CLINICAL PROFILE AND OUTCOMES OF OPSOCLONUS MYOCLONUS ATAXIA SYNDROME

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INTRODUCTION

Opsoclonus myoclonus ataxia syndrome (OMAS) is a rare autoimmune condition characterized by **opsoclonus, myoclonus, behavioral problems, ataxia and sleep disturbances.** The onset is usually in the second year of life and almost half of the cases are associated with neuroblastoma. In rest, the causes might be post-infectious, other autoimmune or idiopathic.

Long term cognitive, language and behavioral outcome is less than satisfactory, while the motor outcomes are better. High intensive immunomodulatory therapy (front load approach) using corticosteroids (CS) , intravenous immunoglobulin (IVIG) , rituximab (RTX)/cyclophosphamide (CYC) has been shown to improve the cognitive outcome in retrospective studies as compared to staggered approach. However, literature evidence is scarce with regard to the treatment response and outcome in children with OMAS, hence this study was planned

OBJECTIVES

To study the clinical profile and outcomes of OMAS in children admitted in our institute, a tertiary care level hospital

MATERIALS AND METHODS

- Children aged less than 12 years, satisfying three out of four criteria for OMAS—**opsoclonus, ataxia and/or myoclonus,** behavioral changes and/or sleep disturbances and neuroblastoma from 2018 were included in this ambispective study.
- The demography, clinical profile, investigations, treatment received and outcome (OMAS rating scale, number of relapses and remission) were recorded. Descriptive statistics were used for analysis.

RESULTS

- 5 out of 12 patients were prospectively enrolled after written informed consent

Table 1: Demographic and clinical characteristics

Demographic parameters	**Number of patients =12 (%)**
Mean age of onset in months (standard deviation, SD)	35.3 (51.3)
Sex, females	7 (58.3)
Clinical parameters	
Antecedent febrile illness	8 (66.7)
Ataxia	12 (100)
Opsoclonus	9 (75)
Myoclonus	7 (58.3)
Irritability	6 (50)
Sleep disturbance	3 (25)
Etiology	
Neuroblastoma	3 (25)
Anti-GAD positivity	1 (8.3)
Idiopathic	8 (66.7)

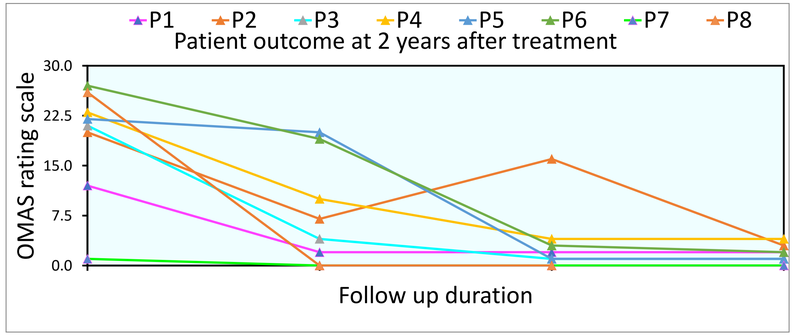

Figure 1: OMAS outcome at baseline and at 2 years

Table 2: Treatment and outcome

Treatment	**Number of patients =12 (%)**
Staggered approach	7 (58.3)
Frontload approach	5 (41.7)
Outcome	**n=8 patients**
Duration of follow up in months, mean (SD)	43.5 (24)
OMAS rating scale, mean (SD)	19 (8.6)
6 month	7.8 (8)
1 year	3.4 (5.3)
2 years	1.9 (1.4)
Complete remission	5 (62.5)
Residual behavioural problems	1 (12.5)
Residual cognitive deficit	1 (12.5)
Residual motor deficit	1 (12.5)
Relapses	7 (87.5)

CONCLUSIONS

- OMAS has significant long term sequelae with high rate of relapse.
- Early aggressive and prolonged immunomodulatory therapy is required for clinical remission

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