Respiratory Failure due to Vocal Cord Paresis in Myasthenia gravis

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Abstract
This report describes a female patient with myasthenia gravis who developed respiratory failure due to vocal cord paresis. The diagnosis was delayed due to the absence of other myasthenic symptoms (e.g. ptosis, muscle weakness and dysphagia). On direct laryngoscopy, her vocal cords were seen to be in the paramedian position and to move apart after the intravenous injection of edrophonium. The patient initially presented with ocular myasthenia and later returned with isolated respiratory failure. A review of the pertinent literature revealed few reports on myasthenia gravis presenting in this manner.

Introduction
Myasthenia gravis is frequently complicated by respiratory failure, but myasthenic involvement presenting as respiratory distress alone is quite rare. In this report, we present an 82-year-old woman with myasthenia gravis who presented with stridor due to bilateral vocal cord paresis. It is important to consider the possibility of myasthenia gravis in patients with stridor due to bilateral vocal cord paresis.

Case Report
An 82-year-old woman presented to our hospital with dyspnea on January 10, 2000. She had been diagnosed as having ocular myasthenia at 68 years of age, and was started on anticholinesterase at that time. She had no previous history of myasthenic crisis.

On admission, physical examination revealed wheezing over both lungs, but no myasthenic symptoms (e.g. dysarthria, ptosis, dysphagia or generalized weakness). On direct laryngoscopy, her vocal cords were seen to be in the paramedian position and to move apart after the intravenous injection of edrophonium. The patient initially presented with ocular myasthenia and later returned with isolated respiratory failure. A review of the pertinent literature revealed few reports on myasthenia gravis presenting in this manner.
Respiratory Frequency 42/min) with a regular breathing rhythm and narcosis. She was intubated and mechanical ventilation was instituted. She improved following intubation, and could be extubated 6 days later.

On February 25, 2000, she suddenly developed stridor and wheezing. Two days later, she was intubated again for respiratory distress with hypoxia. She improved following intubation, and was extubated 5 days later. In the meantime, she was treated with intravenous antibiotics, steroids, and bronchodilators. Chest X-ray examination was normal, showing no findings that could explain her respiratory distress. Upper airway obstruction associated with stridor was suspected.

The direct laryngoscopy revealed that her vocal cords were seen to be in the paramedian position (fig. 1a), and showed weakness of vocal cord abduction that was caused by paralysis of its abductor, namely, the posterior cricoarytenoid muscle. After the intravenous injection of 3 mg of edrophonium, the vocal cords moved apart (fig. 1b). The patient did not have serum acetylcholine receptor antibodies. CT scans of the neck and chest were normal, showing neither thymoma nor findings that could cause paresis of the larynx. She was treated with ambenonium chloride at 10 mg/day and prednisolone at 5 mg/day, and her condition improved. Five months after the second attack her arterial gases had almost normalized, pH 7.369, PaO<sub>2</sub> 67.7 mm Hg, and PaCO<sub>2</sub> 57.6 mm Hg with room air. She did not undergo thymectomy or plasmapheresis.

**Discussion**

Myasthenia gravis is an autoimmune disorder of the neuromuscular junction caused by antibodies directed against the acetylcholine receptor of the striated muscle end plate [1]. This results in variable muscle weakness which is characteristically made worse by exercise and can frequently be complicated by respiratory failure [2, 3]. In severe myasthenia gravis, involvement of the respiratory muscles, sometimes necessitating mechanical ventilation, is well known [4].

However, myasthenic involvement limited solely to the vocal cords is poorly documented. Only 6 cases of vocal cord paresis associated with myasthenia gravis have been reported in the literature [2, 4–8]. The incidence of vocal cord paresis and upper airway obstruction in myasthenia gravis is unknown [9, 10]. Involvement of the larynx in myasthenia gravis results in weakness of both adduction and abduction of the vocal cords. Clinically, the symptoms are breathy, low-pitched voice, and disorders of phonation. With increasing weakness of abduction, stridor and dyspnea may develop.

The characteristic fatigue of speech and the response to edrophonium strongly support the diagnosis of myasthenia gravis [6]. Edrophonium is the agent commonly used for the anticholinesterase test because of its rapid onset (30 s) and short duration (about 5 min) [11]. If there is unequivocal improvement in an objectively weak muscle, the test result is considered to be diagnostic [12]. In our case, the patient’s vocal cords were seen to move apart on direct laryngoscopy after the injection of 3 mg of edrophonium.

The inspiratory flow-volume loop should be used to evaluate upper airway obstruction in myasthenia gravis with respiratory distress [5]. In an analysis of 61 myasthenia gravis patients, Putman and Wise [13] found 7 patients with spirometric evidence of extrathoracic upper airway obstruction, thus emphasizing the importance of flow-volume loops in the evaluation of their respiratory impairment. We could not perform such measurement on myasthenic attacks because of her unstable
mental conditions. About 6 months later, treatment resulted in a normal curve of her inspiratory flow-volume loop, which means no upper airway obstruction.

Our patient was diagnosed as having tracheobronchitis with bronchospasm at the time of admission, and received steroid therapy. Her first attack of stridor leading to wheezing resulted from infection. After the first extubation, she received steroid pulse therapy, which probably induced the second attack of stridor, since it has been reported that myasthenia gravis may be exacerbated during the initial stages of corticosteroid therapy. However, it is more difficult to explain why our patient’s myasthenia gravis led to severe respiratory failure due to vocal cord paresis. Laryngeal stridor is a life-threatening complication of myasthenia gravis. It is important to consider the possibility of myasthenia gravis in patients with stridor due to bilateral vocal cord paresis.

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References