Case Report

Juvenile Myasthenia Gravis in a Black Zambian Child

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ABSTRACT

A 7 year old girl with progressive ptosis and worsening generalised body weakness presented to the out-patient clinic of a general paediatric unit at the University Teaching Hospital (UTH) after having been to a number of health institutions before a diagnosis of Juvenile Myasthenia Gravis was made. She was commenced on a steroid and anticholinesterase therapy with great improvement. This case report summarises the progression of symptoms, challenges in diagnosis and improvement on initiation of pyridostigmine therapy.

BACKGROUND AND LITERATURE REVIEW

Myasthenia Gravis (MG) is an autoimmune disease in which antibodies are directed at the postsynaptic membrane of the neuromuscular junction leading to varying degrees of muscle weakness and fatigability. Where MG presents before 19 years of age, it is termed juvenile myasthenia gravis (JMG)[1]. In South Africa, the median age of onset of symptoms was seven years, with ocular MG being present in 26% of patients and was more common among younger children (mean age of 5.1 years), compared to generalized MG (74%) with a mean age of 10.2 years[2]. In the same study it was reported that children with ocular MG presented at a younger age than those with generalised MG. Subjects presenting with generalised symptoms were diagnosed after a longer delay than those presenting with ocular symptoms mean 1.2±2.1 vs 0.8±1.4 years, respectively[2].

The incidence of MG in black African children is unknown[2]. In Tanzania, the incidence among children less than 10 years of age was 2.2/1,000,000[3]. In Zambia the incidence of MG is unknown and there are no reported cases of the condition in children.

CASE PRESENTATION

A 7 year old female presented to the out-patient clinic of UTH, Department of Paediatrics and Child Health with progressive ptosis for two years, easy fatigability and weight loss for one year, and falling down when walking for five months. She had earlier on presented to a local clinic with itchy red eyes and treated for conjunctivitis. She then developed difficulties keeping her eyes "glued" at one object and drooping of both eyes lids. She was then seen at a Central Hospital by the ophthalmologist who made a diagnosis of myasthenia gravis and referred her to UTH.

Over the following months, she had difficulties bathing herself, and became progressively slow when eating and eventual weight loss. She was inactive and at school she was having difficulties copying notes from the board and would miss several words. As she had difficulties in walking, she would sometimes fall as she walked from the school gate to the classroom. Watching television had also become a daunting task and she had to "peg" her eye lids with her fingers in order to keep them open.

Clinically, she was a well and alert school going child who interacted well with surroundings. She

Key words: *Myasthenia Gravis, Bilateral ptosis, Delayed diagnosis, Child*

had obvious bilateral ptosis which became more apparent with an upward gaze. She tired easily during repetitive opening and closing of the fists, had difficulties elevating her arms for more than five minutes and was noted to have difficulties combing her hair. She also had difficulties ascending and descending stairs.

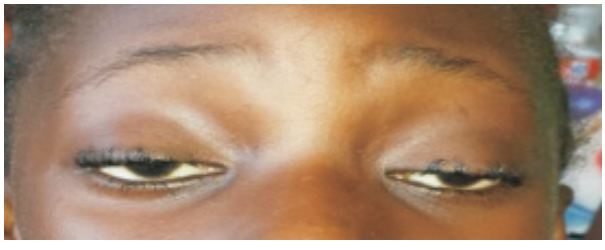
Gait, muscle bulk and power were all normal though the reflexes were rather diminished. Her vitals were all normal and both pupils were reacting to light. Her BMI fell just below the 85th percentile for age.

INVESTIGATIONS

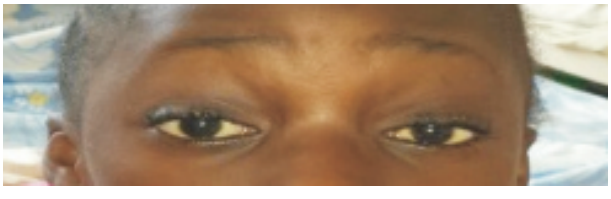
A full blood count and chest radiograph were normal. The neostigmine test was remarkable with improvement of the ptosis within 20 minutes of administration and a diagnosis of *juvenile myasthenia gravis* was confirmed.

Figure 1. Neostigmine intramuscular test at 20 and 40 minutes after adminstration

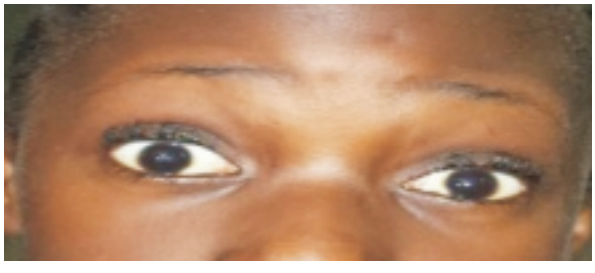
Before neostigmine test



20 minutes after neostigmine administration



40 minutes after neostigmine administration



TREATMENT

She was started on prednisolone 25mg (1mg/kg) on alternate days and pyridostigmine 30mg once a day titrated to optimal response over two months. She was on pyridostigmine 60mg 8 hourly and showed good response. She has regained full physical activity. Over time, she needed higher doses of pyridostigmine and prednisolone but developed cushingoid facies and prednisolone had to be withdrawn at some point. Overtime she has been on and off prednisolone while the dose of pyridostigmine has been adjusted according to response. She is currently on prednisolone at 15mg/day (0.32mg/kg/day) and pyridostigmine at 60 mg 8 hourly (3.8mg/kg/day)[180mg/day]. She has been followed up for four years now and currently doing well.

DISCUSSION

In sub-Saharan Africa, there is delay in the diagnosis of MG, and in our patient the diagnosis was made after two years. The mean duration from onset to diagnosis of MG in Tanzania was 3.6 years[3]. In the Republic of South Africa it was 1.2±2.1 and 0.8±1.4 years for those presenting with generalised symptoms and for those with ocular presentation respectively. Though JMG is rare, health workers need to be sensitized about this condition.

The diagnosis is based upon clinical symptoms and signs, with laboratory and electrophysiological studies used for confirmation[4]. Probably, the referral system need to be streamlined especially that specialists are becoming increasingly available at Provincial General Hospitals. However, even in resource limited settings such as Zambia, a high index of suspicion with limited but available tests can lead to the confirmation of the diagnosis of MG. Intravenous neostigmine is a safe, simple and efficient alternative diagnostic test for MG in regions around the world where the other standard tests for MG are either unavailable or technically not feasible[5]. Treatment, like what was used in the patient, has proven very effective and her quality of life was restored within a short period though the challenge has been the non availability of

pyridostigmine in the public health service as the family had to procure it privately.

CONCLUSION

Juvenile Myasthenia Gravis should be considered in a child who presents with unilateral or bilateral ptosis and especially so if accompanied by easy fatigability.

ACKNOWLEDGEMENT

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