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Opsoclonus-Myoclonus syndrome

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DESCRIPTION

Opsoclonus-Myoclonus syndrome is a rare dyskinesia due to an underlying infectious, metabolic or neoplastic process.¹ It is frequently caused by neuroblastoma in children and breast or lung carcinoma in adults. It is characterised by high amplitude, multidirectional, arrhythmic and conjugate ocular saccadic intrusions without intersaccadic latency causing oscillopsia² and it is associated with myoclonus of axial and/or appendicular musculature³ and ataxia. Autoimmune dysfunction of Purkinje cells in the dorsal vermis and subsequent disinhibition of oculomotor fastigial region seems to be the most probable mechanism.⁴ A 53-year-old female, diagnosed with breast carcinoma 2½ years before the first ophthalmologic assessment. She had refused mastectomy and had nausea, vomiting and cerebellar gait ataxia followed by dysarthria, opsoclonus-related oscillopsia (could not read) and cervical myoclonus. The syndrome was exacerbated by tactile, sound or bright light stimuli and reduced by convergence (video 1). The patient was submitted to left radical mastectomy and monthly chemotherapy (adriamycin 60 mg/m² and cyclophosphamide 600 mg/m²). Due to persistence of the opsoclonus-myoclonus syndrome with severe gait ataxia, intravenous immunoglobulin (IVIG), 2 g/kg, was started. Progressive improvement of oscillopsia and complete remission of opsoclonus was evident. Six months after initiating therapy, she had complete

remission of the myoclonic component. Mild broad base gait is at the moment, possible without aid. We report a severe Opsoclonus-Myoclonus syndrome starting 2½ years after breast carcinoma. Mastectomy and adjuvant chemotherapy were essential in stabilising the underlying pathologic mechanism but the immune-modulating action of IVIG seems to be helpful since clinical improvement was observed after initiating therapy with complete remission after 6 months.

Video 1 Opsoclonus-Myoclonus video: high amplitude multidirectional and irregular saccadic intrusions, without intersaccadic latency, exacerbated by bright light stimuli and reduced by convergence. Vestibulo-ocular reflex and opto-kinetic nystagmus are present. We can see concomitant cervical axial musculature myoclonus. 10.1136/bcr.09.2011.4834v1

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Guedes ME, Almeida AC, Patricio MS, Costa JM. Opsoclonus-Myoclonus syndrome. *BMJ Case Reports* 2011;10.1136/bcr.09.2011.4834, date of publication

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