Opsoclonus

Pattern of regression in a child with neuroblastoma

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Opsoclonus, although a rare disorder of ocular motility, presents such a dramatic clinical picture that the condition should be easily recognized by ophthalmologists and neurologists. The spontaneous semi-conjugate eye movements are grossly chaotic, rapid, of large amplitude and random direction, and continue during eye closure or sleep. Of primary importance is the relationship of opsoclonus to occult neuroblastoma in childhood, and the recognition of this association may be life-saving.

We have recently had the opportunity to observe an infant with opsoclonus and neuroblastoma, and have cinematographically documented the regression of the eye movement disorder after removal of the tumour. The pattern of this regression and the description of an unusual head-thrust movement are the subjects of this report.

Case report

A 1-year-old girl developed unsteadiness when standing, during the month before examination. Five days before she was admitted to hospital, both parents noted the appearance of peculiar spontaneous eye movements. Examination performed by Dr Stuart Brown, at the University of Miami, revealed an irritable child with involuntary jerking of limbs, head and trunk, ataxia, and wildly chaotic, random eye movements. A left-sided abdominal mass was detected, and proved to be a large retroperitoneal tumour; this mass was totally removed and histopathologically diagnosed as neuroblastoma.

In the immediate postoperative period the opsoclonus persisted, but during the following 10–14 days regression was evident. The chaotic continuous movements were replaced by flutter-like bursts of saccades accompanied by a characteristic head shudder. These eye movements were separated by quiet intervals which became progressively longer and more frequent.

During the third postoperative week the parents noted occasional violent thrusting of the child's head. Examination

revealed that such head-thrusting occurred exclusively upon viewing targets to the left. Stop-frame analysis of the movement (Figure) showed that the eyes first made a leftward saccade, initiating the movement, with the head following, but violently overshooting the end-point. A controversial eye movement ended the thrust.

The child's overall condition improved so that 8 weeks after surgery she could sit up, although she still exhibited head and truncal ataxia. Gaze to the left was accomplished smoothly without thrusting, and all flutter and opsoclonus had ceased, although occasional dysmetric eye movements could be detected.

Discussion

Sixty years ago Orzechowski (*cited* 1927) first used the term 'opsoclonus' to describe this particular eye movement disorder. Subsequent reports stressed the benign nature of the condition and the complete recovery of those afflicted (Smith and Walsh, 1960; Cogan, 1968). Although Foster Kennedy described 'gross nystagmoid movements of the eyeballs' in a child with neuroblastoma (*cited* by Cushing and Wolbach, 1927), it was not until later that Solomon and Chutorian (1968) postulated a direct link between neuroblastoma and opsoclonus.

Opsoclonus in children is most frequently due to encephalitis. However, a significant number of cases will harbour neuroblastomas, and this association must be recognized because of its great clinical significance. In adults, on the other hand, malignancy with opsoclonus is exceedingly rare (Alessi, 1940; Ross and Zeman, 1967; Ellenberger, Campa, and Netsky, 1968), the great majority of cases being attributable to a viral encephalitic process.

In children with neuroblastoma, even with successful treatment, a large percentage will have permanent neurological sequelae, while only a few will recover completely (Senelick, Bray, Lahey, Van Dyk, and Johnson, 1973). In the encephalitic group the reverse is the case, and full recovery is the rule in the majority of patients (Weiss and Carter, 1969). In encephalitic adults full recovery is anticipated, while in the rare cases associated with neoplasms, the prognosis is that of the primary tumour.

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FIGURE Stop-frame analysis of head-thrusting on looking left. Conjugate eye movement leads the head-thrust, which overshoots with such velocity that the head violently strikes the side of the push-chair

Neuroblastoma is responsible for most paediatric malignancy mortalities, except the acute leukaemias, central nervous system cancer, and lymphoma (Miller, 1969). However, neuroblastoma also has a high rate of spontaneous regression (Everson and Cole, 1966). Before the report of Solomon and Chutorian (1968) opsoclonus was not recognized as being associated with neuroblastoma, but several

authors have since suggested that many children with this syndrome may harbour silent neuroblastomas (Bray, Ziter, Lahey, and Myers, 1969). The question arises, how many of the previously reported instances of opsoclonus in infancy were actually associated with an occult, spontaneously-regressed neuroblastoma. It has been shown unequivocally that opsoclonus and other cerebellar signs regress, with or without the benefit of steroid therapy, for months or years before the discovery of a tumour (Moe and Nellhaus, 1970; Larbre, Bethenod, Guibaud, Mamelie, and Genoud, 1972; Senelick and others, 1973).

In the present case, after removal of a neuroblastoma, the opsoclonus regressed through phases of *ocular flutter* and then *dysmetria*. Considering this pattern of eye movement regression in the light of histological reports in patients with opsoclonus (Ross and Zeman, 1967; Ellenberger and others, 1968; Senelick and others, 1973), it would seem that the ocular motor phenomena are surely related to cerebellar dysfunction. We agree with Ellenberger, Keltner, and Stroud (1972) that opsoclonus, flutter, and dysmetria are a continuum of ocular motor disturbances related to cerebellar disease. However, Keane and Devereaux (1974) have recently reported a case of opsoclonus associated with hemispherical glioblastoma which extended into the upper mesencephalic tegmentum, yet spared the cerebellum; they suggested that the upper brainstem may be the critical area for lesions producing opsoclonus.

Our patient also exhibited a most peculiar unilateral head-thrusting which was not apparently related to saccadic ocular palsy, since stop-frame ciné analysis revealed that the eyes lead the horizontal head-thrust (see Figure). Conjugate eye movement occurring before head movement is opposite to what is seen in the classically described instances of Cogan's congenital ocular motor apraxia (Cogan, 1952). We interpreted this head-thrust as indicating a *dysmetric head movement* due to cerebellar disease rather than being caused by an ocular motor abnormality. We cannot explain why the head movement was unilateral and occurred during the recovery phase of the ocular motor disturbance.

Summary

A child with opsoclonus associated with occult neuroblastoma is presented, in whom regression of the eye movement disorder through phases of flutter and dysmetria was observed. It is speculated that these ocular motor abnormalities represent a continuum of cerebellar dysfunction. A peculiar dysmetric head movement not related to saccadic palsy is documented.

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