Vertebralbasilar dolichoectasia and exertional headache

Vertebralbasilar dolichoectasia is a rare anomaly of the intracranial arteries, consisting of elongation, widening, and tortuosity of the vertebral and basilar arteries. The neurological symptoms and signs are highly variable.1 Headache associated with exertion is uncommon but well known to clinicians. Usually it is benign and only rarely associated with intracranial disease.2 We report a 29 year old patient with occipital headache, associated with exertion and vertebralbasilar dolichoectasia shown by magnetic resonance angiography (MRA).

A previously healthy 29 year old man came to our clinic because of severe throbbing occipital headache for three years. The pain always occurred a few minutes after exertion such as lifting weights or during jumping or jogging, and once after sexual intercourse. It commonly lasted for several days and subsided gradually without medication. He engaged in sports and the headache did not recur in this period. His mother and father both had a history of headache.

Physical examination showed an asthenic Marfan-like habitus. His height was 201 cm. His blood pressure was 120/80. Neurological findings were normal. Flow velocities assessed by ultrasound in the V1, V2, and V3 segments of the vertebral arteries were normal, and the V4 segments and basilar artery were not found. Magnetic resonance imaging and MRA showed mild ectasia and severe elongation of both vertebral arteries and the basilar artery to the left, almost as far as the left cerebellopontine angle (figure). Transversal axial coded duplex ultrasonography did not visualise the dolichoectatic arteries, probably due to a maximal occlusion angle. The patient was advised to take propranolol (20 mg twice daily) and to resume physical exercise gradually. Six weeks later, still taking propranolol, he was able to perform physical exercises without headache.

Vertebralbasilar dolichoectasia is a rare elongation and distension of the vertebral and basilar arteries. A prospective MRI study showed dolichoectatic intracranial arteries in 0.91% of all examinations, and only two out of three patients had symptoms or signs.3 Vertebralbasilar dolichoectasia is diagnosed when the basilar artery lies lateral to the margin of the clivus or dorsum sellae or above the level of the suprasellar cistern (elongation) and when its diameter exceeds 4.5 mm (ectasia).4 Magnetic resonance imaging and MRA in our patient showed that the basilar artery was elongated as far as the cerebellopontine angle. According to Smoker et al this is the most severe elongation of the basilar artery.4 Symptoms and signs associated with vertebralbasilar dolichoectasia are cranial nerve deficits due to compression, long tract signs, cerebellar signs, and hydrocephalus.5 To our knowledge, vertebralbasilar dolichoectasia associated with exertional headache has never been reported.

Exertional headache has been known for more than two millenia, and is mostly benign and not associated with intracranial pathology. It is characterised as always occurring with exertion, prompt in onset, and brief in duration.2 Early studies of patients with exertional headache did not find any cerebral or cranial abnormality, and only later were lesions and abnormalities such as Arnold-Chiari deformity, patybasia, basilar impressions, subdural haematomas, intracranial cysts, or brain tumours reported.6 There were no patients with dolichoectasia. In our patient the elongation of both vertebral arteries and the basilar artery may have caused the occipital headache, which always occurred during sports and lasted for several days. During physical exercise there is an increase in systemic blood pressure. This may lengthen the already elongated vessels, cause stretching of pain sensitive fibres around blood vessels, or even cause minute and painful tears of the vessel walls.

The headache was successfully treated with propranolol, which has a negative inotropic and chronotropic effect on the heart and reduces blood pressure overall and the rate of change of blood pressure with time. Propranolol has been shown to slow progression of aortic dilatation in Marfan’s syndrome and is also beneficial in abdominal aortic aneurysms.7 This might explain the beneficial effect on vertebralbasilar dolichoectasia, the pathogenesis of which is probably related to aortic aneurysms and which has been reported to occur in conjunction with abdominal aortic aneurysms.

We thank Dr B Porcellini, Bern, for performing the MRA.

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Cholinergic supersensitivity of the iris in Alzheimer’s disease

As well as the damage to basal forebrain cholinergic neurons, there is evidence in Alzheimer’s disease for degeneration of ganglion cells of the retina, a decreased parasympathetic cardiac response to changes in posture,2 and reduction of acetylcholinesterase in adrenal glands.7 This suggests that in this disease the brain is not the only site of abnormalities. Thus we explored the possibility of cholinergic dysfunction in the iris. We studied 21 women and five men with probable Alzheimer’s disease (mean age 72.3, range 56 to 83 years).

Mini mental state examination (MMS), C1, EEG, and CSF and blood tests were performed. Reasons for exclusion were Parkinsonism, stroke, depression, diabetes, renal disease, cancer, alcoholism, syphilis, and ophthalmological pathology. Twenty three healthy subjects (mean age 72.8, range 60 to 83 years) were also studied.

Dilute pilocarpine (0.0625%) was used to assess cholinergic supersensitivity. Ambulatory light was kept constant. Two drops of pilocarpine were instilled in one conjunctival sac and the diameter of both pupils was measured before and 20 minutes after instillation. In 10 control subjects pupillary diameter was obtained by holding a transparent ruler against the bridge of the nose and measuring to the nearest 0.5 mm; the results were compared with photographs of the pupil taken with a polarising camera, and there were no discrepancies between the two methods. In the remaining cases the ruler alone was used. The untreated eye served as a control. Also, the systolic blood pressure response to standing up was recorded at one minute intervals for three minutes. The Wilcoxon signed rank test was used for statistical analysis of the MMS, and Fischer’s exact test for the pupil data.

The mean (SD) MMS score was 11.1 (1) for patients with Alzheimer’s disease and 28.1 (4) for controls (p < 0.0001). Basal pupillary diameter was assessed in every case by two independent observers and there were no discrepancies between the

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*J Neurol Neurosurg Psychiatry* 1994 57: 1544
doi: 10.1136/jnnp.57.12.1544

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