CASE REPORTS

Mondor meets Trendelenburg: penile vein thrombosis after varicose vein surgery

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Aetiological factors in superficial dorsal penile vein thrombosis include trauma, vigorous sexual activity, pelvic tumours and a distended bladder[1,2]. Isolated penile vein phlebitis was originally described in 1958[3] as a variant of Mondor's disease, which is a more generalized superficial venous thrombosis of the chest wall.

CASE HISTORIES

Three cases of superficial penile vein thrombosis were identified prospectively from a series of 231 men who underwent long saphenous vein surgery over a 4-year period. A total of 350 legs were operated on—241 legs for primary and 109 for recurrent varicose veins. A standard surgical procedure was used including flush ligation of the saphenofemoral junction with braided polyester and exposure of the femoral vein to identify all tributaries. The long saphenous vein was removed by inversion pin-stripping to the knee, with retrograde stripping for recurrences. The operations were performed by a single surgeon (AN) and all patients were seen for clinic review at 2 weeks.

Case 1

A man aged 39, previously fit and well, underwent bilateral long saphenous and left short saphenous surgery. At follow-up he was noted to have thrombophlebitis in one of the tributaries of the right long saphenous vein extending up onto the shaft of the penis. This settled without treatment.

Case 2

A man aged 44 underwent bilateral repeat long saphenous vein surgery, having been operated on 4 years previously.

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At follow-up he had an area of superficial thrombophlebitis down one of the left lateral dorsal veins of the penis. This was uncomfortable during sexual intercourse. He was seen again 4 months later when all had resolved.

Case 3

A man aged 61 had bilateral long saphenous vein surgery. Operation on the left side was complicated by the removal of a subcutaneous malignant melanoma from the skin over the knee. Postoperatively he developed thrombophlebitis on the right side of the penile root, extending onto the dorsum of the penis. This settled over 4–5 weeks.

COMMENT

We have been unable to find previous reports of thrombophlebitis of the superficial dorsal vein of the penis as a complication of long saphenous vein surgery. The incidence in this series, 3 per 231 patients, exceeds the 1% level of risk above which patients are usually warned as part of the consent process.

Anatomical studies of penile venous outflow have been performed by selective venography[4,5]. The superficial dorsal vein drains the prepuce and skin of the penis into the right and left external pudendal veins and thence into the long saphenous vein at the groin. Superficial ventral veins also drain into the superficial dorsal vein. There is almost complete separation of the superficial and deep venous systems in the penis with the exception of occasional direct communications between the deep and superficial dorsal veins. It is therefore anatomically possible that a surgeon performing bilateral ligation of the tributaries of the saphenofemoral junction will interrupt the superficial venous drainage of the penis with consequent venous stasis and thrombophlebitis. In 20% of cases the superficial dorsal vein drainage is unilateral, presumably increasing the risk of venous occlusion after surgery.

Superficial thrombophlebitis of the penis presents with cord-like induration of the vein, associated with mild discomfort and inflammation. Most cases are reported to resolve completely, as did ours, with no permanent adverse effects on sexual function. Treatment with anti-inflammatory drugs is said to relieve symptoms but not to hasten resolution.

It is possible that this condition is underreported since it is self-limiting and most patients who undergo long saphenous surgery are not followed up.

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Reduced consciousness with a runny nose

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In a patient with an old head injury, violent nose-blowing can have dangerous consequences.

CASE HISTORY

A man aged 54 had a three-week history of nasal discharge and reduced hearing with pain in the left ear. The earache, accompanied by a sensation of fluid in the ear but no discharge, had begun after an episode of violent nose-blowing. He also had a persistent watery nasal discharge, which he had been trying to relieve by further blowing of the nose. His general practitioner treated him unsuccessfully for otitis, with antibiotics and syringing. A few hours before admission to hospital the patient had become confused and disoriented. In his medical history the only noteworthy incident was a childhood left occipital depressed skull fracture without any intracranial injuries.

On admission to our department the Glasgow Coma Score was 11 (eyes 4, verbal 1, motor 6). He had fluid behind the left tympanic membrane and temperature was

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37.3 °C; there were no focal neurological deficits. Leaning him forward produced large amounts of watery fluid leakage from his nose.

A computed tomographic scan revealed extensive intraventricular air in the lateral and third ventricles. There was also evidence of previous skull trauma with underlying encephalomalacia in the left occipital region but no base-of-skull fractures (Figure 1). A magnetic resonance scan ruled out a cerebral abscess. There was no evidence of infection in cerebrospinal fluid (CSF) obtained by lumbar puncture. Subsequently, a left mastoidectomy was performed; a CSF fistula was found and repaired and a lumbar drain was left *in situ* for 5 days. Postoperatively the Glasgow Coma Score improved to 15. There was no further CSF leak, and he was discharged home with strict instructions not to blow his nose.

COMMENT

The term pneumocephalus—a collection of air within the skull—was coined by Wolff in 1914[1], but Chiari in 1884 was the first to report a case of intracranial pneumocephalus, in a patient with severe ethmoiditis[2]. Most cases of pneumocephalus today are due to head trauma[3,4], followed in order of frequency by neoplasia, infections (such as otitis[3,5], sinusitis[3,6] and intracranial infection[3,7]) and medical interventions[3].

Two main theories have been suggested for the pathophysiological basis of pneumocephalus. The first is the 'ball-valve' mechanism, whereby air enters the intracranial space (by coughing, sneezing or nose-blowing)
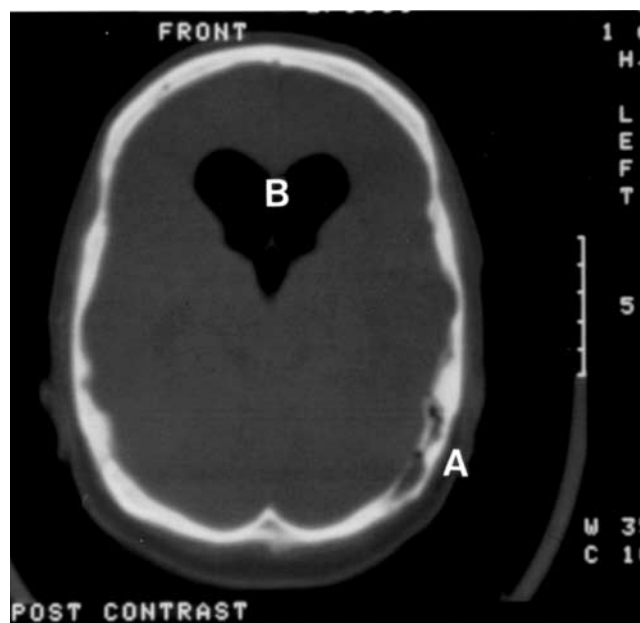

Figure 1 **Bone window computed tomographic scan showing thin plate of bone at site of old occipital skull fracture (A) and intraventricular air (B)**

through a cranial defect and becomes trapped by the meninges or brain, increasing the intracranial pressure. Usually there is no associated CSF leak[8]. The second is the 'inverted-bottle' mechanism: as CSF leaks from the intracranial space, negative pressure within the cranium causes air to be sucked in[3].

In our case pneumocephalus developed unusually late after trauma. Following the childhood head injury, the breeched dura and mastoid air cells were separated only by a thin plate of bone. This was eventually fractured by violent blowing of the nose, causing CSF to leak first into the mastoid air cells and then into the middle ear, down the eustachian tube and out through the nose. The CSF leak in turn caused intracranial hypotension, permitting air entry into the cranial cavity. The initial pneumocephalus therefore seems in this case to have developed through the inverted-bottle mechanism. The pneumocephalus, however, was made worse by the patient's continuous nose-blowing to clear the troublesome rhinorrhoea. A ball-valve mechanism therefore may also be implicated here whereby air entered the cranial cavity via the middle ear and the eustachian tube as a result of a rise in the nasopharyngeal pressure during nose blowing[8]. Air reached the intraventricular space either by direct migration or through the foramina of Luschka and Magendie. Initially, the patient's ventricular system was buffering against changes in intracranial pressure until it was sufficiently drained of CSF such that it could no longer compensate, leading to symptomatic intracranial hypotension.

This case illustrates how post-traumatic pneumocephalus can develop many years after a head injury. In a patient with transient watery nasal discharge and a history of cranial trauma, the possibility of CSF rhinorrhoea should be borne in mind.

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Four children crushed in their driveways

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Pedestrian injuries are a leading cause of mortality and morbidity in childhood[1]. In most cases the child is struck by a fast-moving vehicle, but injuries sustained in the driveway form a distinct subgroup[2]. We report four cases seen over a 5-year period (1995–1999) in a regional paediatric surgical centre.

CASE HISTORIES

Case 1

A girl aged 2 sustained a severe crush injury to the chest and abdomen after being run over by a neighbour who was reversing the car in the driveway. In the accident and emergency department she was noted to have a widespread petechial rash and suffusion over her upper chest, neck and face. There was patterned bruising on the abdomen. On primary survey her airway was patent, but she was tachypnoeic and required face mask oxygen to maintain a normal oxygen saturation. Her peripheral perfusion was poor but her haemodynamic status became normal after an intravenous bolus of 20 mL/kg of crystalloid.

A computerized tomographic (CT) scan of the abdomen and chest revealed evidence of pulmonary contusion, a capsular tear in the spleen, contusion around the second part of the duodenum and superficial lacerations of the liver, particularly in the region of the caudate lobe. Her injury severity score (ISS)[3] was 26. She was electively intubated and ventilated and received intravenous analgesia, broad-spectrum antibiotics, chest physiotherapy and nasogastric enteral feeding. She was managed non-operatively and serial ultrasound imaging demonstrated resolution of her intra-abdominal injuries. She was extubated after 6 days and discharged home after 17 days, eventually making a complete recovery.

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Case 2

A 15-month-old toddler sustained a blunt injury to the abdomen as a result of being run over by her mother, who was reversing the car in the driveway. On arrival at hospital she was normotensive, mildly tachycardic and neurologically normal. CT imaging of her head, chest and abdomen demonstrated a small left pneumothorax with ipsilateral apical pulmonary contusion and a right parieto-temporal skull fracture. She also had a closed fracture of the right femoral shaft. Injury severity score was 14. Her injuries were treated non-operatively and she was discharged home 17 days after admission.

Case 3

A toddler aged 13 months was transferred from another hospital having been crushed by the bumper of a car reversing in her driveway. At the referring hospital she required insertion of a chest drain for a right pneumothorax which was associated with radiological signs of pulmonary contusion. Her ISS was 11. After transfer she remained clinically stable apart from a mild pyrexia which resolved after treatment with intravenous broad-spectrum antibiotics. She was discharged home 3 days after admission.

Case 4

A 4-year-old boy was admitted after sustaining a crush injury of the chest. Sitting on the back seat of a car without a seatbelt he had opened the rear door, fallen out and been run over whilst his father was reversing in the driveway. A CT scan of the chest revealed fractures of the right second and fifth ribs with a small ipsilateral haemopneumothorax and right upper lobe pulmonary contusion. Abdominal CT scan was normal and his ISS was 11. Treatment included insertion of a chest drain, chest physiotherapy and the administration of broad-spectrum intravenous antibiotics and opioid analgesia. He remained haemodynamically stable and was discharged home after 3 days, making a complete recovery.

COMMENT

Reported experience of driveway injuries in children has been largely confined to the USA, where they represent about 10% of injuries in child pedestrians and are commonly fatal[4–7]. Is our experience of four cases in 5 years, all non-fatal, likely to reflect the true picture? We looked at two other local sources, in Leeds and Sheffield. During the same period, 1995–1999, five fatalities from motor vehicle accidents in children under 5 were recorded in the accident and emergency departments of the Leeds Teaching Hospitals NHS Trust. None was the result of a driveway injury.

In the accident and emergency records at Sheffield Children's Hospital 67 road traffic accidents affecting children in the 0–5-year age group could be identified over the same period. 53 were pedestrian injuries, 6 of which were fatal. Again, none was from a driveway injury. Perhaps the difference from American experience reflects the proportion of residences with cars and driveways.

In this series, all the children sustained blunt chest/abdominal injuries and one had additional limb and head injuries. The age distribution is consistent with previous evidence that 1–2-year-old toddlers are most at risk from driveway injuries[5–7]. In the USA, these accidents occur most often in the spring and summer; two of our children sustained their injuries during the autumn and two in summer. Winn et al.[7] reported five fatal driveway injuries at a median age of 14 months. Four of the children died as a result of a head injury and one from head, chest and abdominal injuries. In the largest series of driveway injuries, 37% of young children ($<$4 years) sustained a closed head injury, 32% had torso trauma, 46% had skeletal trauma, and the fatality rate was 20%[4].

Potential risk factors for driveway injuries include a children's play area adjacent to the driveway, shared driveways, male sex, and low socioeconomic status[1,2,8]. Most incidents occur during daylight hours[1,7]. Unsupervised children in a vehicle are capable of setting the car in motion and injuring themselves directly or other children playing in the driveway[4,9].

Separation of the driveway and children's play area by a low-level fence may be one way to prevent these injuries[2] but probably more important is to make parents aware of the hazard. Toddlers may be difficult to see in the rear-view mirror, particularly with the large four-wheel-drive vehicles now popular, but the frequency of these injuries is not high enough to demand redesign to improve rear visibility (and there may be competing safety considerations). A car reverse alarm may be a helpful warning of danger.

Although uncommon, driveway injuries tend to be severe and may be fatal. They are devastating for parents and others who are responsible for the accident. These injuries deserve wider recognition in accident prevention programmes.

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Stiff-man syndrome in childhood

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Stiff-man syndrome is a disease of sporadic occurrence. An autoimmune pathogenesis is thought to involve the spinal interneuronal circuits, resulting in isolation of the motor neurons from their inhibitory influence[1]. Most of the reported cases have been in adults.

CASE HISTORY

A boy aged 11 was seen because of intermittent stiffness and painful spasms of the limb muscles with difficulty in walking of 5 years' duration, all of which worsened after taking a few steps (Figure 1). There was no family history of a similar illness. On examination there was an exaggerated lumbar lordosis. Mental functions and cranial nerves were normal. There was no demonstrable muscle weakness. On palpation of his thighs and calves the muscles felt firm and were tender. They would suddenly stiffen when touched or during a startle induced by a loud sound. The muscles around the forearm, knee and ankle would stiffen with twisted posturing and the patient would find it impossible to move these joints until the spasm subsided. This resulted in an awkward posture and a clumsy wide-based gait when he was asked to walk a few steps. The trunk muscles were

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little affected. He could not get up from the squatting position, since any activity aggravated the painful spasms. All the deep tendon reflexes were brisk. There was no clinical evidence of either peripheral nerve or spinal cord disease. Autonomic and sphincter function were normal. No myoclonus was ever noticed.

The following were within normal limits: routine haematological tests, thyroid function, antinuclear antibody, anti-double-stranded DNA antibody, blood glucose, serum lactate (pre and post exercise), liver function, serum copper and serum ceruloplasmin. Nerve conduction was normal but needle electromyography showed continuous motor unit activity (CMUA) at rest and complete disappearance of motor activity with intravenous administration of diazepam (Figure 2). CMUA was seen in the gastrocnemius, quadriceps and abductor pollicis brevis muscles.

He was started on oral diazepam 2 mg thrice daily (body weight 19.5 kg). Before treatment he could walk 4–5 m on a level floor and was then halted by painful spasms. After 5 days of diazepam, the stiffness and spasms were much less severe and he could walk 40–50 m without difficulty. In addition he could get up from the squatting position. He was discharged on the same dose of diazepam, now able to perform activities of daily living with little discomfort.

COMMENT

The diagnosis in this patient was based on the clinical features of stimulus-sensitive and activity-induced painful
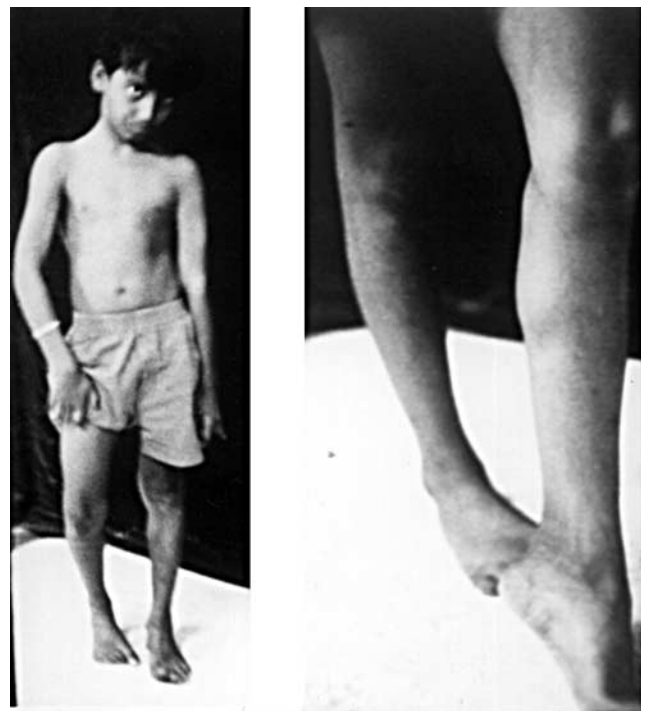

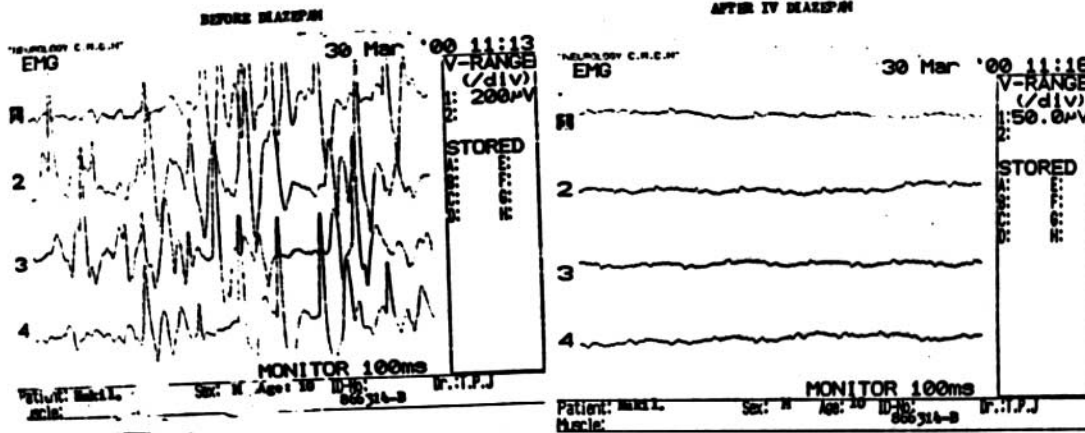*Figure 1* **Contracted muscles of legs**

Figure 2 **Electromyogram of left gastrocnemius at rest with continuous motor unit activity, seen to disappear completely after administration of diazepam (5 mg, intravenous)**

muscle spasms in association with continuous motor unit activity seen on the EMG. The pattern of muscles involved was consistent with the stiff-limb syndrome described by Barker *et al.*[2], and the response to diazepam strengthened the diagnosis. Differential diagnoses such as myotonia, pseudomyotonia, myokymia, neuromyotonia, paroxysmal kinesigenic dystonia, paroxysmal exertion-induced dystonia, extrapyramidal syndromes, pyramidal dysfunction, tetanus, hyperexplexia, spinal cord disorders and reflex sympathetic dystrophy were considered and excluded.

Various subgroups of the stiff-man syndrome have been described[2]. Progressive encephalomyelitis with generalized rigidity[4] usually manifests as part of a paraneoplastic syndrome. It invariably follows a progressive and unrelenting course leading to death within a few months. Stiff-trunk (man) syndrome associated with autoantibodies[5] is characterized by rigidity predominantly of the neck, trunk and proximal leg muscles, painful spasms of axial muscles and lumbar hyperlordosis. This is the commonest presentation. Typically, the upper limbs, distal lower limbs, sphincters and cranial nerves are spared. Almost all patients have antibodies to glutamic acid decarboxylase in blood and cerebrospinal fluid. Some also have antibodies to pancreatic beta-cells, thyroid cell microsomes, gastric parietal cells and smooth muscle cells[6]. A stiff-man syndrome associated with malignancies[7,8] has been described in patients with breast and small-cell lung carcinomas. These patients have antibodies to a synaptic vesicle protein, amphiphysin. Stiff-limb syndrome[2] is characterized by rigidity and abnormal postures of the legs with painful spasms. The trunk is less involved. The sphincters and the brainstem are involved in many patients. A few of these patients have antibodies to glutamic acid decarboxylase. There have been reports of patients with associated generalized myoclonus[9], also called jerking stiff-man syndrome, possibly a subgroup of this variety.

Autoantibodies are typically present in the stiff-trunk subgroup, but are occasionally seen in the stiff-limb variety too. Therefore, insulin-dependent diabetes, anaemia, and other associated autoimmune manifestations are more common in the former.

The most striking aspect of this patient is the onset at age 6, since most reports have been on adults. Familial stiff-man syndrome has been described with rigidity and muscle spasms inherited in an autosomal dominant pattern[10], but this childhood case seems to have been sporadic.

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Bilateral hypoperfusion retinopathy

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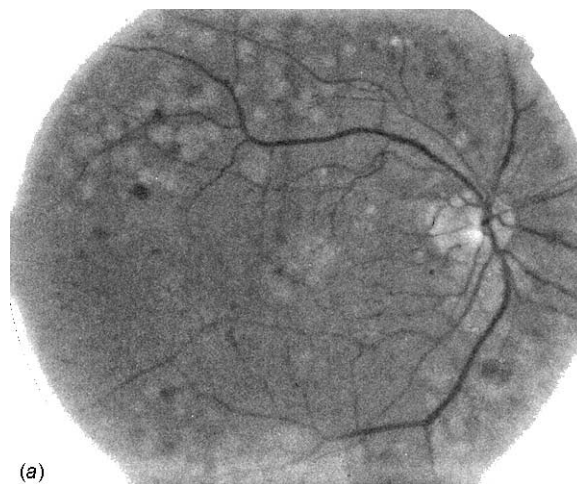D McCormack DCH DRACOG[1]
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Hypoperfusion retinopathy, also known as venous stasis or slow flow retinopathy, is a severe and blinding manifestation of carotid artery disease. Treatment by carotid endarterectomy, combined with panretinal photocoagulation, can stabilize vision.
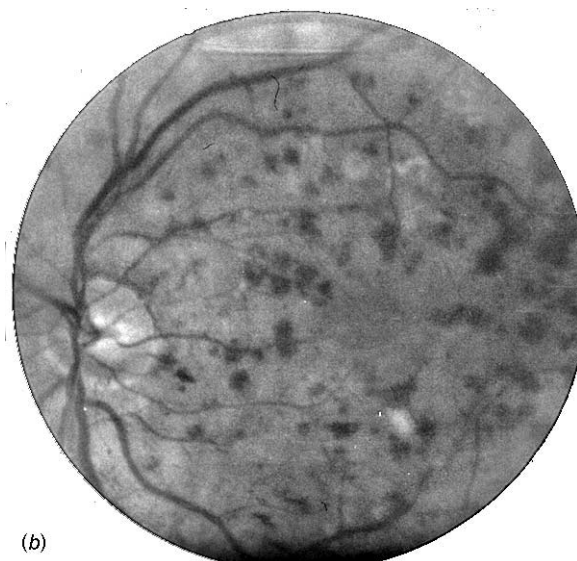
CASE HISTORY

A man aged 83 described bilateral progressive visual loss over three weeks, worse in the left than the right eye and with a notable deterioration during the past three days. Medical history included non-insulin-dependent diabetes mellitus, hypertension, ischaemic heart disease, and peripheral vascular disease. On examination corrected visual acuity (VA) was 6/9 in the right eye and 6/12 in the left. The intraocular pressure was slightly raised in the left eye. Fundoscopy revealed small intraretinal haemorrhages in all quadrants in the right eye. In the left eye, superficial and deep intraretinal haemorrhages were densely scattered throughout the fundus and one cotton-wool spot was seen (Figure 1). The retinal veins were dilated and of irregular calibre. The initial diagnosis was of right background diabetic retinopathy and left non-ischaemic central retinal vein occlusion. Blood tests were normal except for moderately raised cholesterol, glucose and HbA_{1c}.
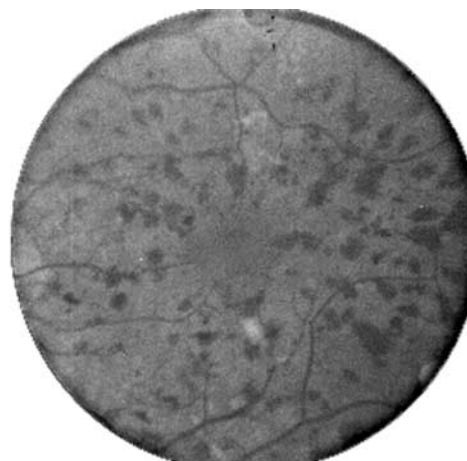
Within three days the vision had deteriorated to 6/18 in the right eye and 6/60 in the left. The pupillary margin of both irides showed small dilated vessels, consistent with early neovascularization of the iris, leading to a diagnosis of bilateral hypoperfusion retinopathy. Bilateral retinal argon laser photocoagulation was begun immediately. A carotid duplex ultrasound scan revealed calcified atheromatous plaques in both common carotid arteries and a 90–99% stenosis of the right internal carotid artery (ICA). No flow could be seen in the proximal left ICA; the distal ICA could

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Figure 1 **Fundus photographs obtained on second visit.** (*a*) Right fundus: some midperipheral retinal haemorrhages superotemporally to the vascular arcade. (*b*) Left fundus: numerous posterior pole and midperipheral retinal haemorrhages. (*c*) Higher magnification of left posterior pole. One cotton-wool spot can be seen below the macula

not be displayed. A consultation with a vascular surgeon, with a view to possible carotid endarterectomy, was arranged, but this would have been a high-risk procedure for a person in such poor general health and the patient opted for conservative management. Within two months of presentation he underwent three sessions of panretinal photocoagulation to both eyes. The progression of ischaemia was stopped in the right eye, but the left developed iris neovascularization and rubeotic glaucoma, necessitating cryoablation of the anterior retina and cyclocryotherapy to the ciliary body to bring the intraocular pressure under control. Visual acuity is currently stable at 6/18 in the right eye and hand movements in the left eye.

COMMENT

Chronic hypoperfusion retinopathy is seen in severe stenosis or complete occlusion of an internal carotid artery. Patients often have a history of hypertension, peripheral vascular, cerebrovascular or ischaemic heart disease, or diabetes[1]. The condition is usually ipsilateral to the more severely affected carotid artery. The pathophysiology is that chronic low arterial perfusion pressure leads to retinal hypoxia. Slowing of the retinal circulation time causes dilatation and tortuosity of the retinal veins, breakdown of capillary walls, superficial (flame-shaped) and deep (dot-blot) retinal haemorrhages, macular oedema and eventual neovascular proliferation in retina and iris which occurs as a response to the release of angiogenic factors from the ischaemic retina[2,3]. The fundoscopic picture resembles diabetic retinopathy but there are two distinguishing signs: in the early stages, hypoperfusion retinopathy affects the retinal midperiphery rather than the posterior pole; and it is usually unilateral[2,3]. Contrasting features to a central retinal vein occlusion are the absence of optic disc swelling and the midperipheral location of the haemorrhages. In more profound ocular hypoperfusion, known as ocular ischaemic syndrome, the retinopathy is associated with anterior segment ischaemia as reflected in corneal oedema and ischaemic uveitis, neovascularization of the iris and raised intraocular pressure secondary to neovascular glaucoma. A poorly reactive pupil is often seen and patients complain of severe orbital pain[2]. Visual prognosis is poor[1].

The differential diagnosis includes diabetic retinopathy, non-ischaemic central retinal vein occlusion, hyperviscosity syndromes such as polycythaemia, Waldenström's macroglobulinaemia, haemoglobinopathies, myelomatosis, and lymphoma[3], all of which need to be excluded by investigations.

Chronic ocular ischaemia is treated by panretinal photocoagulation, which reduces the production of angiogenic factors by the hypoxic retina[2]. Reduction of intraocular pressure to improve ocular perfusion can be achieved by topical β-blockers and/or topical or systemic carbonic anhydrase inhibitors[1]. If possible, this is followed by management of the carotid occlusion by endarterectomy or a bypass from the superficial temporal artery to the middle cerebral artery. Reports on the outcome of carotid endarterectomy in patients with ocular ischaemic syndrome show conflicting results[1,4,5]. The decision about cerebrovascular surgery needs to be made on an individual basis, and the risk of perioperative complications, especially strokes, needs to be balanced against the expected benefit— namely, stabilization of vision[2].

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Gitelman's syndrome

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Gitelman's syndrome is a primary renal tubular hypokalaemic metabolic alkalosis with hypocalciuria and hypomagnesaemia, a mild variant of Bartter's syndrome.

CASE HISTORY

A man aged 37 had experienced episodes of generalized weakness along with recurrent cramps for the past year. The weakness, which was precipitated by resting after exertion and by a heavy carbohydrate meal, predominantly affected the proximal muscles of all four limbs. Each episode lasted a few hours, and between attacks his only

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symptoms were polyuria and nocturia. There was no vomiting or diarrhoea and he was not taking any medications. No other family member had a similar illness and there was no history of parental consanguinity.

On examination during an attack the quadriparesis was flaccid and associated with depressed tendon jerks. Muscles of the eyes, face, tongue, pharynx, larynx, diaphragm and sphincters were not involved. Blood pressure was normal. Serum potassium was persistently low even between the attacks—1.7 mmol/L (normal range 3.5–5.0). An excessive loss of potassium, chloride and magnesium was detected in the alkaline urine—potassium 300 mmol/24h (25–125), chloride 650 mmol/24h (110–250), magnesium 56 mmol/24h (2–5). He also had hypomagnesaemia, hypochloraemia and metabolic alkalosis—magnesium 0.5 mmol/L (0.8–1.2), chloride 89 mmol/L (98–106), bicarbonate 33 mmol/L (22–30), blood pH 7.48 (7.35–7.45). The urinary calcium excretion was subnormal at 1.0 mmol/24h (2.5–7.5). Serum calcium was 2.5 mmol/L (2.2–2.6) and sodium 140 mmol/L (136–145). Thyroid function tests (T3, T4, TSH) were normal.

The hypomagnesaemia responded to oral magnesium supplements, but the hypokalaemia persisted despite large doses of oral and parenteral potassium. Oral indomethacin 25 mg 8-hourly was then started empirically and the patient became normokalaemic within 24 hours. He was discharged with advice to continue oral potassium and magnesium supplements along with indomethacin. He remains symptom-free and normokalaemic after four months of follow-up.

COMMENT

Episodic weakness beginning after age 25 is almost never due to primary periodic paralysis[1]. Further, a low serum potassium between attacks and absence of a similar family history should raise strong suspicion of a secondary disorder[2]. Thyrotoxicosis can mimic periodic paralysis, especially in Asians, and had to be ruled out here[3]. Secondary hypokalaemic periodic paralysis with normotension, alkaline urine and metabolic alkalosis is seen in hyperplasia of the juxtaglomerular apparatus with hyper-aldosteronism[2,3]. Also known as Bartter's syndrome, this condition begins in childhood and presents with short stature, polyuria, polydipsia, and a tendency to dehydration during infancy or before school age. It is associated with polyhydramnios or premature delivery[4,5]. Classic Bartter's syndrome is a severe congenital disease that is inevitably recognized before the age of 6 years[5]. Gitelman's syndrome is a milder variant, with more episodes of tetany and a later presentation. It also differs from Bartter's in being associated with hypocalciuria, so these two variants of primary renal tubular hypokalaemic metabolic alkalosis can easily be distinguished by measurement of urinary calcium[4,5]. Renal magnesium wasting is seen in all patients with Gitelman's syndrome and in about one-third of those with Bartter's syndrome[5]. The tetany may be attributable to exacerbation of alkalosis and consequent low ionized plasma calcium in the presence of hypomagnesaemia. Usually the underlying condition is obvious, but recurrent episodes of transient weakness can sometimes be difficult to distinguish from primary hypokalaemic periodic paralysis[3]. The paroxysmal nature of the attacks is unexplained and it is not known whether the ionic shifts during the attacks are the same as in the primary hypokalaemic form. The timing of the attacks may relate to fluctuations in catecholamine levels and associated regulation of sodium potassium ATPase function[2]. Insulin causes movement of potassium into cells which may account for the precipitation of paralysis by large carbohydrate meals.

The laboratory characteristics of classic Bartter's syndrome may be mimicked by treatment with loop diuretics, which bind to and inhibit the luminal sodium-potassium-chloride cotransporter found in the thick ascending limb of loop of Henle. In the case of Gitelman's syndrome the laboratory characteristics resemble those induced by thiazides, which bind to and inhibit the luminal sodium-chloride cotransporter in the distal convoluted tubule[5]. The clinical features of Bartter's syndrome (and possibly also Gitelman's syndrome) are to a large extent caused by raised concentrations of prostaglandins[6]. By direct action and through stimulation of natriuresis, these compounds stimulate renin secretion, thereby promoting potassium wasting. They also have a direct effect on aldosterone biosynthesis. Indomethacin has been used in both syndromes, but especially Bartter's, for the beneficial effects of inhibiting prostaglandin synthesis.

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Surgical treatment of cortical tremor

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Cortical tremor is easily mistaken for parkinsonism. Ablation of the underlying lesion can be curative.

CASE HISTORY

A woman aged 50 sought advice because of continous shaking of her left hand, which interfered with her daily life. She was right-handed. The tremor had begun gradually about six weeks earlier, worsening to the point where she could not get food to her mouth with a fork in the left hand. Alcohol made it worse. There was no precipitating cause that she could recall; she had not been exposed to antiemetics, neuroleptics or antidepressants. On examination the tremor was present at rest and was aggravated by posture and action. There was no stimulus sensitivity. She had difficulty in performing rapidly alternating tasks and fine finger movements with the left hand. Neurological findings were otherwise normal. Early parkinsonism and epilepsia partialis continua were prominent in the differential diagnosis. Magnetic resonance imaging revealed an arteriovenous malformation, approximately 3 cm in maximum diameter, at the medial aspect of the right cerebral hemisphere just superior to the rostrum of the corpus callosum, affecting the cingulate gyrus (Figure 1). Digital subtraction arteriography indicated that its blood supply was from the right anterior cerebral artery (pericallosal and callosomarginal branches). Venous drainage was to the superior sagittal sinus (Figure 2).

The nidus of the malformation was catheterized (1.2F microcatheter) and the lesion was partly embolized with glue mixture. Some of the arterial supply to the malformation was from small perforating vessels which could not be catheterized. The tremor improved somewhat. The malformation was then excised completely via a right parasagittal frontal craniotomy, with preservation of the pericallosal and callosomarginal arteries. Complete excision was confirmed by intraoperative angiography. The patient's symptoms resolved completely and she was discharged
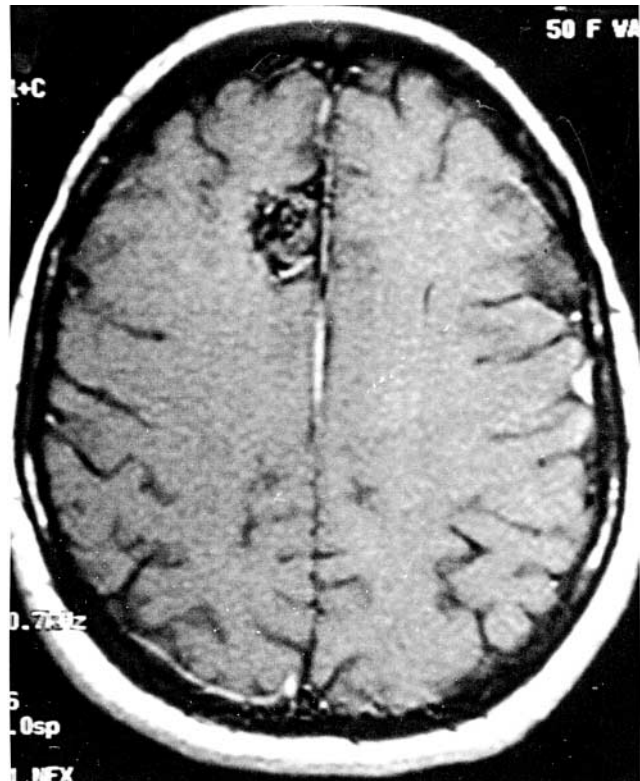
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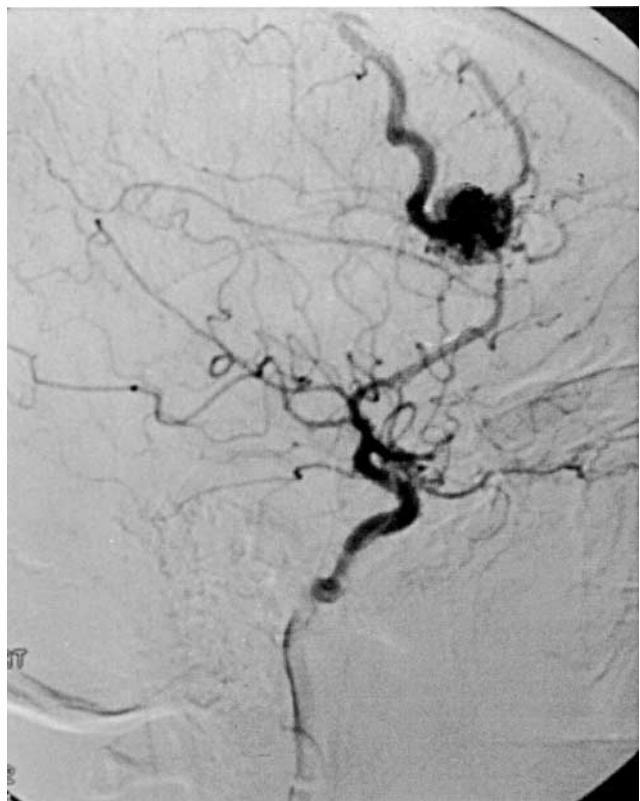Figure 1 **Sagittal magnetic resonance scan showing the right frontal lesion**

Figure 2 **Cerebral angiogram of right internal carotid artery demonstrating the arteriovenous malformation** (shown as the blush of vessels with early draining veins into the sagittal sinus)

without further disability. On review at one year she had a very mild tremor in the left hand which did not interfere with her social or domestic activities.

COMMENT

Various tumours and vascular lesions, including arteriovenous malformations, have been reported as causing kinetic tremors[1–6]—hence the calls for brain scanning in patients with parkinsonsism[7]. Evidente *et al.*[5] described improvement, though not abolition, of tremor after removal of a convexity meningioma. Others, by contrast, have described development of tremor *after* meningioma surgery[3,8]. The mechanism in our patient may have been rapid growth or enlargement of the malformation, with disruption of the pathways in the right motor cortex. With tumours, some workers suggest that the basal ganglia can be involved via oedema or vascular insufficiency; others hypothesize dysfunction of the premotor cortex or deafferentation of the primary motor cortex[5]. Wang *et al.*[3] suggest that the sensory cortex is normally under inhibitory influences from the ipsilateral frontal motor cortex; thus damage from trauma or ischaemia might cause loss of such inhibition. In our patient we think the presentation and outcome suggestive of an ischaemic mechanism. The arteriovenous malformation was in the cingulate gyrus and we hypothesize that a vascular 'steal' phenomenon, more pronounced on activity, was responsible for the tremor and its variability.

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Puzzle prizewinner

In the puzzle set by Richard Krysztopik the final diagnosis was emphysematous cystitis (see May issue, p. 247). Fourteen correct answers were received, the most favoured alternative diagnosis being caecal volvulus. The prizewinning entry, drawn from a hat, was from John Thurston (Dartford), to whom goes a copy of Robert Richardson's *Larrey: Surgeon to Napoleon's Imperial Guard*. The other thirteen who sent entirely correct answers were: R Basu (Southampton), A Bhattacharyya (Manchester), J Hopkins (Birmingham), J Houghton (Chester), A Huang (London), R Linton (London), M Long (Harlow), R Shaw (Leamington Spa), J Singleton (Salernes, France), R Sivakumar (London), D Stoker (Henley on Thames), T Terry (Leicester), S Ward (Stafford).