weight, mild, diffuse upper limb weakness, and severe lower limb weakness. The weakness was predominantly distal and left-sided. 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 ankle. She was not able to 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 alanine 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 in the left limb was no longer detectable at rest. Brain CT showed left frontal atrophy despite her normal intelligence. On the morning of the third day in hospital, she complained of pain in and around the left temple. Blood gas analysis showed severe metabolic acidosis (pH 7.01). Despite an intravenous infusion of 400 mL 7% sodium bicarbonate, her metabolic acidosis remained (pH 6.97). 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, with multiple organ failure, including acute respiratory failure, 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 U/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 with marked angiectatic fibres 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 mtDNA was performed on DNA obtained from the patient's peripheral blood. Genomic DNA was isolated from 300 mg of blood collected from each patient. The DNA was digested with restriction enzymes and analysed by Southern blotting. The DNA was hybridized with a radiolabelled probe for mitochondrial DNA. The probe was a 1.4-kb fragment of the mitochondrial genome containing the tRNAI-u(uR) gene. The probe had been subcloned into a plasmid vector. The DNA was hybridized to the probe, and the resulting autoradiographs were analysed by computer-generated densitometry.

In the clinical course, 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 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. Among the patients with MELAS reported by Marksby et al.1 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 creatine kinase was 506 U/L on the 7th day and it 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. Fourthly, we 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.

Papilloedema and visual failure in a patient with nocturnal hyperventilation

A morbidly obese woman treated for polymyositis developed symptoms of raised intracranial pressure and visual failure. Invasive CSF pressure monitoring showed normal pressures; however, she had repeated periods 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 had a history of recurrent deep vein thrombosis, pulmonary emboli, and polymyositis (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 eye. 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 analysis showed no abnormality, and CT of the head was normal.

A clinical diagnosis of benign intracranial hypertension was made and a neuro-ophthalmological opinion sought. A left optic nerve fibre count showed a normal result. Over the following months, she was treated with a right frontal venous cutdown connected to a subcutaneous reservoir. Continuous monitoring of intracranial pressure was undertaken from the reservoir (Camino optical transducers) through a fluid filled transcatheter butterfly needle (21G). Radial arterial blood pressure, middle cerebral artery flow velocities by trancranial Doppler (Scimed), and continuous measurements of peripheral oxygen saturation by Multimex oxymetry were also taken, and displayed graphically on a portable computer. The cerebral perfusion pressure (mean arterial blood pressure-intracranial pressure) was 150 mmHg. The middle cerebral artery flow velocity (FV) were calculated (pulsatility index = FV amplitude/FV mean) were calculated. Recordings were carried out during the night, with the patient in her usual sleeping position (20 degrees head up) for up to 10 hours. Before sleep, baseline intracranial pressures of 5–15 mmHg 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 mmHg (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 hypoxia (mean PaO₂ 91 mm Hg with nocturnal capillary PaO₂ 52 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,
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 angioma is an arteriovenous malformation that occurs on conventional angiography (hence "cryptic") vascular malformations (CVMs) but have a characteristic appearance on MR image. The definitive diagnosis and distinction from other cryptic vascular malformations depends on histological examination. The clinical manifestations of cavernous angiomas include epilepsy, acute signs secondary to (recurrent) bleeding, and rarely progressive neurological deficit due to expansion of a mass effect 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 neck, which affected 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 site of the lesion the burr hole was made and a silastic tube was passed to the border of the lesion with a Backlund catheter implantation set. After craniotomy the lesion was reached with the catheter as a guide. The mulberry like vascular lesion was removed completely, including two small haemorrhages.

Histology (figure B) showed a conglomerate of arteriovenous 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 pigmentation 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 natural course 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. Scott et al state that in some paediatric institutions cavernous angiomas are the most common cerebrovascular malformations encountered. Most cavernous angiomas, however,
Papilloedema and visual failure in a patient with nocturnal hypoventilation.

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