PostScript

LETTERS

Internal carotid artery vasospasm syndrome: demonstration by neuroimaging

Recurring episodes of temporary hemiparesis in young adults are often regarded as ischaemic or migrainous attacks, although non-vascular pathogenesis such as multiple sclerosis should not be ruled out. Vasospasm is known to accompany migraine, but spontaneous vasospasm as a cause of temporary hemiparesis has been rarely demonstrated

except in those cases induced by subarachnoid haemorrhage and those induced by mechanical manipulations.

We report a case of repeated internal carotid artery (ICA) vasospasm. Magnetic resonance angiography (MRA), colour coded duplex sonography (CCDS), and [99mTc]-hexamethylpropyleneamine oxime single photon emission computed tomography (HMPAO-SPECT) together were successful in demonstrating the ICA vasospasm and resulting cerebral ischaemia.

Case report

A 35 year old right handed man presented at our department for repeated transient one

sided weakness. His parents were second cousins. His father died of stroke at the age of 46 years and his mother dies of a severe asthma attack aged 52 years. His 33 year old sister did not have a history of any neurological disorders.

He first experienced right hemiparesis and contralateral visual blurring at 20 years of age. This was not accompanied by headache. These symptoms continued for seven days and then resolved spontaneously. Since then he had had similar episodes almost monthly. The affected side had varied with each attack. Weakness was worse on standing but without significant orthostatic hypotension. He recovered completely within three to seven

Figure 1 (A) Magnetic resonance imaging (MRI) on February 2003 during remission. The arrow indicates the small infarct that occurred after administration of ergotamine at the age 27 years. (B) Internal carotid artery (ICA) image disappeared when he suffered from right hemiparesis during an attack in August 2003. (C) ICA image reappeared in follow up MRA done in September 2003, when the symptoms had ameliorated. (D) Intensity of vascular images of the right ICA was decreased, indicating narrowing of the ICA on October 17. HMPAO-SPECT was carried out between attacks (E) and immediately after MRA (F). Resulting right brain hypoperfusion is shown. (G) Immediate colour coded duplex sonography during an attack on October 23 showing severe stenosis of the left ICA. (H) One hour after left stellate ganglion block, left ICA stenosis was reversed. HMPAO-SPECT, [99mTc]-hexamethylpropyleneamine oxime single photon emission computed tomography.

days. On one occasion after ingesting Cafegot, a preparation of ergotamine with caffeine, when aged 27, he experienced a myocardial infarct. Although we did not undertake a neurological examination at that time, magnetic resonance imaging (MRI) showed a small new lesion in the white matter of the left frontal lobe. Calcium channel antagonists, β adrenergic blockers, antidepressants, anticonvulsants, and aspirin had been used to prevent attacks. However, these drugs had a marginal effect, if any.

In August 2002, he first presented to us between attacks. Neurological examination revealed right handed clumsiness with a somewhat increased tone of the right arm. Pes cavus and equinovarus were also noticed. Electrocardiography supported the history of a previous myocardial infarct. Laboratory studies—including coagulation profile, fibrinogen level, protein C and S levels, screening for collagen diseases, anticardiolipin antibody, and homocysteine—did not show any cardiovascular risk factors. Cerebrospinal fluid was normal (0/3 cells/mm³, protein 27 mg/dL, glucose 65 mg/dL, IgG index 0.52). Genetic screening was carried out after obtaining informed consent, and was negative for mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS), myoclonic epilepsy associated with ragged-red fibres (MERRF), voltage sensitive P/Q-type calcium channel mutation, and NOTCH3 mutation. Pattern reversal visual evoked potentials were normal. Our first MRI examination (28 February 2003) revealed only the above mentioned old lesion in the left brain (fig 1A) with normal MRA findings (data not shown). Disorders of the peripheral nervous system were not detected clinically or electrophysiologically. Disorders of language, agnosia, or apraxia were never noticed. Drugs selected from propranolol, arotinolol, nifedipine, diltiazem, isosorbide, nicorandil, nisoldipine, diazepam, phenytoin, valproate, and aspirin were used for prevention. Each was taken for at least three months. However, no clear beneficial effect was observed.

Repeated MRI did not yield any findings until 22 August 2003, when left eye pain and decreased vision started and developed into a right hemiparesis. Emergent MRA showed disappearance of the left ICA, indicating absence or little flow in the ICA (fig 1B). However, no new lesion was found in the brain parenchyma (data not shown). The symptoms resolved six days later. The left ICA was of normal appearance on follow up MRA in September 2003 (fig 1C). Narrowing of the right ICA was also confirmed as decreased intensity of vascular image on 17 October 2003, on the first day of the attack. Stenosis appeared to start a little distal to the carotid bifurcation in the petrous segment (fig 1, panels D and E). Subsequent SPECT revealed decreased blood supply in the right brain (fig 1, panels F and G). The symptoms disappeared by 23 October 2003.

We concluded that the transient hemiparesis resulted from ICA vasospasm. To reverse ICA vasoconstriction at an early stage, we undertook a stellate ganglion block by the standard anterior paratracheal approach at the C6 transverse process, and he immediately recovered. Vasodilatation was successfully demonstrated by CCDS (fig 1, panels G and H). Although a single application of stellate ganglion block was able to ameliorate the vasospasm, further blocks to the affected side were carried out on three consecutive

days to thoroughly suppress sympathetic activity. Sadly, a preventive effect on ICA vasospasm was not observed and the frequency of the attacks was unchanged.

Comment

We describe a patient with repeated ICA vasospasm of unknown origin. Because of the consanguinity of his parents and the Friedreich's foot seen in various genetic neurological disorders,¹ genetic screening was undertaken, but a gene disorder was not identified.

We initially diagnosed him as having hemiplegic migraine because migraine manifesting aura alone is well documented, and narrowing of the large or small arteries during a migrainous attack has been reported.² However, vasodilatation or hyperperfusion seen after the aura in migrainous attacks³ was not observed even when MRA, CCDS, and SPECT were done on several occasions from the time of onset to just after the attacks ended. Worsening on standing indicated that insufficient blood supply rather than cortical spreading depression was the primary cause of his hemiparesis. The affected side was not fixed, in contrast to familial hemiplegic migraine caused by a genetic disorder of the voltage sensitive P/Q-type calcium channel.³ As multiple sclerosis can produce a transient neurological deficit, it should be ruled out. However, normal cerebrospinal fluid and visual evoked potential findings and lack of demyelination on MRI did not support multiple sclerosis as a diagnosis.

We used stellate ganglion block to reverse ICA vasoconstriction by blocking sympathetic activity.⁴ The favourable effect observed confirmed that the neurological deficit resulted from ICA spasm. Hypersensitivity of the ICAs to neuropeptide Y and noradrenaline may be related to vasospasm.

Spontaneous vasospasm as a cause of ischaemic attacks should be carefully considered using neuroimaging techniques such as MRA and CCDS. As successful prevention of ICA vasospasm by treatment with a calcium channel antagonist has been reported,⁵ and as stellate ganglion block was able to ameliorate the vasoconstriction in our case, evidence of the presence of vasospasm may change the treatment and management of patients with ischaemic attacks.

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References

- 1 Patten JP, editor. *Neurological differential diagnosis*, 2nd edition. London: Springer-Verlag, 1996.
- 2 Boussier MG, Welch KM. Relation between migraine and stroke. *Lancet Neurol* 2005;4:533–42.
- 3 Welch KM. Contemporary concepts of migraine pathogenesis. *Neurology* 2003;61:S2–8.
- 4 Nitahara K, Dan K. Blood flow velocity changes in carotid and vertebral arteries with stellate ganglion block: measurement by magnetic resonance imaging using a direct bolus tracking method. *Reg Anesth Pain Med* 1998;23:600–4.
- 5 Proust F, Callonac F, Clavier E, et al. Usefulness of transcranial color-coded sonography in the diagnosis of cerebral vasospasm. *Stroke* 1999;30:1091–8.

Adult onset leucoencephalopathy with brain stem and spinal cord involvement and normal lactate

Leucoencephalopathy with brain stem and spinal cord involvement and raised lactate (LBSL) is a white matter disease that has recently been described based on typical magnetic resonance imaging (MRI) findings.^{1,2} These include widespread, homogeneous or inhomogeneous white matter abnormalities, corpus callosum involvement, and selective involvement of certain brain stem and spinal cord tracts. Increased white matter lactate levels are usually found on magnetic resonance spectroscopy (MRS). Clinically, the condition is characterised by a childhood onset slowly progressive course. Here we report two patients who presented with adult onset LBSL and normal lactate on MRS.

Case report

A 23 year old woman and her 25 year old brother were referred to our department because of a slowly progressive gait disorder. At the age of 20 (in the female patient) and 23 years (in the male patient), the siblings had first noted an unsteady gait, stiffness in both legs, and bilateral clumsiness. They had both had normal development, and their former medical history was unremarkable. They had achieved unsupported walking at ages of 14 months (female patient) and 15 months (male patient). Both had been examined during early adolescence, with normal neurological findings. They were the only affected members in their family, and their parents were non-consanguineous.

Neurological examination on admission revealed mild spastic tetraparesis (affecting the legs more than the arms), generalised ataxia, saccadic smooth pursuit, saccadic dysmetria, and horizontal, gaze evoked nystagmus. Both patients had hyperreflexia, and plantar reflexes were extensor. Sensory testing was remarkable for a distal symmetrical decrease in position and vibration sense. Neuropsychological testing (Wechsler Memory Scale Revised, Rey-Osterrieth Complex Figure Test, logical reasoning, dual task paradigm, Multiple Choice Vocabulary Test B) revealed that they were of average intelligence and did not show signs of cognitive decline. The following laboratory investigations