
Letters

Midbrain locked-in state with oculomotor subnucleus lesion

Sir: The term locked-in syndrome had been synonymous with the ventral pontine syndrome until Karp1 reported a locked-in state caused by bilateral midbrain infarcts. Subsequently, three cases of the syndrome were reported in association with midbrain lesions. We present here a midbrain locked-in state with a characteristic ocular finding.

A 60-year-old man with a 5 year history of hypertension and diabetes mellitus had been suffering from recurrent vertigo since 29 March 1983. On 13 April, unsteadiness of gait developed and persisted for 2 weeks. On 12 August, he noticed weakness of the extremities. He became quadriplegic and anarthric on 15 August and was admitted to our hospital. On admission he was anarthric but fully conscious communicating with us by blinking or by some eye movements. Neurological examination revealed mild memory disturbance, slight conjugate eye deviation to the left, rightward gaze palsy, paralysis of bilateral upward eye movements, paralysis of VII, IX–XII cranial motor nerves and flaccid quadruplegia with bilateral extensor plantar response. Torsional eye movements were normal. Oculovestibular or oculocephalic reflex revealed a full eye deviation in the horizontal plane but no upward deviation. Bell’s phenomenon was absent. Cranial CT scan demonstrated low density areas located in bilateral cerebral peduncles, mesencephalic tegmentum, occipital lobes (fig a), and left cerebellar hemisphere sparing the ventral pons (fig b). Left vertebral angiography revealed occlusion of the basilar artery at its origin. One month after admission, faint vocal and some motility of head and left leg appeared. He is now gradually improving.

The classical locked-in syndrome identified by Plum and Posner2 is a clinical state consisted of full consciousness and of total immobility except for vertical eye movements and blinking. However, there have been some clinical variations of ocular motility in subsequently reported cases. The cases with eye movements except for those originally reported should be termed locked-in state. Karp1, Dehaene3, Forti4, and Kobayashi5 described the locked-in states caused by midbrain lesions. Full voluntary control of all eye movements was preserved in the cases of Karp and Kobayashi in which the lateral two-thirds of the cerebral peduncles were involved in infarcts. On the other hand, Dehaene’s case with horizontal gaze paralysis had infarcts located in the bilateral medial cerebral peduncles. The difference in horizontal ocular motility depends on the anatomical evidence that the horizontal gaze pathways are situated in the medial parts of the cerebral peduncles. Our patient’s gaze palsy may be due to the lesion of left medial cerebral peduncle. The characteristic ocular finding of our case was the paralysis of upward eye movements. Recently, much attention has been paid to the rostral interstitial nucleus of the medial longitudinal fasciculus as an organising centre of upward gaze. In the present case, however, absence of upward deviation in Bell’s phenomenon, oculovestibular and oculocephalic reflexes was indicative of nuclear or infranuclear oculomotor paralysis rather than upward gaze paralysis. Normal extorsion of the eyes indicated normal inferior oblique function. Crossed innervation of the superior rectus muscle was first investigated by Warwick.4 Clinically, Pierrot-Deseilligny6 indicated that a lesion in the unilateral oculomotor nucleus resulted in not only ipsilateral oculomotor paralysis but also contralateral paralysis of superior rectus function. Bienfang7 assumed that a lesion in the unilateral superior rectus subnucleus could cause a

Fig (a)  CT scan showing low density areas in the bilateral cerebral peduncles (long arrow), left mesencephalic tegmentum (short arrow), and bilateral occipital lobes. (b) CT scan showing normal upperpons and low density area in the left cerebellar hemisphere.
bilateral paralysis of superior rectus function based on his experimental result that the axons from the superior rectus subnucleus of one side crossed the midline and passed through the same subnucleus of the other side. However, isolated paralysis of the bilateral superior recti found in our case is a rare ocular finding. To our knowledge, no other case has been reported in the literature. The above anatomical and clinical evidence and CT findings suggest that the most probable lesion in our case is the superior rectus subnucleus in the left oculomotor complex.

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References


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Fluid chronic epidural haematoma: a rare complication of ventriculo-peritoneal shunt.

Sir: Epidural haematoma is a rare complication of ventricular shunting procedures. Fluid chronic epidural haematoma occurring after a shunt has not been described before. Recently, we encountered such a case following a ventriculo-peritoneal shunt in a young adult with obstructive hydrocephalus due to a giant-cell astrocytoma of the third ventricle. CT scanning was invaluable in achieving prompt diagnosis and treatment.

A 16-year-old right-handed student was admitted with a three month history of morning headaches and rapid deterioration of vision one week prior to admission. Examination showed multiple hairy moles all over the body, but no overt stigmata of tuberose sclerosis. The boy was intelligent, alert and orientated. Bilateral sixth nerve palsies and severe papilloedema were evident. Visual acuity was markedly diminished to 6/24 in both eyes. There was otherwise no motor or sensory impairment. Blood chemistry was normal. Skull radiographs showed erosion of the sella turcica. Computerised scanning with contrast demonstrated a large contrast-enhancing tumour at the foramen of Monro, projecting upwards and more to the left (fig a and b). Marked dilatation of both lateral ventricles was evident.

Because of rapid deterioration in vision, a ventriculo-peritoneal shunt was performed as an emergency through a right posterior parietal burr-hole. A medium pressure (60 to 90 mm of water) Holter valve was used. Intraoperatively, about 20 ml of cerebrospinal fluid were collected for biochemical and cytological analysis. Following the shunt there was rapid relief of headache and improvement of visual acuity. On the eleventh post-operative day, he noticed recurrence of headache. Examination revealed a left homonymous hemianopia and a mild left hemiparesis. Computerised scanning with contrast was repeated which demonstrated a large hypodense biconvex extracerebral collection with an enhancing inner membrane. It was situated at the right frontal parietal region, anterior to the burr hole site (fig a).

A large epidural dark brown fluid collection measuring about 150 ml was evident at operation. The dura was adherent to the site of the previous posterior parietal burr-hole, and no bleeding point could be identified. Complete drainage of the fluid haematoma was achieved via a small craniectomy measuring 4 × 4 cm. He made a prompt recovery after the drainage of the haematoma. One week later, he underwent a left fronto-transventricular approach for a subtotal removal of the tumour, which proved to be a giant cell astrocytoma. He was subsequently discharged from hospital with no neurological deficit.

Subdural haematoma is a known complication of ventricular shunts. Its incidence is reported to vary from 0·4 to 1·2 percent.1 In contrast, epidural haematoma is extremely rare. On reviewing the literature, we found only three reports describing this complication following valve regulated internal shunts.2–4 The details of these cases are summarised in the table. These haematomas occurred mostly in young adults. The underlying cause was chronic obstructive hydrocephalus in each case. The haematoma was usually distant to the site of burr-hole with predilection for the anterior half of the cranial vault. Presentation varied from 6 hours to three weeks after the insertion of the shunt.

The mechanism of formation of epidural haematoma in these situations is unclear. It is generally believed to be the result of sudden lowering of the intracranial pressure after ventricular decompression. This may create a potential space between the dura and the inner table of the skull with subsequent bleeding. An unusual feature in our case is the presence of liquid blood, which was apparent both on the CT scan (hypodense) and also at operation. Although chronic subdural haematoma is almost always fluid, it is extremely rare to find fluid in a chronic epidural haematoma. Jameison stated that secondary liquefaction in a chronic epidural haematoma is virtually unknown.5 However, three such cases have been recorded,6–8 but no satisfactory explanation on its pathophysiology has been offered. In chronic subdural haematoma, the mechanism of formation is believed to be related to increase fibrinolysis and repeated microhaemorrhages within the haematoma capsule.6 The inner layer of the dural mater is important in squiggle.6 Conversely, it can be argued that in the case of a chronic epidural haematoma, the outer layer of the dura mater does not have the same potential for cellular organisation, so secondary events leading to liquefaction of the haematoma does not occur to the same extent. The role of cerebrospinal fluid remains controversial.11–12 In our case, CSF might have leaked around the ventricular catheter into the epidural space, thus forming a mixed hygroma and haematoma, and epidural hygroma associated with a small clot and a
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