“CONFUSION AT THE JUNCTION”: JUVENILE MYASTHENIA GRAVIS AT A TERTIARY HEALTH CARE FACILITY IN GHANA

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INTRODUCTION

Juvenile Myasthenia gravis (JMG) is seen in children under 18 years of age.

It is a disorder of neuromuscular transmission due to binding of autoantibodies to components of the neuromuscular junction –commonly the nicotinic acetylcholine receptor, and results in variable muscle weakness, with characteristics of fatigability and diurnal variation.

BACKGROUND & OBJECTIVES

Korle Bu Teaching Hospital is the premier tertiary facility in Ghana and serves as the third largest referral facility in Africa.

The Department of Child Health (DCH) has an average annual out-patient attendance of 36,000 with several sub-specialty clinics, including a neurodevelopmental clinic where patients with various neurologic conditions are followed up.

Clinical data on JMG in Sub-Saharan Africa is generally lacking.

This case series aims to characterize the clinical and socio-demographic features of JMG in patients in a tertiary hospital in Ghana.

This was a retrospective observational study in which the medical records of children with myasthenia gravis who presented to DCH between January 2019 and November 2023 were retrieved and analysed.

DESCRIPTION OF CASES

Over the period, a total of ten patients were seen. Their ages ranged from two to sixteen years with a median age of 7.7 years old.

Similar to literature, the majority of our patients were female and the most common initial feature was ptosis. 50% of patients also had generalized weakness at their index visit, 20% had dysphonia and 10% had dysphagia at initial presentation.

All patients seen had an ice pack test done which was positive.

Unfortunately, due to financial constraints only 30% had serology done. Two out of three of these patients had acetylcholine receptor antibodies and the third patient was seronegative.

Single fibre electromyography showed 11% decremental response for the one patient it was performed on.

None of our patients had evidence of thyroid dysfunction or other autoimmune disease.

Oral pyridostigmine was prescribed for all patients and 70% had combination therapy with oral corticosteroids due to progression of their symptoms.

One child developed a myasthenic crisis, was admitted to the paediatric intensive care unit, given intravenous immunoglobulin and mechanically ventilated, but died after three days.

RESULTS

Fig. 1. Clinical Features of Patients with JMG

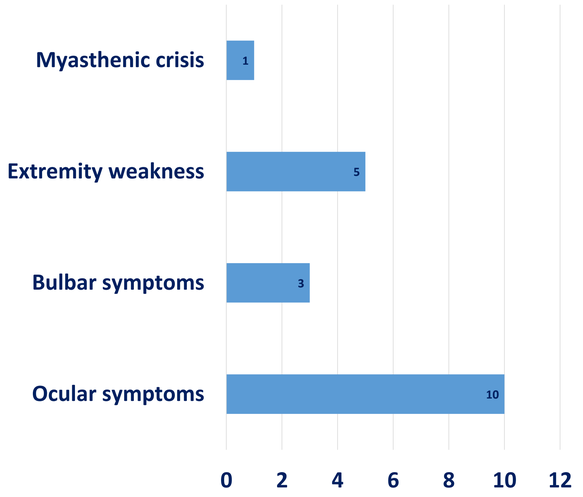
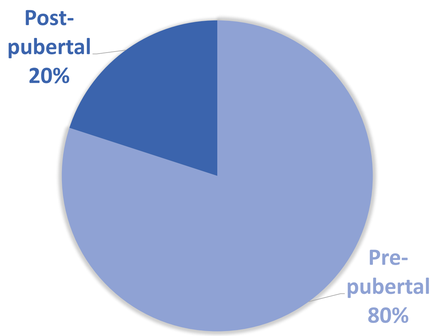


Fig. 2. Classification Of JMG By Age Of Onset



TEACHING POINTS

- JMG is a rare, non-hereditary autoimmune condition which results in varying degrees of muscle weakness.
- Generally, onset of symptoms pre-pubertally is associated with a better outcome[1].
- Early recognition, diagnosis and treatment are key due to the potential risk of mortality from respiratory muscle involvement and myasthenic crisis.
- Cholinesterase inhibitors alone or with corticosteroids are usually first-line therapy[2].
- Multicenter studies in the paediatric population are recommended to help in providing standardized treatment guidelines and improving outcome.

REFERENCES

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