Opsoclonus
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DESCRIPTION
An Iranian boy aged 15 years presented with a few day’s history of abnormal gait and sudden repetitive jerky limbs’ movements. The past histories were unremarkable. He was fully conscious. Examination revealed ataxia of stance and gait and generalised myoclonic jerks. Ocular examination revealed opsoclonus (video 1). Routine blood tests and brain MRI with gadolinium were normal. The boy’s father stated that he intends to go back to Iran and manage his son there. In Iran, brain MRI, chest/abdomenopelvic CT scans, paraneoplastic autoantibodies panel and CSF analysis were unremarkable. The boy was diagnosed with idiopathic opsoclonus; partially improved on intravenous immunoglobulin.

Opsoclonus is an ocular dyskinesia. There are sudden, involuntary, chaotic, arrhythmic and multidirectional (upwards, downwards and torsional) conjugate saccadic ocular movements. The resulting symptoms of oscillopsia and visual blurring are attributed to their large amplitude and high frequency. Opsoclonus must be differentiated from nystagmus (rapid jerks followed by a slow corrective saccade) and ocular flutter (horizontal back-to-back saccades). Frequently, generalised myoclonus and ataxia coexist; hence, the term opsoclonus-myoclonus-ataxia.

Paraneoplastic (neuroblastoma in children; small-cell lung cancer in adults), parainfectious, metabolic and toxic aetiologies can be identified; however, it can be idiopathic. The cell-mediated and humeral immunities contribute to the pathogenesis of the paraneoplastic and idiopathic varieties. Although it has been associated with several autoantibodies, several patients are seronegative. The treatment is directed towards the underlying cause; tumour surgery, chemotherapy and immune suppressive therapy, including intravenous immunoglobulin. Clonazepam may be a useful symptomatic treatment in those who did not respond to immune suppressants.1–3

Learning points
▸ Opsoclonus should be differentiated from nystagmus and ocular flutter.
▸ Opsoclonus may be associated with neuroblastoma in children and small-cell lung cancer in adults, as a paraneoplastic manifestation.
▸ Frequently, opsoclonus is seen with ataxia of stance and gait as well limbs and truncal myoclonus jerks.

Video 1 The patient was asked to look forward. Note the repetitive, involuntary, high-amplitude, chaotic and multidirectional conjugate eye movements; these represent opsoclonus (dancing eyes; saccadomania). Opsoclonus is present during fixation, pursuit movements and convergence. In addition, it persists during sleep or eyelid closure. Frequently, limbs and truncal myoclonus as well as ataxia of stance and gait coexist; therefore, the term opsoclonus-myoclonus-ataxia was coined. The term dancing eyes-dancing feet is also sometimes used.

REFERENCES