

CASE REPORT

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Myasthenia gravis crisis: A case report

Christine Sykalo, Babajide Adio, Sandra Birchem

ABSTRACT

Introduction: Myasthenia gravis (MG) is an autoimmune disease targeting the postsynaptic acetylcholine receptors, which typically presents with fatigue symptoms relieved by rest and is often associated with a thymoma. Myasthenic crisis requires intubation and ventilation, and only 2–3% of MG patients are admitted with crisis per year. Impeding crisis may have a presentation of dyspnea, bulbar symptoms, or both together, requiring immediate attention. **Case Report:** We present a seropositive patient without a thymoma, who presented multiple times with dyspnea, which led to anxiety-triggered crisis within minutes. The patient experienced the crisis within two years of being diagnosed with MG, and required intensive care unit (ICU) care for five days, including intravenous immunoglobulin (IVIG) and steroids. **Conclusion:** The importance of symptomatic assessment is crucial to avoid and treat myasthenic crisis. Approximately 13–20% of patients, who experience a myasthenic crisis, have it as their initial presentation upon diagnosis, and require ICU care and intubation.

Keywords: Bulbar symptoms, Myasthenia gravis, Myasthenic crisis, Respiratory failure

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INTRODUCTION

Myasthenia gravis is an autoimmune disease targeting the postsynaptic acetylcholine receptors, which typically presents with fatigue symptoms relieved by rest [1, 2]. It has a bimodal age distribution, with peak onset in the 20–30s with female prevalence, and male prevalence in the 60–80s age range [2]. Majority of patients present with symptoms of fatigue relieved by rest and intact reflexes that may become progressively weaker on repeated use. More than half of patients present with ocular symptoms of ptosis and/or diplopia, which develop into generalized disease within two years. About 15% of patients present with bulbar symptoms, including dysarthria, dysphagia, and fatigable chewing, which requires increased attention due to risk of crisis [2].

Myasthenia gravis often occurs in conjunction with a thymoma, the resection of which may lessen disease severity or cure it [3]. The disease can be diagnosed by the presence of anti-acetylcholine receptor antibodies (anti-AchR-Abs), which are predominantly the binding antibodies, although there are also blocking and modulating antibodies. These antibodies are present in 85% of patient with generalized MG, and almost 100% of those who concurrently have a thymoma [4]. The MuSK antibodies are present in 38–50% of myasthenic patients who are AchR-Ab negative, and are seen more frequently in patients without a thymoma. Finally, anti-striated muscle antibodies target heterogeneous striated muscle proteins and are present in 80% of patients with a thymoma, but only 36% of those with generalized disease. Thus, the presence of different antibodies can sway the treatment regimen, although some 6–12% of myasthenic patients are seronegative.

The bedside ice pack test can be used as an extension of the neurologic examination to suggest the need for further evaluation, if ptosis resolves with direct cooling of the eyelid. The Tensilon test, using edrophonium, which was previously an initial testing modality, is no longer

available in the United States as of 2017 [4, 5]. In terms of prevalence, MG is seen in approximately 70 people per 320 million. Within this group, around 10% experience a myasthenic crisis requiring intubation and ventilation within the disease course, and only 2–3% of MG patients are admitted with crisis per year [6–8]. Impending crisis may have a presentation of dyspnea, bulbar symptoms, or both together, requiring immediate attention. Here, we present a seropositive patient without a thymoma, who presented multiple times with dyspnea, which led to anxiety-triggered crisis within minutes.

CASE REPORT

A 57-year-old male presented 07/2017 to the emergency room (ER), with three days of left eye ptosis with onset upon awakening. For a week, he had noticed double vision, which was somewhat corrected by his glasses, but he had not seen an optometrist since 1998. He continued his normal activity, but noticed issues of balance. The patient reported past medical history of psoriasis, no allergies, no medications, and no alcohol or drug use. Vital signs included blood pressure (BP) of 116/95 and body mass index (BMI) of 31 kg/m². On physical exam, his pupils were equal, round, and reactive to light, with left lateral gaze palsy and ptosis. No other abnormalities were noted on labs, chest X-ray, and head computerized tomography (CT). Neurology was consulted, and the ordered myasthenia blood panel was positive for the following antibodies: AchR binding, AchR modulating, GAD65 (0.04), and striated muscle.

The patient was seen by neurology in 07/2017, for progression of symptoms of weakness in the lower extremities and two days of jaw weakness while eating. At the time, he had recurrence of left eye ptosis and diplopia, without dysphagia, dysarthria, or shortness of breath. He was admitted for a myasthenia flare on the same day to the hospital, and treated with IVIG (400 mg/kg IV QD × five days), prednisone (20 mg po QD), and pyridostigmine (60 mg po TID). Symptoms improved, the patient was discharged five days later. A chest CT done during this hospital stay showed no evidence of thymoma.

The patient was slowly weaned off steroids, completing course in 09/2018. In October 2018, he presented to the emergency department (ED) with difficulty in breathing over two days, diplopia, neck locking, and hard stools. Similar symptoms of dyspnea were present when he was originally diagnosed with MG. Leukocytosis of 13.4×10^3 u/L and mild hyperglycemia, felt related to prednisone use, were noted. A chest X-ray was normal. The patient was discharged from ER with resuming prednisone and albuterol inhaler. He did not complete recommendations of follow-up with neurology.

In January 2019, he presented to the ED with gradual onset of worsening weakness. He ran out of his prednisone on 01/06/2019 and scheduled neurology appointment was in two weeks. Symptoms included

ptosis, worsening bulbar symptoms of difficulty chewing, and overall trouble with moving. He was discharged on 10 mg prednisone TID for 14 days, until his neurology appointment. On 01/24/2019, he presented to the ER with dyspnea. His bulbar symptoms were more pronounced with dysarthria, ptosis, and he reported a feeling of “100 lbs pushing on my neck and I cannot keep my head up.”

Exam was in process in the ER and the patient stated he needed to urinate. He became notably anxious with sudden worsening respiratory distress. Within minutes of reporting the need to urinate, patient became severely cyan with respiratory failure and required emergent intubation. Given clinical picture of cyanosis, hypophonia, dyspnea, and poor respiratory effort, the patient was then hospitalized in the ICU for five days with respiratory intubation and mechanical ventilation. After the passing of a spontaneous breathing trial, the patient was monitored on the floor and discharged home. He received five days of IVIG and stably discharged on 60 mg of prednisone and 50 mg of azathioprine daily. Currently, he is being followed by neurology, with consideration of surgery for thymus removal if symptoms will not be controlled with the medical regimen.

DISCUSSION

The patient presentation was unique in multiple ways. Antibodies suggest a rich autoimmune process, and his seropositivity is suggestive of myasthenia gravis with a likely thymoma, although one was not visualized on chest CT. GAD65 antibodies indicate susceptibility to autoimmune disorders, but relate more commonly to endocrine disorders like type 1 diabetes mellitus, thyroiditis, and pernicious anemia. Neurological disorders more commonly have a GAD65 < 0.03 nmol/L, and anything greater than that suggests a diagnosis of stiff-man syndrome, autoimmune encephalitis, cerebellitis, brain stem encephalitis, or myelitis [9]. Although GAD65 elevation is rare in MG [10], it does not exclude the diagnosis, which should always focus on the presenting symptoms more than the lab values, given possibility of seronegativity and incomplete test sensitivities.

The importance of symptomatic assessment is crucial to avoid and treat myasthenic crisis. Approximately 13–20% of patients, who experience a myasthenic crisis, have it as their initial presentation upon diagnosis [11–13], and require ICU care and intubation. Thus, it is important to recognize bulbar and respiratory symptoms as signs of impending crisis and to admit these patients to the ICU immediately [14]. In the presented case, the patient had bulbar and respiratory symptoms for weeks prior to crisis, but it was finally set off by emotions and anxiety. This has been described in a previous case report [15], but such presentation is rare and requires prompt recognition. Patients in crisis require IVIG or plasmapheresis, and the treatment modality is based on hospital availability. Current research suggests IVIG and

plasmapheresis to have similar treatment effects [16], and indications of superiority do not yet exist. These patients should also begin immunomodulating therapy with high-dose glucocorticoids and should be considered for additional therapy, such as azathioprine, cyclosporine, or mycophenolate mofetil [8, 17]. If medical management does not achieve desired results, a thymectomy should be considered [18]. Once the patient's respiratory muscle strength is improving, mechanical ventilation weaning should be initiated.

CONCLUSION

In patients with a thymoma, thymectomy is indicated regardless of disease severity. In patients similar to the one presented in this case without evidence of a thymoma, it is recommended to undergo thymectomy in symptomatic MG with positive AchR-Abs and all those presenting with disabling ocular symptoms. The MGTX trial showed thymectomy to be beneficial in symptomatic treatment of MG without thymoma, thus, decreasing the degree of disease severity. While a definitive cure for MG does not currently exist, there are ways to increase patients' quality of life and lower their risk of crisis, when recognized in a timely manner.

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Author Contributions

Christine Sykalo – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Babajide Adio – Conception of the work, Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Sandra Birchem – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Verbal informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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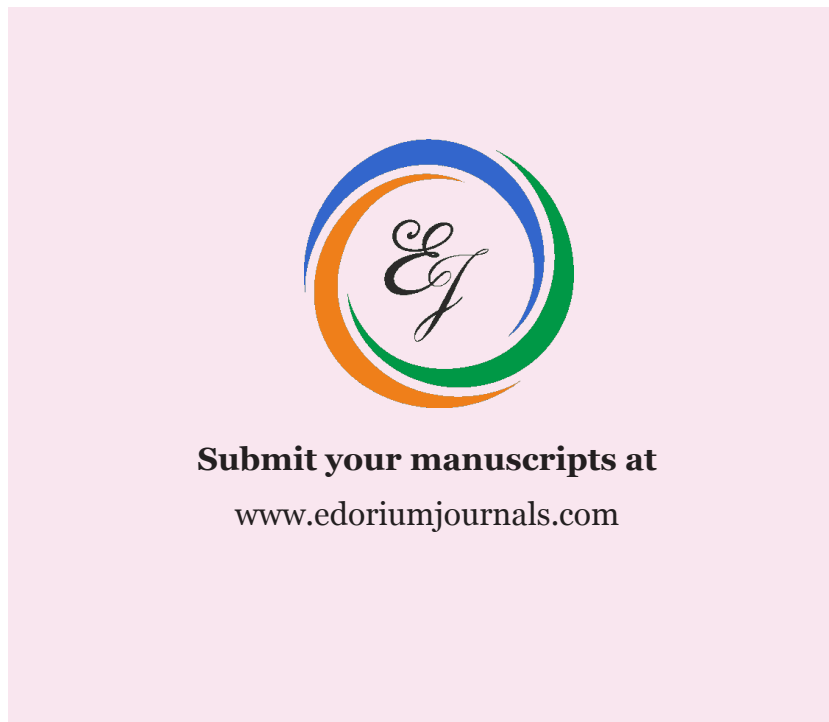
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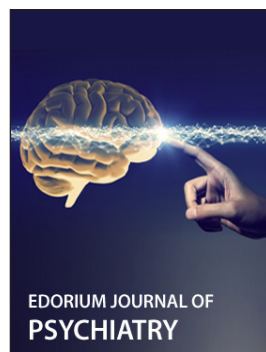
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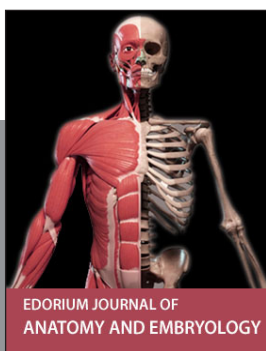
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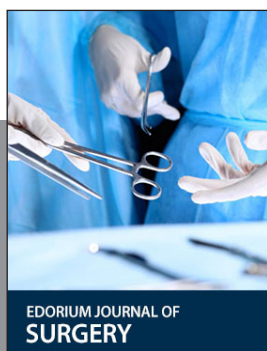
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