Opsoclonus Myoclonus Syndrome: a Brazilian perspective

Renata Barbosa Paolilo[1,2]; Mariana Braatz Krueger[3]; Roberta da Silva Mendonça Thurler[4]; Ricardo Silva Pinho[5]; Ingrid Lacerda Pessoa[1]; Ming Lim[6], Fernando Kok[1].

1. Department of Neurology. Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, São Paulo, Brazil; 2. Hospital Israelita Albert Einstein, São Paulo, Brazil;3. Hospital Infantil Albert Sabin, Fortaleza, Brazil; 4. Instituto Nacional do Câncer, Rio de Janeiro, Brazil; 5. Grupo de Apoio ao Adolescente e à Criança com Câncer, São Paulo, Brazil; 6. Children's Neurosciences, Evelina London Children's Hospital at Guy's and St Thomas' NHS Foundation Trust , London, United Kingdom.

18TH ICNC 2024 Footprints to the future
INTERNATIONAL CHILD NEUROLOGY CONGRESS
Cape Town, South Africa I May 6–10, 2024

INTRODUCTION

Opsoclonus myoclonus syndrome (OMAS) is a rare neurological disorder characterized by acute onset of opsoclonus, myoclonia, ataxia, and mood/sleep disorders. There is compelling evidence supporting it is an immune-mediated disorder, paraneoplastic in half of cases. This syndrome is linked with potential cognitive impairment.

OBJECTIVE

To describe demographic, clinical, paraclinical and prognostic aspects of OMAS patients from five centers in Brazil.

MATERIALS AND METHODS

Multicentric study including five centers in Brazil.

Retrospective data collection: RedCap survey with a structured questionnaire.

Inclusion criteria: patients fulfilling Genova's diagnostic criteria + ≤ 5y of symptom onset.

Exclusion criteria: other CNS condition, infection or systemic inflammatory disorder.

Outcome at last follow-up (FU): clinical remission, Michell-Pike scale, social/behavioral problems, sleep problems, learning disabilities.

Approval by the locals and the National Ethical Committee (approval number: 32733020.5.0000.0068)

RESULTS

Twenty-seven patients were included.

Most were female (59.3%) from mixed background ethnicity (59.3%).

Median (IQR) age at symptom onset was 15.7 (2.2-22.5) months.

Fifteen (55.6%) patients were diagnosed with a neuroplastic tumor.

The first disease manifestation is disclosed in the Figure 1.

Median (IQR) time to diagnosis was 28 (12-48) days. Most patients were misdiagnosed with other conditions such as cerebelitis, encephalitis, or epilepsy.

Most patients (76%) relapsed, with a median (range) of 2 (1-6) relapses by patient.

During FU all patients presented ataxia, 96.3% of patients presented opsoclonus and tremor/dysmetria and 88.9% presented behavioral/sleep problems.

Intravenous immunoglobulin was the most common prescribed immunotherapy (85.2% patients), followed by oral steroids (77.8% patients).

After a median (IQR) 1.7 (0.5-3.22) years follow-up, one patient died, and five (19.2%) patients reached clinical remission. Most common long-term sequelae are summarized in Figure 2.

Longer time to diagnosis (0.02) and longer time to initiate therapy (0.007) was associated with learning problems at last FU.

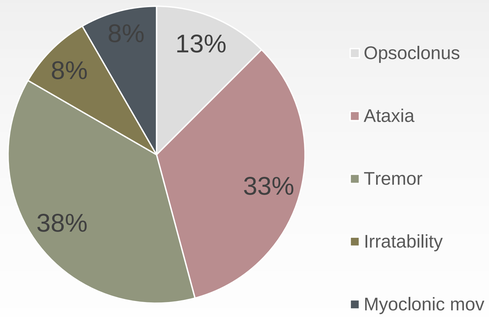

Figure 1. First symptom at disease onset.

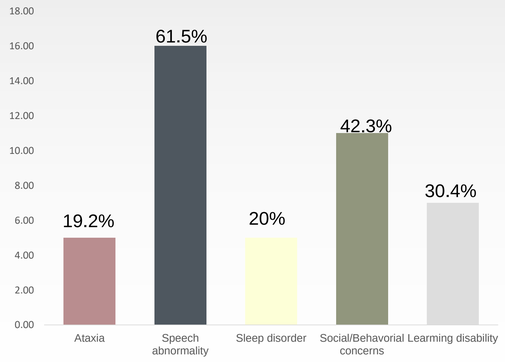

Figure 2. Neurological sequelae at last follow-up of 25 patients.

CONCLUSIONS

The demographic findings are similar to those of previously reported cohorts. To our knowledge, this is the first Brazilian characterization of OMAS.

Most patients had a paraneoplastic etiology.

Tremor and ataxia were the most common symptoms at disease onset.

OMAS is a highly disabling condition. Less than 20% of patients reached remission of symptoms.

In this multicentric Brazilian cohort, poor outcomes at the last FU were influenced by disease course and treatment.

REFERENCES

Matthay KK, Blaes F, Hero B, Plantaz D, De Alarcon P, Mitchell WG, et al. Opsoclonus myoclonus syndrome in neuroblastoma a report from a workshop on the dancing eyes syndrome at the advances in neuroblastoma meeting in Genoa, Italy, 2004. Cancer Lett. 2005;228(1-2):275-82.

Pike M. Opsoclonus-myoclonus syndrome. Handb Clin Neurol. 2013;112:1209-11.

Pranzatelli MR, Tate ED, McGee NR. Demographic, Clinical, and Immunologic Features of 389 Children with Opsoclonus-Myoclonus Syndrome: A Cross-sectional Study. Front Neurol. 2017;8:468.

Rossor T, Yeh EA, Khakoo Y, et al. Diagnosis and Management of Opsoclonus-Myoclonus-Ataxia Syndrome in Children: An International Perspective. Neurol Neuroimmunol Neuroinflamm. 2022;9(3):e1153. Published 2022 Mar 8.

Yıldırım M, Öncel İ, Bektaş Ö, et al. Clinical features and outcomes of opsoclonus myoclonus ataxia syndrome. Eur J Paediatr Neurol. 2022;41:19-26. doi:10.1016/j.ejpn.2022.09.002

ACKNOWLEDGMENTS AND CONTACTS

Acknowledgment: Dr José Albino da Paz

Contact: renata.paolilo@hc.fm.usp.br