Myasthenia gravis with isolated bulbar syndrome on presentation, two case reports

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Objective

Analyzing the clinical signs at first presentation of Myasthenia Gravis patients and bringing into attention cases with bulbar syndrome as the only presenting sign

Introduction

Myasthenia Gravis (MG) is an autoimmune disorder mediated from antibodies clinically presented with easy fatigability of various groups of muscles. The most common antibodies causing the clinical features are anti-AChR and anti-MuSC antibodies. Unilateral or bilateral palpebral ptosis is the most common symptom at initial presentation with more than 75% of cases. Less frequently the disease may present with generalized fatigability in the limb muscles or bulbar syndrome. Up to 20% of MG Cases may have prominent oropharyngeal symptoms early in the disease course but usually associated with the other clinical symptoms. Anti-MuSC MG has several characteristics that differ from anti-AchR MG. The bulbar involvement is more common and can more often be the presenting symptom

Methods and Materials

From our MG database we selected 2 cases who have recently been diagnosed after presenting solely with bulbar symptoms in their first clinical examination

Case 1

A 55 year old man presented with a 2 months history of dysphagia and dysarthria, not associated with easy fatigability or ocular symptoms. After excluding other causes of bulbar syndrome (vascular etc) the patient underwent electrophysiological and laboratory tests and was diagnosed with anti-MuSC MG. No thymus abnormality was found in chect CT. Later in the course of the disease he developed bilateral facial paralysis and respiratory distress. He was twice under mechanical ventilation and did 2 cycles of plasmapheresis before being stabilized with a corticosteroid regimen.

Case 2

A 84 year old man presented with a one year history of intermittent dysphagia and dysphonia. He was hospitalized because of severe difficulty swallowing without easy fatigability, diplopia or ptosis. The lab studies showed anti AChR Ab level of 4.98 nmol/L. After the treatment with Pyridostigmine the symptoms resolved quickly and the patient has been in optimal conditions with a 180mg/day dose of Pyridostigmine

Discussion

Bulbar syndrome as a clinical entity can be due to several causes ie. vascular, degenerative, inflammatory, autoimmune etc. In MG the isolated bulbar presentation can be in up to 20.73% of the cases[1]. In the Anti-MuSC MG the facial and bulbar involvement can be as high as 84% [2] while in the anti-AChR group the percentage is much lower.
Anti- MuSC MG patients often don't respond to anticholinesterase drugs[2,3] and immunupressants might be necessary

Conclusions

Approximately 20% of patients with Myasthenia Gravis present with bulbar symptoms especially in the patients positive for anti Anti-MuSC antibodies. Anti MuSC MG patients don't respond to Pyridostigmine and are more prone to myasthenic crisis. Myasthenia Gravis should be included in the differential diagnosis of patients presenting with bulbar syndrome regardless having or not other MG features

References

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