CASE REPORT

Postintervention acute opsoclonus myoclonus syndrome

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ABSTRACT

Opsoclonus myoclonus syndrome (OMS) is a rare, neurological condition affecting 1 in 10 000 000 people annually. Opsoclonus, defined as involuntary rapid, multivectorial oscillations of the eyes, together with ataxia and myoclonus are usually present. OMS may be paraneoplastic: often associated with occult neuroblastoma in childhood and with breast carcinoma or small cell lung carcinoma in adults. Other aetiologies include viral or toxic agents. The pathogenesis is thought to be immune mediated. A 37-year-old woman with previous inflammatory cranial mononeuropathies was admitted for elective dilatation and curettage (D&C). Immediately after she complained of left-sided paraesthesia and later became disoriented, with incoherent speech, inability to obey commands, opsoclonus of the eyes and myoclonic jerks. Investigations including onconeural antibodies, cerebrospinal fluid analysis, and imaging were normal. She was treated with intravenous methylprednisolone with rapid improvement. Previous surgeries with anaesthesia were uncomplicated. The anaesthetic agents used for the D&C were fentanyl and propofol.

BACKGROUND

Opsoclonus myoclonus syndrome (OMS), also known as dancing eyes-dancing feet syndrome, is a serious and often chronically disabling neurological condition. It is an extremely rare syndrome, affecting as few as 1 in 10 000 000 people every year.¹ It presents with opsoclonus; defined as rapid and repetitive, involuntary, conjugate ocular saccades which are irregular in amplitude and frequency and occur in all directions without an intersaccadic interval; ataxia, myoclonus, irritable behaviour and sleep disturbances. Neurocognitive and psychiatric sequelae may also feature.^{2,3}

OMS is associated with occult neuroblastoma in up to 40% of cases, especially when the onset occurs in childhood. In adults, it is commonly paraneoplastic in origin, accompanying breast carcinoma or small cell lung carcinoma. Other aetiologies involving viral or toxic agents have also been reported. The pathogenesis is thought to be immune mediated on the basis of the paraneoplasticity and symptomatic response to immunosuppressive therapy. However, no immunological marker has yet been identified.⁴ There is classically no diagnostic test, with investigations showing non-specific results and brain imaging reported as normal.

Therapeutic benefit has been described with the use of steroids, intravenous immunoglobulins, cyclophosphamide, azathioprine and rituximab, but randomised trials have proven to be extremely difficult because of the rarity of the condition.⁴

The prognosis of OMS depends on early management and successful treatment, and therefore prompt identification of the syndrome and the underlying cause is essential.

To the best of our knowledge, this is the first reported case of OMS occurring acutely in association with an intervention under anaesthesia.

CASE PRESENTATION

A 37-year-old woman was being seen by neurologists in view of a 10-year history of recurrent facial palsies and optic neuritis which were thought to be autoimmune in origin due to the fact that she responded well to treatment with steroids. All investigations were negative.

She had first presented in 2000 with a left-sided Bell's Palsy followed by complete remission with oral prednisolone. Over 2006 and 2007, she developed three separate episodes of right retrobulbar neuritis, treated with intravenous methylprednisolone; and in 2008 she presented with a right-sided Bell's Palsy which was severe enough to require management with tarsorrhaphy.

In April 2015, she was admitted by the gynaecologists for an elective dilatation and curettage (D&C) for menorrhagia. She had no history of anaesthetic complications and a surgical history included two Caesarean sections performed under general anaesthesia, a right-sided carpal tunnel syndrome release and an appendectomy. The anaesthetic agents used for the D&C were fentanyl and propofol.

Immediately after the procedure, she developed an acute confusional state with transient left-sided numbness, paraesthesia and weakness. Within the first 24 hours she became disoriented, with incoherent speech and inability to obey simple commands. Her eyes were kept closed, but on examination, opsoclonus with rapid jerky eye movements in all directions of gaze was revealed. She also had repetitive involuntary myoclonic jerks of both her upper limbs. Examination of her face revealed right facial synkineses due to her previous lower motor neuron facial palsy but no other focal signs. Breast examination and ultrasonography as well as gynaecological assessment did not reveal abnormalities.

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INVESTIGATIONS

Investigations performed included: routine blood tests, serum B12 and folate levels, erythrocyte sedimentation rate and C-reactive protein, viral screen, vasculitic screen, onconeural antibodies (including anti-Ri and anti-NMDA receptor antibodies), cerebrospinal fluid analysis, chest radiography, CT and MRI of the brain, which were all normal. Electroencephalography on the first day showed background slowing compatible with a non-specific encephalopathy.

DIFFERENTIAL DIAGNOSIS

OMS is uniquely autoimmune, though may have paraneoplastic, parainfectious or idiopathic autoimmune causes.

TREATMENT

She was treated with intravenous methylprednisolone for a duration of 5 days.

OUTCOME AND FOLLOW-UP

She responded immediately with significant improvement and no residual features of her opsoclonus myoclonus and subsequently discharged. Subsequent 2-year follow-up for possible occult underlying malignancies did not reveal any such developments.

Learning points

- ▶ Opsoclonus myoclonus syndrome (OMS) is a rare neurological syndrome
- ▶ Usual aetiologies are autoimmune, paraneoplastic, viral or toxic
- ▶ To our knowledge this is the first case report of OMS after an intervention under anaesthesia.

DISCUSSION

This case report illustrates the occurrence of acute OMS after a minor surgical (gynaecological) intervention. In this patient, a possible predisposition may be autoimmune in view of her previous multiple steroid responsive cranial mononeuropathies. Given this phenotype, the OMS in this patient is almost certainly autoimmune. Perioperative autoimmune disorders have been reported in the literature including perioperative inflammatory neuropathies and plexopathies.⁵ Moreover, as described in the Mayo clinic series, autoimmune OMS may be antibody negative but still respond well to immunotherapy.⁶

There was no evidence for a paraneoplastic neurological syndrome even after subsequent surveillance.

OMS and its relationship with anaesthesia has not been studied in detail. Fentanyl and propofol have been reported to be safe with established OMS,⁷ while other anaesthetic agents as ketamine and etomidate aggravate the opsoclonus and myoclonus.⁸

Contributors AA wrote the case report with the guidance of the firm responsible for the patient's care. Resident specialist, neurology: ND. Consultant neurologist: JA.

Competing interests None declared.

Patient consent Obtained.

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