

showed 33 lymphocytes/mm³, 7.33 g/l protein, and 1.4 mmol/l sugar (blood sugar 6.6 mmol/l). Subsequent CSF culture was positive for *Mycobacterium tuberculosis*, which was sensitive to isoniazid, pyrazinamide, and rifampicin but highly resistant to streptomycin. The patient was HIV and cryptococcal antigen negative. He was diagnosed as having tuberculous meningitis and treated with isoniazid (500 mg/day), rifampicin (600 mg/day), pyrazinamide (2 g/day), and streptomycin (750 mg/day).

Although there was an initial improvement in his symptoms after three weeks in hospital there was a rapid deterioration in his condition over a period of 24 to 48 hours, with dysphagia, dysarthria, and the development of a spastic tetraparesis that initially allowed him to walk with the support of one person. An enhanced brain CT was normal. It was assumed that the patient had had a brainstem infarct, which was confirmed on MRI of the posterior fossa. The possibility of spinal cord complications of his tuberculous meningitis was considered. Spinal cord MRI showed a normal cord but clearcut gadolinium enhancement in the spinal meninges (figure). Dexamethasone was added to his antituberculous treatment for four weeks.

The patient remained profoundly disabled with deteriorating lower limb function. He became paraplegic with pronounced flexor spasms. He was catheterised and fed by a gastrostomy tube. Muscle wasting developed in the upper limbs and there was associated upper limb areflexia. Because of his increasing disability, a repeat scan was carried out four months after the initial spinal MRI. This showed that almost the whole of the cervical spinal cord was filled with loculated discrete hypointense areas outlined by an intensely gadolinium enhancing rim.

Repeat CSF examination showed < 3 white blood cells/mm³, > 1 g/l protein, and 2 mmol/l sugar (blood sugar 4.4 mmol/l). The patient's antituberculous regime was changed to 900 mg/day rifampicin, 2 g/day pyrazinamide, and 500 mg/day isoniazid. Dexamethasone (16 mg/day) was reintroduced.

There was no major change in the patient's overall condition. He was discharged home six months after his initial admission. He resumed oral feeding but remained paraplegic with limited upper limb function: He remained catheterised. Cerebral function was intact. Follow up third MRI eight months after the initial scan and four months after the second showed no dramatic change from the second. A new area of loculation had appeared at the cervicomedullary function. Some of the earlier lesions were less obvious whereas others had remained unchanged.

The prominent tuberculous arachnoiditis described by Wadia and Dastur^{1,2} can now be shown by the presence of gadolinium enhancement in the spinal dura-arachnoid complex on MRI. This was first shown in three out of the five cases reported by Chang *et al.*³ This was the only obvious abnormality on first MRI of the present patient. Without the serial studies he would have been defined as a straightforward case of spinal arachnoiditis.

The second and third scans showed multiple massive loculated lesions within the cervical cord. These were far too large for isolated tuberculomas. The radiological

features fit most closely with intramedullary abscesses. In particular the high signal on T2 weighted scans and the thick rim of enhancement correspond well with the known morphological features of pyogenic abscesses. Intracranial tuberculous abscesses can develop while patients are on antituberculous chemotherapy.⁴

Earlier medical literature has tended to separate tuberculous radiculomyelitis completely from syringomyelia secondary to chronic arachnoiditis. Appleby in 1969 perhaps first clearly established the link between delayed myelopathy after chronic meningitis and syringomyelia. More recently MRI studies have confirmed this link.⁵ The myelopathy in these cases is usually delayed by many years and gadolinium enhancement is not seen in the wall of these syrinxes.

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Bickerstaff's brainstem encephalitis associated with cytomegalovirus infection

Cytomegalovirus infection rarely induces encephalitis in a non-immunocompromised host. Bickerstaff's brainstem encephalitis was characterised by the acute onset of external ophthalmoplegia and cerebellar ataxia with CNS signs and symptoms.¹ We describe here the first case of Bickerstaff's brainstem encephalitis associated with cytomegalovirus infection.

A previously healthy 17 year old girl developed fever, headache, and general

fatigue. Ten days later, drowsiness, diplopia, and walking difficulty developed. She had low grade fever, drowsiness, and neck stiffness. The right eye showed esophoria. External ophthalmoplegia was severe in both lateral directions and moderate in the right upward direction. Triceps reflexes were absent and biceps reflexes were reduced. Finger to nose and heel to knee tests showed dysmetria and decomposition. Romberg's sign was negative. She had a broad based gait and tandem gait was impossible. Muscle strength and sensory function were normal. Blood count, liver function, and cold agglutinins were normal. Serum cytomegalovirus-IgM antibodies were negative and HIV 1 antibody was absent. Serum IgG anti-GQ1b antibody tests² were positive. A lumbar puncture showed a high pressure (300 mm H₂O), and high values for cell count, protein, albumin, IgG, IgA, and anticytomegalovirus antibodies (table). Cytomegalovirus DNA was detected in the CSF by the polymerase chain reaction. Brain CT and MRI were normal. Electroencephalography showed irregular slow waves with an enhanced and prolonged response to hyperventilation. Electromyography and sensory conduction studies were normal. The patient was treated with acyclovir (1500 mg/day) for seven days. Clinical symptoms disappeared as follows; low grade fever disappeared on the 17th day of illness, drowsiness and neck stiffness on the 22nd day, ataxia on the 24th day, and the eye movement disorder on the 30th day; deep reflexes returned to normal on the 36th day. On the 30th day of illness, euphoria with pathological laughing appeared; she burst out laughing when our eyes met or we began to talk and this lasted for seven days. Reduction of anticytomegalovirus antibodies and cytomegalovirus DNA by the polymerase chain reaction in the CSF paralleled the clinical improvement. The serum anti-GQ1b antibody titre was also decreased. The EEG abnormalities disappeared. Titres of antibodies against herpes simplex virus, varicella-zoster virus, and Epstein-Barr virus in both serum and CSF showed no significant changes.

Although brain MRI and CT showed no abnormalities, the rapidly developed ataxia, drowsiness, ophthalmoplegia without downgaze disturbance, and abnormal EEG findings suggested the presence of a CNS lesion.³ We therefore diagnosed the patient as having Bickerstaff's brainstem encephalitis.² Reduction of the increased CSF anticytomegalovirus antibodies and positive cytomegalovirus DNA paralleled the

Clinical data

| | Day of illness | | | | |
|--------------------------------------|----------------|------|------|------|------|
| | 21 | 28 | 42 | 56 | 76 |
| CSF: | | | | | |
| Cells (/mm ³) | 63 | 67 | 29 | 24 | 13 |
| (monocytes (/mm ³)) | (59) | (65) | (27) | (22) | (11) |
| protein (mg/dl) | 99 | 76 | 52 | 35 | 27 |
| IgG (mg/dl) | 9.6 | 8.0 | 4.9 | 2.8 | 2.8 |
| CMV-IgG-FA (normal <1 ×) | 1 × | 1 × | <1 × | <1 × | <1 × |
| CMV-IgG-ELISA (normal <200) | 596 | 485 | 304 | <200 | <200 |
| CMV-DNA-PCR | + | ND | ND | — | ND |
| Serum: | | | | | |
| IgG anti-GQ1b antibody (normal <400) | 6400 | ND | 3200 | ND | 1600 |
| CMV-IgG-ELISA | 3870 | 2700 | 3320 | 2270 | 3370 |

FA = indirect fluorescent antibody technique; ELISA = enzyme linked immunosorbent assay; ND = not done; PCR = polymerase chain reaction; CMV = cytomegalovirus.

improvement in neurological symptoms, indicating that the cytomegalovirus infection was associated with her neurological disease. The aetiology of Bickerstaff's brainstem encephalitis is still unclear. A relation with herpes simplex virus infection has been noted,⁴ but no patients with Bickerstaff's brainstem encephalitis associated with cytomegalovirus infection have been reported.

With regard to the pathogenesis of Bickerstaff's brainstem encephalitis, an immune mechanism has been considered.¹⁻⁴ In this patient, the presence of serum anti-GQ1b antibody, which is common in Fisher's syndrome,⁵ indicated that humoral auto-immune mechanisms, common to Fisher's syndrome, function in the development of Bickerstaff's brainstem encephalitis.¹ The typical signs of meningoencephalitis—namely, fever at the onset of neuritic symptoms, meningeal irritation and CSF pleocytosis—and detection of cytomegalovirus DNA in the CSF may indicate the involvement of cytomegalovirus infection. Both cytomegalovirus infection and a post-infection autoimmune mechanism may have caused clinical symptoms in this patient.

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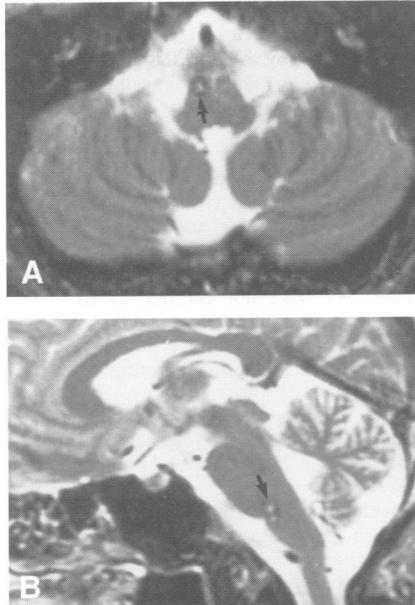
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Raymond syndrome (alternating abducens hemiplegia) caused by a small haematoma at the medial pontomedullary junction

Raymond syndrome¹ is characterised by ipsilateral abducens nerve palsy and contralateral hemiplegia. Pure Raymond syndrome is extremely rare, as many nuclei and fibres exist near the root fibres of the abducens nerve. This is the first report in which the precise localisation of a pure form of Raymond syndrome was determined by MRI.

A 39 year old man awoke with horizontal diplopia, especially on right lateral gaze. Five days later, a Hess chart examination performed by an ophthalmologist showed paresis of the right lateral rectus muscle. On admission 19 days after onset, the patient showed a mild paresis of the right abducens



(A) Axial (TR = 2500 ms, TE = 110 ms) and (B) sagittal (TR = 2500 ms, TE = 100 ms) MRI sections. There are two small high signal intensity spots surrounded by low signal intensity areas at the right pontomedullary junction (arrows).

nerve and a subtle weakness of his left leg with moderate hyper-reflexia in the left upper and lower limbs. The Babinski reflex was positive and the abdominal reflex was absent on the left side and the Babinski reflex was negative and the abdominal reflex was positive on the right side. No facial weakness or deviation of the tongue on protrusion was found. All other general and neurological examinations were normal. Routine blood and urine examinations were normal. Evaluations of short latency somatosensory evoked potentials to posterior tibial nerve stimulation, brainstem auditory evoked potentials, and blink reflex proved normal. Head CT was normal, but a brain MRI done 31 days after onset showed two punctate high signal intensity spots surrounded by low signal intensity areas at the medial pontomedullary junction on both the T1 and T2 weighted images (figure). Vertebral angiography showed no abnormality. Thus the lesion was probably produced by a haemorrhage from a cavernous haemangioma at the pontomedullary junction.

Both Millard-Gubler syndrome (facial palsy and contralateral hemiplegia) and Raymond syndrome are well known to induce crossed paralysis due to a caudal pontine lesion. The pure form of either syndrome has, however, rarely been reported. The lesion producing the pure Millard-Gubler syndrome² is located more laterally than seen in our patient, whereas that producing isolated abducens nerve palsy³ is located more dorsally. As the haemorrhage was restricted to the ventral and medial pons, our patient was considered to show pure Raymond syndrome.

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Pupillary dilatation and arm weakness as negative ictal phenomena

Transient ictal hemiplegia is an uncommon feature of epileptic attacks that were classified by Gastaut and Broughton as unilateral atonic seizures.¹ The present case was of particular interest because hemiplegia was accompanied by dilatation of the pupil on the side of the hemiplegia.

A boy aged 9 years had a history of episodic weakness of his left upper and lower limbs, sometimes preceded by a sensation "like a dog shivering" in his head since the age of 5. His mother said that he would stare and his left arm then dropped limply to his side while his left leg became weak for about 10 to 40 seconds. During this time his left pupil dilated. In some episodes his left eyelid fluttered and the left side of his mouth turned up and his left arm and leg remained weak. The attacks increased in frequency until he was having two to eight each day, but subsided to once daily when carbamazepine treatment was started. There was no history of head injury or other relevant illness and no family history of epilepsy. His EEG showed an almost continuous sharp and slow wave discharge arising in the right parietal region. Brain CT was normal but MRI four years later showed a hyperintense area involving both grey and white matter in the right parietal lobe; there was no mass effect or evidence of blood products surrounding the lesion.

At the age of 13 he underwent craniotomy and electrocorticography confirmed the presence of an epileptic focus in the area surrounding an atrophic gyrus in his right parietal cortex. The abnormal area was then excised. The histology report (Dr W A Evans) concluded that "I find this lesion hard to classify. It is most likely a hamartoma, possibly of a similar nature to the focal dysplasia of the cerebral cortex described by Taylor *et al*".²

There was no postoperative neurological deficit and he was free of seizures until eight months later when his carbamazepine dosage was reduced from 1000 mg to 400 mg daily. Three years after the operation his carbamazepine dose was again reduced, when he had a recurrence of daily attacks of fluttering of his left eyelid and weakness of his left arm, but not the left leg, lasting 20 seconds. His EEG showed focal right parietal slow activity without epileptogenic features. Since then he has been subject to episodes about every 10 days with dilatation of the left pupil, weakness of the left arm, and some twitching of the left side of his face lasting about 10-20 seconds. He has never had any jerking or involuntary movement of his left arm.

Constriction of the left pupil in association with hallucinations projected into the left visual field was reported as an ictal phenomenon by Lance and Smee and