CLINICO-EPIDEMIOLOGICAL PROFILE OF CHILDREN WITH OPSOCLONUS MYOCLONUS ATAXIA SYNDROME: A DECADE'S EXPERIENCE FROM A TERTIARY CARE CENTRE IN NORTH INDIA

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INTRODUCTION

- Opsoclonus-myoclonus ataxia syndrome (OMAS), is an inflammatory neurologic syndrome with a strong connection to cancer
- A paraneoplastic etiology can only be proven presently in about 50% of the cases

OBJECTIVE

- To study the clinico-epidemiological profile and treatment strategies of children with OMAS

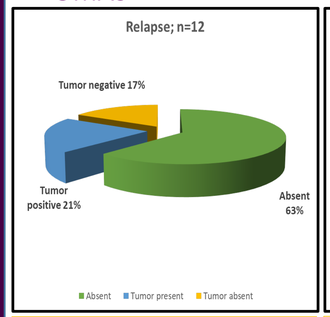

Fig 1. Prevalence of tumor positiveness in children with relapse, in OMAS

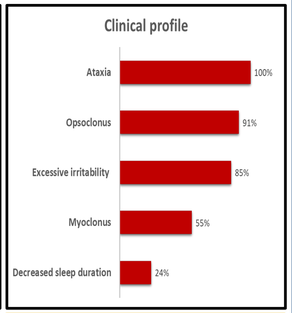

Fig 2. Clinical profile of children with OMAS

MATERIALS AND METHODS

- A retrospective review and analysis of case records of children presenting with OMAS (January 2012 to March 2022) to a tertiary centre was performed

RESULTS

- A total of 40 children (23 females, 17 males) were identified
- The median age of symptom onset was 21.5 months (IQR :16.3- 24.5)
- 17/40 (42.5%) were non ambulatory at presentation
- Underlying neuroblastoma was present in 19/40 (47.5%)
- In the non-tumor group (n=21), 4/21(19%) were para-infectious and 17/21 (80.9%) were idiopathic
- Antineuronal antibodies (2 for anti-Hu and 1 for anti PNMA2) were detected in 7.5 % (3/40) of children and they showed early response to immunotherapy
- 42.5% received IVIG+ACTH, 40% ACTH alone, 7.5% IVIG+IV pulse methylprednisolone and 10% only steroids as the 1st line therapy
- Irritability (58%) was the first symptom to improve
- 15 children (37.5%) showed relapse on first line therapy (median time: 5 months)
- Relapse was treated with IVIG + ACTH in 7/13 (53.8%), 6/13 (46%) required rituximab (375mg/m2 - 1.12mg/m2) and 8/13 (61.5%) required long term immunosuppression

CONCLUSIONS

- Children in the tumor group had an earlier age of onset, attained delayed remission and had more relapses in comparison to non-tumor group.
- Early diagnosis, periodic tumor surveillance and aggressive combined immunotherapy is the key in improving outcomes.
- With the emergence of CNS autoantibodies, its worthwhile to look at the antineuronal antibody status in these children as it has important therapeutic implications

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