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Unmasking Weakness: A Rare Case of Juvenile_Myasthenia Gravis in a Young Child

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ABSTRACT:

Juvenile Myasthenia Gravis (JMG) is an uncommon autoimmune disorder characterized by muscle weakness and fatigue, with a prevalence of 10-15% among all myasthenia gravis cases. This report presents a rare case of JMG in a 4-year-old boy, detailing the clinical evaluation, diagnosis, and comprehensive management strategies. The patient exhibited bilateral ptosis and generalized muscle weakness, confirmed by elevated acetylcholine receptor antibodies and repetitive nerve stimulation tests. Management involved acetylcholinesterase inhibitors and corticosteroids, resulting in significant clinical improvement. Early diagnosis and a multidisciplinary approach are crucial for optimal outcomes in pediatric JMG cases.

Keywords: Juvenile Myasthenia Gravis, pediatric autoimmune disorder, muscle weakness, ptosis, acetylcholinesterase inhibitors, corticosteroids, acetylcholine receptor antibodies.

1. Introduction

Juvenile Myasthenia Gravis (JMG) is a rare autoimmune neuromuscular disorder in children, characterized by fluctuating weakness of voluntary muscles. The incidence of JMG in children is approximately 1 in 500,000. The condition often presents with ptosis, diplopia, and generalized muscle weakness. Early diagnosis and appropriate management are essential to prevent complications and improve the patient's quality of life.

Case Presentation

A 4-year-old boy was brought to the pediatric clinic with concerns of drooping eyelids and generalized muscle weakness, which worsened throughout the day. Symptoms had been progressively worsening over the past two months. The child had no history of respiratory issues, difficulty swallowing, or significant weight loss. His developmental milestones were appropriate, and there was no family history of neuromuscular disorders.

On examination, the boy appeared alert and cooperative, with bilateral ptosis more pronounced on the left side and noticeable facial weakness with a "snarling" smile. Muscle strength testing revealed weakness in the proximal muscles of the limbs, with normal deep tendon reflexes. There were no signs of muscle atrophy or fasciculations.

Initial laboratory investigations, including complete blood count, electrolytes, and thyroid function tests, were normal. A repetitive nerve stimulation (RNS) test showed a significant decremental response in the amplitude of evoked muscle action potentials, consistent with a neuromuscular junction disorder. Serum acetylcholine receptor (AChR) antibodies were elevated, confirming the diagnosis of juvenile myasthenia gravis.

Management

The patient was started on oral pyridostigmine, an acetylcholinesterase inhibitor, at a dose of 1 mg/kg per dose, given every 4-6 hours. Due to the severity of symptoms, prednisolone was initiated at a dose of 1 mg/kg/day. The child's parents received education about the disease, medication adherence, and the importance of regular follow-ups. Additionally, the family was referred to a pediatric neurologist for further management and support.

Over the following weeks, the boy showed significant improvement in muscle strength and a reduction in ptosis. Pyridostigmine dosage was adjusted based on clinical response, and prednisolone was gradually tapered over several months to minimize potential side effects. Regular follow-up visits were scheduled to monitor for disease progression and treatment side effects. During these visits, the patient was also evaluated for potential complications, such as corticosteroid-induced growth suppression and cataracts.

Outcome and Follow-Up

After six months of treatment, the boy exhibited marked improvement, with a significant reduction in muscle weakness and ptosis. He remained stable on a maintenance dose of pyridostigmine and low-dose prednisolone. Regular follow-ups were conducted every three months to monitor his condition and adjust medications as needed. The family was provided with ongoing support and education to manage the condition effectively, and the patient continued to see a pediatric neurologist for specialized care.

2. Discussion

Juvenile Myasthenia Gravis is a rare but treatable condition in children. The clinical presentation can vary, but common symptoms include ptosis, diplopia, and generalized

muscle weakness. Diagnosis is confirmed through clinical evaluation, RNS tests, and detection of AChR antibodies.

Management typically involves acetylcholinesterase inhibitors like pyridostigmine to improve neuromuscular transmission. Corticosteroids, such as prednisolone, are often used to suppress the autoimmune response. In refractory cases, immunosuppressive agents like azathioprine or mycophenolate mofetil may be considered. Thymectomy is another therapeutic option, especially in patients with thymoma or those not responding to medical treatment.

Long-term prognosis for JMG is generally favorable with appropriate treatment, but regular monitoring and follow-up are essential to manage potential relapses and treatment side effects. Multidisciplinary care involving pediatric neurologists, endocrinologists, and psychologists is crucial for comprehensive management and support.

3. Conclusion

This case highlights the importance of considering juvenile myasthenia gravis in the differential diagnosis of pediatric patients presenting with muscle weakness and ptosis. Early diagnosis and a multidisciplinary approach to management can significantly improve outcomes and quality of life for affected children. Continued research and awareness are necessary to enhance understanding and treatment of this rare condition.

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