weight, mild, diffuse upper limb weakness, and severe lower limb weakness. The weakness was predominantly distal and deep tendon reflexes were absent in all four limbs. All sensation modalities were impaired in her cheeks, trunk, and limbs, predominantly distal. She complained of dysaesthesia in her left cheek, forearm, and legs. She could barely stand with support. Mild myalgia was present in the thigh muscles. Laboratory tests showed a moderate increase in serum creatine kinase (496 IU/l) and lactate dehydrogenase (355 IU/l) and an increase in glutamic oxaloacetic transaminase, glutamic pyruvic transaminase, and lactate dehydrogenase. On the second day in hospital, motor conduction velocity was normal in all limbs, but a low amplitude motor potential was noted in the tibial nerves. Sensory conduction velocity was normal in the median nerves, but sural nerve action potentials and F waves in the median and tibial nerves were absent. A conventional needle EMG showed that the interference pattern in limb muscles during maximal voluntary contraction was reduced but the motor unit potential amplitudes were not decreased. Motor unit potential amplitudes were normal at rest. Brain CT showed left frontal atrophy despite her normal intelligence. On the morning of the third day in hospital, she complained of nausea and vomiting. She was treated with antacids and metoclopramide. Blood gas analysis showed severe metabolic acidosis (pH 7.01). Despite an intravenous infusion of 400 ml 7% sodium bicarbonate, her metabolic acidosis persisted (pH 6.97-7). She went into a state of shock in the evening and was given continuous haemodialysis for 52 hours. Two plasma exchanges were also given. During the next several days, her condition improved, but the patient convulsed with each breath and severe acidosis, acute renal failure, acute heart failure, and rhabdomyolysis. Even after her acute pulmonary oedema improved, she required mechanical ventilation for four months because of respiratory muscle weakness. The peak values of serum myoglobin and creatine kinase were 18 000 ng/ml and 20 800 IU/l, respectively at this time. Lactic acid and pyruvic acid had reached 294-4 mg/dl and 8-94 mg/dl, respectively by 10 hours after we first became aware of her acidosis. Her CSF protein and cell count were normal in three examinations carried out during the course of her illness. Muscle biopsy from her left quadriceps femoris, taken on the seventh day in hospital, showed ragged red fibres, a mitochondrial myopathy, and scattered small angular fibres. No apparent grouping of fibre type was evident. A sural nerve biopsy three months after admission showed a severe decrease in myelinated fibres. Teased fibre preparations showed myelin ovoid formation and no demyelination.

Mitochondrial DNA analysis after amplification of the tRNAI-u(uuR) region and digestion with HpaII showed 3243 A to G point mutation in blood and muscle samples from the patient, as well as in blood samples from her mother, brothers, and sister. The point mutation was about 40 in her blood and 70 in the muscle sample. She was treated with 3-0 g of sodium succinate, 150 mg of coenzyme Q, and 30 mg of idebenone daily. One month after admission motor conduction velocity of the left tibial nerve was slightly reduced (38-0 m/s) and the amplitude was reduced to 200 µV. Seven months after admission, the motor conduction velocity had recovered to normal, with low amplitude motor potentials in the left tibial nerve. Needle EMG showed polyphasic long duration motor unit potentials in her limb muscles. After 18 months, she was able to stand without support. Her reflexes were improved but the chilulhes were still absent. Sensory disturbances also disappeared but painful dysaesthesia remained in her lower legs. In this case, the diagnosis of MELAS was based on the history, the existence of lactic acidosis, ragged-red fibres, and mitochondrial DNA analysis. There were several new clinically important points in this patient with MELAS.

Firstly, she developed symptoms during her first and second pregnancies; therefore, gestation was possibly an aggravating factor for her. Secondly, she had acute axonal neuropathy, which has rarely been reported with MELAS, although ophthalmoplegia plus or myoclonus epilepsy with ragged Red fibres has been reported to be associated with neuropathy of the patients with MELAS reported by Markesbery and Katsuragi et al. had major features of neuropathy such as sensory disturbance or muscle weakness.

Thirdly, rhabdomyolysis has not been reported in MELAS. Our patient certainly developed rhabdomyolysis after an episode of critical acidosis, because her serum creatinine kinase, which was 506 IU/l on admission, rose to 20 800 U/l the next morning. Hypotension, acidosis itself, or two days of treatment with betamethasone followed by its withdrawal might cause rhabdomyolysis. Fourth, she developed critical lactic acidosis, which improved with continuous haemodialysis and plasma exchange. Therefore, haemodialysis or plasma exchange should be considered in severe lactic acidosis in MELAS. Finally, this case adds a new clinical manifestation associated with the 3243 A to G mutation in mitochondrial DNA.

Hajime Haras Yoshishiro WAKAYAMA
Department of Neurology, Division of Neurology, Department of Medicine, Showa University Fujigaoka Hospital, 1-30 Fujigaoka, Midori-ku, Yokohama 227, Japan

MASASHI TANAKA TAKAYUKI OZAWA
Department of Biological Chemistry, Faculty of Medicine, University of Nagoya, 65 Tsuruma-cho, Showa-ku, Nagoya 466, Japan

Correspondence to: Dr Haras, Division of Neurology, Department of Medicine, Showa University Fujigaoka Hospital, 1-30 Fujigaoka, Midori-ku, Yokohama 227, Japan.

Papilloedema and visual failure in a patient with nocturnal hypoventilation

A morbidly obese woman treated for polymysitis developed symptoms of raised intracranial pressure and visual failure. Invasive CSF pressure monitoring showed normal pressure. Serological investigations suggested episodes of raised intracranial pressure during sleep. These episodes were associated with hypoxia and hypercarbia, and supported a possible relationship between increased intracranial pressure, visual failure, and nocturnal hyperventilation. A 45 year old woman was admitted for investigation of headaches, papilloedema, and progressive visual failure. She was morbidly obese (weight 126 kg, body mass index 52-7 kg/m²) and gave a history of recurrent deep vein thrombosis, pulmonary emboli, and polymysitis (treated with prednisolone and azathioprine). Examination showed bilateral papilloedema with visual acuities of 6/24 in the right eye, and just able to count fingers in the left. A lumbar puncture had only been successful in the sitting position; hence an accurate measurement of the CSF opening pressure had not been recorded. Biochemical and cytological analyses were normal, and the CT of the head was normal. A clinical diagnosis of benign intracranial hypertension was made and a neuro-ophthalmological opinion sought. A left orbital nerve fentanyl anaesthesia was performed without improvement, and so detailed assessment of the intracranial pressure was requested. Access to CSF was achieved with a right frontal venflon catheter connected to a subcutaneous reservoir. Continuous monitoring of intracranial pressure was undertaken from the reservoir (Camino optical transducer) through a fluid filled transcutaneous butterfly needle (21G). Radial arterial blood pressure, middle cerebral artery flow velocities by translacranial Doppler (Scimed), and continuous measurements of peripheral oxygen saturation by Multinex oximetry were also taken, and displayed graphically on a portable computer. The cerebral perfusion pressure (mean radial arterial blood pressure-mean intracranial pressure) was measured. The middle cerebral artery flow velocity (FB) were calculated (pulsatility index PI = FB/mean pressure(FM) were calculated. Recordings were carried out during the night while the patient in her usual sleeping position (20 degrees head up) for up to 10 hours. Before sleep, baseline intracranial pressures of 5-15 mm Hg were recorded. Within one hour of sleep, cycles of high pressure waves (40-45 mm Hg) lasting 10-20 minutes were evident, occurring every 60-120 minutes and superimposed on higher background pressures of 15-25 mm Hg (fig 1). The intracranial pressure waves were accompanied by a fall in cerebral perfusion pressure to as low as 40 mm Hg, and were tightly coupled with increases of middle cerebral artery flow velocity and decreases in cerebrovascular resistance.

Respiratory function was evaluated further; arterial blood gases showed daytime hypocapnia (mean PaO2 = 9-9 kPa or Pao2 with normocapnia = 5-2 kPa). Overnight ventilation studies indicated a mean baseline arterial saturation of 88-2% and end tidal Pco2 of 6-1 kPa. During the periods of raised intracranial pressure,
Further arterial desaturation to a mean of 73% occurred with an increased end tidal PCO₂ of 7.8 kPa. These episodes were associated with hyperventilation (reduced abdominal and chest wall movements) without apnoea or airway obstruction. She was given a continuous positive airway pressure device, which provided marked symptomatic relief and improved the mean baseline arterial saturation to 97.5% but failed to abolish the high nocturnal waves of intracranial pressure. A ventriculoperitoneal shunt was therefore inserted and a further period of monitoring undertaken (fig 2). The changes in middle cerebral artery flow velocity and cerebrovascular resistance still occurred with the episodes of desaturation, but the associated increases in intracranial pressure were abolished. At six month follow up her headaches had disappeared, the papilloedema had resolved, and the acuity in the right eye had improved to 6/12.

Several possible aetiological factors for benign intracranial hypertension may have contributed to the visual deterioration in this patient including subclinical cerebral venous thrombosis. Although the primary aetiology in our case is not known, raised intracranial pressure occurred during sleep and accompanied episodes of hypoxia and hypertocnia. These were associated with haemodynamic changes compatible with cerebral vasodilatation. We therefore suggest that nocturnal hyperventilation producing cerebral hypoxia, hypercapnia, and a subsequent rise in ICP may be secondary to increased cerebral blood volume contributed to the symptoms. Chronic respiratory disease with severe hypercapnia has long been recognized as a cause of raised intracranial pressure and papilloedema1 but there were no such features in this case, and although a significantly raised CSF pressure is required for the diagnosis of benign intracranial hypertension, the clinical and radiological features in our patient were typical of this condition.2 Further, low baseline CSF pressures are often found in patients with chronic benign intracranial hypertension despite persisting papilloedema.4 Two important points are raised. Firstly, abnormal CSF dynamics require continued observation over several hours as baseline CSF pressure may be normal and waves of raised intracranial pressure transient. Inadequate attention to CSF dynamics may partly explain why isolated CSF pressure estimations do not predict the development of papilloedema and visual deterioration.5 Secondly, although nocturnal hyperventilation has not been quoted as a contributing factor in benign intracranial hypertension, a relation with raised intracranial pressure has been found. Overnight monitoring of peripheral oxygen saturation may be a useful addition to the investigation of obese patients with symptoms of raised intracranial pressure.

P J KIRKPATRICK
Y MEYER
N SARKKIES
J D PICKARD
H WHITEHOUSE
P SMIELEWSKI
University Department of Neurosurgery, Flinders Medical Centre, Bedford Park, SA and Addenbrookes Hospital, Hills Rd, Cambridge CB2 2QQ, UK.

Correspondence to: Mr P J Kirkpatrick.


Hemichorea reversible after operation in a boy with cavernous angioma in the head of the caudate nucleus

Hemichorea and hemiballism point to a structural lesion in the contralateral basal ganglia with a large list of possible causes, including various vascular malformations. Cavernous angiomas are one of congenital vascular malformations that are occult on conventional angiography (hence "cryptic" vascular malformations (CVMs) but have a characteristic appearance on MR image.1 The definitive diagnosis and distinction from other cryptic vascular malformations depends on histological examination. The clinical manifestations of cavernous angiomias include epilepsy, acute signs secondary to (recurrent) bleeding, and rarely progressive neurological deficit due to expansion of a mass of thrombosed within the angioma. With the availability of MRI the number of clinical reports on the subject of CVMs has increased. Recently a case was reported of cavernous angioma in the lentiform nucleus that was the first to present with a movement disorder, in this case focal dystonia. Complete resolution was followed by resolution of the symptoms.

We report an 11 year old boy with cavernous angioma in the caudate nucleus, presenting with contralateral hemichorea, evidence of recurrent bleeding, and the disappearance of the hemichorea after surgery. The boy complained of involuntary movements of the right half of his body including his face, arm and leg, that had suddenly started the week before admission. He could not suppress these movements. There was no family history of neurological disease.

The neurological examination on admission showed continuous, random, jerking movements of the face and right upper limb and the right side of the body. Muscle strength, sensation, and reflexes were normal. Brain MRI (figure A) showed a lesion in the head of the caudate nucleus, with the typical aspect of a cavernous angioma.

Two weeks later the boy experienced a sudden deterioration, with involuntary movements of a larger amplitude, more appropriately termed hemiballistic. Surgery was considered appropriate.

With the Leksell stereotactic frame (Elekta Co, Sweden) the shortest route to the lesion via the paramedial frontal lobe was estimated. At the spot that was marked a small burr hole was made and a silastic tube was passed to the border of the lesion with a 4 mm round catheter implantation set. After craniotomy the lesion was reached with the catheter as a guide. The multilayer like vascular lesion was removed completely, including two small haemorrhages.

Histology (figure B) showed a conglomerate of cavernous vascular channels. The wall of these channels consisted of a single inner layer of endothelial cells and an outer layer of collagen of varying thickness. Some vascular spaces were occluded by a recent or an organised thrombus and some vessel walls were partly calcified. Iron pigment was found in and around several vessels, as evidence of prior bleeding. The surrounding brain tissue showed pronounced gliosis and deposition of iron.

In the two months after the operation the hemichorea-hemiballism disappeared completely. Control MRI (figure C) showed complete removal of the angioma.

This case is to our knowledge the first in the literature of a histologically confirmed cavernous angioma presenting with hemichorea. Hemichorea has been described in lesions of the caudate nucleus, and is thought to reflect release phenomena caused by a lesion of the striatal neurons projecting to the external globus pallidus.

The incidence of cavernous angiomas remains obscure. In a consecutive series of 11 children operated on for cerebral vascular malformations five were diagnosed to have cavernous angiomas.2 Scott et al3 state that in some paediatric institutions cavernous angiomas are the most common cerebrovascular malformations encountered. Most cavernous angiomas, however,
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P J Kirkpatrick, T Meyer, N Sarkies, J D Pickard, H Whitehouse and P Smielewski

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