/jnnp.52.12.1449

Updated information and services can be found at:
http://jnnp.bmj.com/content/52/12/1449.citation

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/