Ocular bobbing and opsoclonus

Two abnormal spontaneous eye movements occurring in the same patient: case report

H. G. BODDIE

From the Department of Neurology, North Staffordshire Hospital Centre, Stoke-on-Trent

SUMMARY A case in which two rare abnormal spontaneous eye movements, ocular bobbing and opsoclonus, were observed, is reported. Their pathophysiology and distinction from other abnormal spontaneous eye movements are discussed.

Ocular bobbing is a term first used by Fisher (1961) to describe distinctive, abnormal, spontaneous eye movements occurring in comatose patients with paralysis of horizontal conjugate gaze. The movements consist of an abrupt conjugate downward jerk of the eyes followed by a slow return to the mid position. The usual cause is a pontine haemorrhage or infarction. Since then, additional reports have further defined the clinical spectrum of ocular bobbing (Fisher, 1964; Daroff and Waldman, 1965; Hameroff, Garcia-Mullin, and Eckholdt, 1969; Nelson and Johnston, 1970; Susac, Hoyt, Daroff, and Lawrence, 1970).

Opsoclonus is a term first used by Orzechowski and Walichiewicz (1913) and Orzechowski (1927) to describe constant, conjugate, chaotic agitation of the eyes seen in patients with a non-epidemic form of encephalitis. The movements were characterized by periods of violent ocular mobility alternating with periods of relative stillness; they were present during eye closure and sleep; they were not rhythmical and occurred in the horizontal and vertical planes. Associated abnormalities were myoclonic movements of the face and limbs, and cerebellar incoordination. Since the original description, additional reports have further delineated what appears to be a distinct entity (Marmion and Sandilands, 1947; Cogan, 1954, 1968a; Smith and Walsh, 1960).

The occurrence of ocular bobbing and opsoclonus in the same patient has not been reported previously. This paper presents such a case, and

discusses briefly the pathophysiology of ocular bobbing and opsoclonus, and their differentiation from other abnormal spontaneous eye movements.

CASE REPORT

A 39 year old female was admitted as an emergency on 19 September 1970. Essential hypertension had been diagnosed six months previously, and outpatient follow-up showed satisfactory control of blood pressure on propranolol 20 mg t.d.s. For six weeks before admission she had noticed intermittent headaches of an indeterminate nature. On the day of admission she had complained of severe frontal headaches and had collapsed at work.

On admission she was unconscious with no response to painful stimuli; the blood pressure was 140/110 mm Hg; respirations were of the Cheyne-Stokes pattern. General examination was otherwise normal. There was no neck stiffness and the fundi were normal. The pupils were 3 mm in diameter, equal and reacting briskly to direct light, the consensual light reflex being normal. Corneal reflexes were absent. There was a left-sided hypertonia and hyperreflexia with bilateral extensor plantar responses.

The most striking abnormality present was the disturbance of ocular movements known as ocular bobbing, brisk conjugate downward jerks of the eyes followed by a slow return to the mid position at a rate of two to four per minute. No spontaneous horizontal eye movements were present, nor did caloric stimulation (ice-water at 0° C) induce reflex eye movements, or influence the rate of bobbing. The oculocephalic reflex was absent.

INVESTIGATIONS Complete blood counts, urinalysis, serum electrolytes, protein electrophoresis, calcium, phosphate, alkaline phosphatase, cholesterol, transaminases, WR and Kahn tests, 24 hour urinary urea and electrolytes, creatinine clearance, skull, and chest radiographs were normal. An electrocardiogram showed changes consistent with pre-existing hypertension. Lumbar puncture showed a uniformly bloodstained cerebrospinal fluid under a pressure of 150 mm water. Xanthochromia was present; total protein content was 80 mg/100 ml. The CSF contained 3 nucleated cells/cu. mm with 9,400 red blood cells/cu. mm. An electroencephalogram on the day of admission was reported as follows (Dr. Rosemary Cooper): there was a symmetrical 8–10 Hz dominant post-central rhythmic activity with forward spread. There was a moderate amount of background slow activity in the theta range with some delta components seen diffusely but maximally posteriorly. There was no evidence of asymmetry of frequency or amplitude. The patient was unconscious at the time of recording and showed no EEG or behavioural change to arousal stimuli. Conclusion: this is an interesting record with its total disassociation of the clinical and EEG state. The presence of a symmetrical dominant post-central rhythm in the alpha range with this level of consciousness strongly suggests a pontine (tegmental) disturbance. The presumptive diagnosis was that of a pontine haemorrhage secondary to hypertension. Contrast radiographic studies were not done.

During the first 72 hours after admission, the patient remained comatose and the ocular bobbing persisted at a rate of two to four per minute. During the fourth day after admission, her level of consciousness lightened, and the ocular bobbing decreased in frequency, and finally disappeared on the fifth day. She remained in a steady state up to December 1970, since when there has been a slow but steady improvement to reach her present clinical state, which was achieved in May 1971. She is mentally alert and oriented with a marked cerebellar dysarthria and ataxia in all four limbs, predominantly left-sided. The hyperreflexia persists on the left, but both plantar responses are flexor. Myoclonic movements of the face, palate, platysma, sternomastoids, and upper limbs are present. These movements are irregular, not rhythmical and occur at a rate varying between 80 and 120 per minute. Opsoclonus—that is, constant, conjugate, chaotic movements of the eyes—is present predominantly in the vertical plane but also with some movement in the horizontal plane. Its rate varies from 80 to 180 per minute; the movements are present with eye closure and persist during sleep, although their frequency and severity are diminished. They are not

synchronous with the associated myoclonic movements. Despite the severe neurological deficit, the patient went home in May 1971, and when seen in outpatients in October 1971, the signs were unchanged.

DISCUSSION

In the past 25 years a variety of rare abnormal spontaneous eye movements have been increasingly reported in disorders of the brain-stem, cerebellum, and their connections. In some instances their recognition has been of precise localizing value, but in others has led to semantic confusion. The spectrum includes, besides ocular bobbing and opsoclonus, down-beat nystagmus, ocular myoclonus, 'lightning eye movements', ocular dysmetria, and ocular flutter.

Cogan (1968b) defined down-beat nystagmus as nystagmus in which the fast component is downwards. Eight of his 27 cases were associated with radiological evidence of congenital anomalies at the level of the foramen magnum, with or without the Arnold-Chiari malformation. The diagnosis in the rest covered a miscellany of conditions which had in common symptoms and signs suggestive of a lesion in the pons, medulla or cerebellum. It is readily distinguished from ocular bobbing by its regularity, more rapid rate, and the associated clinical picture.

Ocular myoclonus describes the ocular movements seen in palato-pharyngo-laryngo-oculodiahphragmatic myoclonus. These movements are pendular, rhythmical, and synchronous with the myoclonic movements of associated structures. Yap, Mayo, and Barron (1968) using a polygraphic recording technique showed this to be true but nevertheless described the movements as 'ocular bobbing', thus leading, as pointed out by Susac *et al.* (1970) to predictable confusion. Atkin and Bender (1964) described three patients in whom abnormal eye movements were seen, which they called 'lightning eye movements'. These were characterized by rapid bursts of horizontal to-and-fro eye movements, the bursts consisting of small conjugate saccades. Although there was no evidence of myoclonic movements in associated structures, they used the term ocular myoclonus synonymously with 'lightning eye movements'.

In ocular dysmetria, overshooting of the eyes

on attempted fixation is followed by several cycles of diminishing amplitude until precise fixation is achieved. Both this and ocular flutter, the occurrence of intermittent to-and-fro oscillations of the eyes lasting a few seconds which is often spontaneous and precipitated by changes in fixation, are seen primarily in cerebellar disease. Their differentiation from the abnormal spontaneous ocular movements under discussion has not given rise to difficulty.

Opsoclonus—that is, constant, conjugate, chaotic movement of the eyes, has been considered by authors to be a distinctive abnormal spontaneous eye movement seen in encephalitis affecting the brain-stem. The clinical syndrome consists of the onset of a mild non-specific infection accompanied by fever and malaise, which is followed about 10 days later by opsoclonus, cerebellar signs, and myoclonic movements of the face, neck, and upper limbs. Sometimes there is a pleocytosis in the cerebrospinal fluid. The prognosis is good, complete recovery in two months occurring in most cases. However, the fact that opsoclonus, although most commonly seen in the syndrome described, has been reported in other diseases of the central nervous system (Halliday, 1967) limits its usefulness with regard to the underlying pathology. More recently its association with malignancy has been reported (Ross and Zeman, 1967; Solomon and Chutorian, 1968).

Typical ocular bobbing—that is, abrupt conjugate, downward jerking of the eyes followed by a slow return to the mid position and accompanied by paresis of horizontal conjugate gaze—is a valuable clinical sign which is easy to define and recognize. Its importance lies in the fact that the available pathological evidence indicates that the underlying lesion is almost always pontine, and commonly a haemorrhage or infarct. Susac *et al.* (1970) reviewed the literature on 25 reported cases, of whom four survived. Of the 21 who died, post mortem studies were performed on 18, the cause of death being pontine haemorrhage in nine; two cases each of pontine infarction and cerebellar haemorrhage; one case each of pontine glioma, post-traumatic lesions of cortex, brain-stem, and cerebellum, haemorrhagic infarction of cerebellum, metabolic encephalopathy, and a patient with presumptive vertebro-basilar ischaemia.

In their clinical experience of nine cases they added a further dimension to the syndrome by classifying the cases of ocular bobbing as (1) 'typical'—abrupt, spontaneous downward jerks of the eyes with a slow return to the mid position associated with paralysis of horizontal conjugate eye movements; (2) 'monocular' (or parietic)—reflecting a coexisting unilateral third nerve paresis in addition to the bobbing; and (3) 'atypical'—either a variation unexplained by associated oculomotor palsy, or bobbing with intact spontaneous or reflex horizontal eye movements. Since their report, ocular bobbing has also been reported in a case of Leigh's necrotizing encephalomyopathy (Borit, 1971).

The diffuse and varied nature of the underlying pathological processes in the post mortem studies of cases of opsoclonus and ocular bobbing have effectively rendered any precise assessment of the pathophysiology uncertain. The patient described here presented with typical ocular bobbing and subsequently developed opsoclonus. The available clinical evidence at the time of admission (pre-existing hypertension and haemorrhagic cerebrospinal fluid), suggested that there had been an intrapontine or cerebellar haemorrhage. The development in sequence of these abnormal eye movements suggests that these two, at least, have a common pathological background capable of producing one or other of these movements.

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