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Case Report

Opsoclonus-Myoclonus-Ataxia Syndrome Revealing a Neuroblastoma

S Mrhar*, F Taher, K Elfakiri, N Rada, G Draiss and M Bouskraoui

Pediatrics Department A Mother and Child Hospital Center Hospitalier Mohammed VI, Marrakech, Morocco

***Corresponding Author:** S Mrhar, Pediatrics Department A Mother and Child Hospital Center Hospitalier Mohammed VI, Marrakech, Morocco.

Received: May 03, 2023; **Published:** May 26, 2023

Abstract

Opsoclonus-myoclonus-ataxia is a clinical syndrome which may be of paraneoplastic, infectious, post-infectious, post-vaccination or idiopathic origin. We report the case of a 23-month-old infant hospitalized for gait disorders with abnormal eye movements. Examination revealed an irritable infant with cerebellar ataxia and multidirectional nystagmus. Brain MRI was normal. The thoracoabdominal CT scan showed an adrenal neuroblastoma. The treatment was symptomatic by corticosteroid therapy and etiological by surgery and chemotherapy. Neuroblastoma remains the most common etiology of opsomyoclonic syndrome in pediatrics.

Keywords: *Neuroblastoma; Opsoclonus-Myoclonus; Paraneoplastic Syndrome*

Introduction

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare disorder of the nervous system that classically presents with a combination of eye movements and myoclonus, in addition to ataxia, irritability, and sleep disturbance [1]. Also known as opsoclonus -myoclonus syndrome or dancing eye syndrome, myoclonic encephalopathy and Kinsbourne syndrome [2]. It represents above all a paraneoplastic syndrome, through its association with neuroblastoma in children, and with various tumors in adults, with a probable immunological physiopathological mechanism [3]. A rather favorable prognosis of neuroblastoma has already been pointed out.

Observation

23-month-old EH infant, with no particular pathological history, hospitalized in the pediatric department A, Marrakech University Hospital for gait disorder with agitation and abnormal eye movements evolving for a week before admission. The physical examination objectified a conscious, irritable, afebrile infant with gait instability and enlargement of the support polygon, myoclonus and generalized hypotonia. The ocular examination showed disordered movements of the eyes in the type of multidirectional oscillations. This clinical picture, which associated cerebellar ataxia, disordered eye movements and myoclonus, evoked an opsoclonus -myoclonus syndrome. The cerebral MRI was without abnormality, the thoracoabdominopelvic scanner found a localized adrenal neuroblastoma, the biological assessment was normal.

Management was both symptomatic based on corticosteroid therapy at a dose of 1 mg/kg/d and etiological by total excision of the adrenal mass followed by chemotherapy. The evolution was marked by a partial improvement of the neurological signs.

Discussion

Opsoclonus-myoclonus syndrome is clinical, corresponding to ocular dyskinesia which includes involuntary, arrhythmic, chaotic and multidirectional ocular saccades. Saccades have a horizontal, vertical or rotational component. The opsoclonus appears during fixation, slow eye pursuit, or convergence, and it is persistent during sleep or eye occlusion. Due to the large amplitude and high frequency of saccades, it can cause visual blurring or oscillopsia. It differs from nystagmus because there is no slow component but always a saccade after the fixation of an object [4-6]. Opsoclonus is often associated with diffuse myoclonus and other signs of brainstem involvement or cerebellar syndrome as part of the opsoclonus-myoclonus syndrome [4,7]. In about 50% of children with OMAS, there is an underlying neuroblastoma [1,8]. There is good evidence that OMAS is an immune-mediated condition that may be paraneoplastic [1]. As it is very rare, it is necessary to have a high level of clinical suspicion and to consider it among the differential diagnoses of acute-subacute onset ataxias [1]. The earlier the diagnosis is established, the more crucial it is for a good evolution and drastically reduces the chances of long-term neurological sequelae [3]. It is an immune-mediated disorder and its treatment is based on immunosuppressants, immunomodulators and tumor resection in cases secondary to neuroblastoma. Between 70% and 80% of cases may have neurological sequelae, depending on the cause, the initial severity of the symptoms and the speed of introduction of treatment [1,3].

Conclusion

Neuroblastoma remains the most common cause of paraneoplastic opsoclonus-myoclonus-ataxia syndrome, which requires early diagnosis with rapid management to avoid neurological sequelae.

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Volume 12 Issue 6 June 2023