Vertebrobasilar dolichoectasia and exertional headache

Vertebrobasilar 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 verteobasilar 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 stopped 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). Transfemoral colour coded duplex ultrasonography did not visualize the dolichoectatic arteries, probably due to a 50° orientation 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 exercise without headache.

Vertebobasilar 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.1 Vertebobasilar 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 mm (ectasia).2 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.3 Symptoms and signs associated with verteobasilar dolichoectasia are cranial nerve deficits due to compression, long tract signs, cerebellar signs, and hydrocephalus.4 To our knowledge, vertebobasilar dolichoectasia in combination with exertional headache has never been reported.

Magnetic resonance angiography: (A) coronal; (B) axial view. The vertebral and basilar arteries are elongated to the left. The junction of the vertebral arteries to the basilar artery is marked with a star.

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, platybasia, basilar impression, normal haematomyelia, intracranial cysts, or brain tumours reported.3 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.5 This might explain the beneficial effect on verteobasilar dolichoectasia, the pathogenesis of which is probably related to aortic aneurysms and which has been reported to occur in conjunction with abdominal aortic aneurysms.

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Correspondence to: Dr H P Mattle, Department of Neurology, University of Bern, Inselhospital, 3010 Bern, Switzerland.


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 cholinergic response to changes in posture,2 and reduction of acetylcholinesterase in adrenal glands.3 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), CT, 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. Ambient 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 polaroid camera, and there were no differences 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 28.1-1 (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
Rheumatic chorea and lupus anticoagulant

Two patients were diagnosed as Sydenham’s chorea out of 322 patients seen in our movement disorder clinic in 1991–3. Both had lupus anticoagulant.

Patient 1 was a 12-year-old girl who had had three attacks of generalised chorea within two years. The first attack occurred a month after a pharyngitis episode; it was the most severe and the longest, lasting five months. Because the choreoathetotic hypokinetic movements were severe and only slightly improved by sodium valproate (1500 mg daily) and haloperidol (3 mg daily), corticosteroid treatment was required for two months (prednisone, initial dosage 40 mg daily). She had a history of asthma, which was treated with cromoglicate, and frequent episodes of pharyngitis. There was no other relevant personal or family history. Her physical examination was normal. Laboratory investigations showed an antistreptolysin-O titre between 300 and 621 Todd units (normal <250). Several different techniques revealed partial thromboplastin time during the first attack of chorea showed prolonged values (57 sec; control 32.9 sec), not corrected by the addition of normal plasma. In the two subsequent relapses of chorea only a slight prolongation of the activated partial thromboplastin time was detected. Anticardiolipin IgG antibodies ranged between 6.9 and 19 U GPL/ml (normal, <15). Anticardiolipin IgM antibody concentrations were normal. Brain MRI showed a slight asymmetry of the frontal horns of the lateral ventricles, but there was neither change of signal nor gadolinium enhancement. Since February 1993, she has remained asymptomatic, just receiving prophylactic treatment with intra-muscular penicillin.

Patient 2 was a 12 year old girl who presented with mild generalised chorea in July 1992. No other neurological or systemic abnormalities were seen. One year earlier she had had a pharyngitis episode followed by fever and migratory polyarthritis. There was a family history of rheumatic fever without chorea. Laboratory investigations showed antistreptolysin-O titres between 300 and 460 Todd units, and prolonged activated partial thromboplastin time (39 sec; control 30 sec) not corrected by the addition of normal plasma. Anticardiolipin antibody concentrations were normal. Brain MRI was normal. The patient was treated with sodium valproate (1000 mg daily) and propranolol intramuscularly. After a few episodes, the choreic movements disappeared within the next few months. Although an increased titre of antistreptolysin-O antibody is still present, subsequent coagulation studies have been normal.

Extensive investigations showed no other abnormalities in either case. Thus two patients with rheumatic chorea and prolonged activated partial thromboplastin time are reported. Although more specific coagulation tests’ were not done, lupus anticoagulant is the most probable explanation for the increased activated partial thromboplastin time detected in both cases. In patient 1, a high titre of anticardiolipin IgG antibodies was also detected.

Rheumatic fever and lupus anticoagulant are two known causes of immunologically mediated chorea, and lupus anticoagulant seems to represent the main biochemical abnormality responsible for chorea in systemic lupus erythematosus. Although lupus anticoagulant has been associated with chorea gravidarum, and anticardiolipin antibodies with rheumatic fever, to our knowledge, lupus anticoagulant had not been reported in patients with rheumatic chorea.

Although little is known about the role of antiphospholipid antibodies, anticardiolipin antibodies seem to be unrelated to the rheumatic chorea pathogenesis. Different protein confirmations responsible for lupus anticoagulant and anticardiolipin antibodies have however, been isolated. Therefore, the antineuronal antibodies responsible for rheumatic chorea could be more related to lupus anticoagulant than to anticardiolipin antibodies.

RAÚL DE LA FUENTE FERNÁNDEZ
Service of Neurology
Hospital Juan Canalejo,
La Coruña, Spain

Correspondence to: Dr Raúl de la Fuente Fernández, Servicio de Neurología, Hospital Juan Canalejo, Xubias de Arriba, 84 15006 La Coruña, Spain.


Acute peripheral neuropathy, rhabdomyolysis, and severe lactic acidosis associated with 3243 A to G mitochondrial DNA mutation

Neuropathy can be a complication of progressive external ophthalmoplegia, but is rare in mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS). We report here a case of mitochondrial cytopathy with these unusual manifestations.

The patient is a 33 year old woman who had a normal birth and early development. When she was aged 30, after the stillbirth of her first child, she suffered spontaneous, remitting myalgia of her thighs muscles. At the age of 32 she had intermittent headaches and nausea after physical work. At the age of 33, during her second pregnancy, she felt numbness in her toes and weakness in her legs, which spread to all four extremities over three weeks. She also developed facial weakness. Simultaneously, intrauterine growth retardation occurred and abortion had to be induced. By that time she was too weak to walk. She was diagnosed as having Guillain-Barré syndrome and was given betamethasone (8 mg/day) for two days. This was not effective, and she was transferred to our hospital.

Examination on admission on 16 October 1991 showed average height and