Myasthenia Gravis Presenting As Stridor After Aspiration: A Case Report

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Citation

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Abstract

We report a case of bilateral vocal cord paralysis due to myasthenia gravis. His initial presentation was acute respiratory failure requiring tracheotomy. He was diagnosed and treated promptly soon after admission. Although an uncommon cause, myasthenia gravis should be included in the differential diagnosis of stridor.

INTRODUCTION

Bilateral vocal fold paralysis caused by myasthenia gravis is a very uncommon cause of acute upper airway respiratory failure. If it is not treated promptly, poor outcome may occur. We report a case of a 21-year-old man presenting with stridor requiring an emergency tracheotomy. We aimed to discuss differential diagnosis and an overview literature.

CASE HISTORY

A 21-year old man was admitted to the emergency department because of stridor and respiratory distress in December 1999. He had been in good condition until one hour earlier, when aspiration of food developed, despite complaints of muscle weakness and difficulty in swallowing recently.

On initial examination the blood pressure was 110/70 mmHg, pulse was 120, respirations were 40. The patient was cyanotic. Stridor, intercostal and suprasternal retractions were observed. Vocal cord was fixed in paramedian position and velopharengeal failure was observed upon larynx examination. The examination revealed no food in the mouth and no neck masses. Diminished respiration sounds were heard in both lungs. It was decided to perform an emergency tracheotomy. An endotracheal canule was inserted and ventilation assistance was provided. Respiratory failure was improved and blood gases were reverted to normal range. After the first aid the patient was transferred to the department of otolaryngology.

On examination in normal condition, ptosis and weakness of the proximal limb muscles were observed without loss of reflexes or impairment of sensation or other neurologic function. Lactate dehydrogenase level was high (660 U per liter), but there were no abnormal results on routine laboratory investigations and thyroid function tests. Antinucleer, mitochondrial, and smooth muscle antibodies were also normal.

Computed tomography scanning of the cranium and neck revealed no abnormalities that related with this condition. A chest radiograph was normal. We suspected miastenia gravis on the basis of weakness and fatigability in the typical distribution described above. This was confirmed by a positive edrophonium test performed by a consultant neurologist. Repetitive nerve stimulation electric shocks were delivered at the maxillary branch of the left facial nerve and activation potentials were recorded from the left nasal muscle. There was a rapid reduction in the amplitude of the evoked responses of more than 15 percent in repetitive nerve stimulation. Anti-AchR antibodies were detected as high in the serum.

After the diagnosis was confirmed, treatment was begun with oral pyridostigmine 60 mg three times daily, and oral prednisolon 30 mg one time daily. A mediastinal MR scan obtained with the administration of contrast material, showed a retrosternal mass without contrast and smooth margin in 4 x 6 x 7 cm sizes. The patient was transferred to the department of thoracic surgery and thymectomy was applied. Histological findings confirmed the diagnosis of thymoma.

On the tenth day of admission, the vocal folds were mobile and he could breathe comfortably with the tracheotomy tube occluded. So it was decannulated. He was discharged for follow-up in both neurology and ENT clinics one month after admission.

The patient returned to his home on the other side of country. On review one year later, Azothiopurin 100 mg per day had been added his medical therapy to diminish complaints of generalized body weakness. Unfortunately, myastenic criss had been developed and treatment had been carried out in an ICU with invasive mechanic ventilation in spite of the combination of anticholinesterase and immunosuppressive therapy, 6 months later.

DISCUSSION

Numerous acute and chronic neuromuscular disorders may induce acute ventilatory failure. Four different clinical presentations are described by Fitting et al.: slowly progressive (Duchenne muscular dystrophy), rapidly progressive (Guillain-Barre syndrome), chronic with exacerbations (myasthenia gravis), and a form consecutive to critical care (critical care polyneuropathy and myopathy) ([1]). Myasthenic vocal cord dysfunction or paralysis is one of the most rare factors among these pathologies. Ten cases of stridor with MG have been reported in the literature and tracheotomy was required in each of these cases except one ([3,4,5,6,7,8,9]).

Myasthenic crisis, or respiratory failure requiring intubation and mechanical ventilation, may be caused by infections, aspiration, physical and emotional stress, and changes in medication ([2]). In our case, vocal cord paralysis was precipitated by food aspiration. Colp et al reported that respiratory arrest was been precipitated by anesthesia with succinylcholine (4). Our patient demonstrated two unusual features: (I) severe upper airway obstruction due to bilateral vocal cord paralysis at first manifestation of myasthenic syndrome; and (II) precipitation of this syndrome following aspiration of food. We tried to relieve his vocal cord paralysis and other symptoms initially in our myasthenic patient. So tracheostomy was applied to provide enough respiration. Interestingly, Todisco et al reported that vocal cord dysfunction (VCD), presenting with severe inspiratory stridor, was been successfully treated with nasal continuous positive airway pressure (nCPAP), thus giving the medical staff time to make the diagnosis and avoiding intubation or tracheostomy ([10]). It is also reported that a case of myasthenia gravis in a 46-year-old man presenting as acute stridor with bilateral abductor paralysis of the vocal folds which could been treated with pyridostigmine avoided the need for tracheostomy ([11]). This report indicates that it is important to remember the possibility of myasthenia gravis in cases of stridor due to bilateral vocal fold paralysis, since effective medical treatment is available.

The frequency of thymoma associated with autoimmune diseases has been reported to be increased, with 50% of thymoma patients also having myasthenia gravis (MG) ([12]). Venuta et al. reported their 27-year experience with surgical treatment of MG with respect to long-term results and factors affecting outcome. Between 1970 and 1997, they performed 232 thymectomies for MG. According to this trial, the following outcomes were obtained: after a mean follow-up of 119 months, 71% of all patients have improved their clinical status, 18% have a stable disease with no clinical modifications, 5% presented with a deterioration of their clinical status with worse symptoms, required more medications, or both. Six percent died because of MG ([13]). Thymectomy is considered an effective therapeutic option for patients with myasthenia gravis (MG). They concluded that the presence of a thymoma negatively influenced the prognosis (13). After the thymectomy, our patient presented with a deterioration of his clinical status with worse symptoms and needed more medications too.

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