Short report

Ocular bobbing and myoclonus in central pontine myelinolysis

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SUMMARY Ocular bobbing and myoclonus were unusual findings in a patient with central pontine myelinolysis. Pathological findings confirmed the diagnosis of pontine and extrapontine myelinolysis.

Central pontine myelinolysis is the most likely diagnosis when a patient with a background of metabolic, nutritional and electrolyte disorders develops a quadriplegia and pseudobulbar signs over several days. The authors describe a patient with central pontine myelinolysis who presented with diffuse myoclonic jerks and ocular bobbing.

Case report

A 55-year-old man with a history of longstanding excessive alcoholic intake was admitted because of weakness. On initial examination he appeared pale, disoriented and his gait was unsteady and widebased. Muscle strength was normal, reflexes were symmetrical and plantar reflexes were flexor. Serum sodium was 110 mmol/l, serum osmolality was 225 mosm/l with a urine osmolality of 430 mosm/l. Liver function was normal. The electrolyte values were corrected within five days by moderate water restriction and his mental state improved. On the 5th day, bronchopneumonia was diagnosed. On the 6th day, a slight left facial weakness and a prominent jaw jerk were noted. He choked on food on several occasions. Gross proximal and distal myoclonic jerking affected his limbs and head synchronously. The jerking was provoked by any attempt of voluntary movement, by eliciting tendon reflexes, by stretch applied to fingers or by sudden noise. The myoclonus always diffusely involved the head and limbs. It was not provoked by touch or pinprick. Clonazepam 1 mg IV abolished the jerking for about two hours. Lumber puncture showed normal cerebrospinal fluid (CSF), EEG showed a diffuse slow rhythm of the θ range with intermittent spikes with variable temporal relationship to the jerks. The patient became severely dysarthric and unable to swallow. A tracheostomy was performed. Gross myoclonic jerking persisted over six days. On the 15th day, he was stuporous and responded to command with eye opening only. Oculocephalic testing showed minimal abduction of the left eye but was otherwise normal. After horizontal head rotation both eyeballs made conjugate brisk downward movements, about 4 mm in excursion, with slow drift back to the horizontal meridian either immediately or after several seconds delay. The rate was unpredictable and never exceeded a frequency of about 15 per minute. The next day oculococular testing showed complete absence of reflex lateral eye movements with normal vertical movements. Caloric stimulation enhanced the bobbing eye movement with full downward excursion. Corneal reflexes were absent, there was a flaccid quadriplegia. Ocular bobbing was observed over seven days. The patient died on the 25th day.

Post-mortem findings

The leptomeninges were normal. There was minimal atherosclerosis, and in particular, the basilar artery was normal. Coronal sections revealed the integrity of both hemispheres, in particular, the corpus callosum and the mammillary bodies were normal. There was extensive softening of the pons extending to the junction with the medullar oblongata. Histologically, the main lesions were located in the pons which was extensively demyelinated, with the preservation of a narrow rim at its superficial margin (fig). The demyelinated area was invaded by numerous fat-filled "gitter-cells". Many neurons remain intact in the demyelinated area but silver impregnation demonstrated an important axonal destruction with some varicose axons at the border of the lesion. No oligodendrocytes were seen. The corpora mammillaria were intact: no glio-mesenchymal proliferation was noticed. In addition to the...
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central pontine myelinolysis there was a symmetrical demyelination of both anterior columns of the fornix close to their termination in the mammillary bodies. This was well seen on frozen sections as the lesion was packed by many phagocytes containing sudanophilic lipids. Patchy demyelination was also present in the white axis of some cerebellar folia. The Purkinje cells and granular cells were intact in these folia.

Discussion

Central pontine myelinolysis was the most likely diagnosis as the patient with a background of chronic alcoholism developed a quadriplegia and pseudobulbar signs over several days after correction of hyponatraemia.1 The pathological findings in the pontine and extrapontine structures were typical of central pontine myelinolysis.2-4 Ocular bobbing is not mentioned in reviews on central pontine myelinolysis. Recent work on ocular bobbing2-7 does not include central pontine myelinolysis in the growing list of associated clinical conditions. Ocular bobbing has been described mainly, though not exclusively, in intrinsic pontine lesions, so its occurrence in central pontine myelinolysis is hardly surprising. It was a prominent feature in this patient, lasted for several days and appeared shortly before complete paralysis of lateral reflex eye movements. Ocular bobbing usually appears after complete paralysis of voluntary and reflex lateral eye movements with lesions involving the paramedian pontine reticular formation.

Myoclonic jerking occurred early after disease onset. There was diffuse jerking of the limbs and the head in response to stimuli of various nature. Regardless of the limb stimulated, the myoclonic response was elicited in all susceptible sites and was reduced after injection of clonazepam. It thus had the clinical features of reticular reflex myoclonus.6 The patient did not sustain clinically detectable hypoxia and the jerking disappeared with worsening of quadriplegia. The precise mechanism of the genesis of this myoclonus is unknown, but we speculate that it was related to involvement of the pontine reticular formation.

References