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Acute inspiratory stridor: a presentation of myasthenia gravis

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Abstract
We present a rare presentation of myasthenia gravis as acute inspiratory stridor in a 16-year-old girl. Prompt diagnosis and medical treatment avoided the need for tracheostomy. Although an uncommon cause, myasthenia gravis should be included in the differential daignosis of stridor.

Key words: Respiratory sounds; Myasthenia gravis; Vocal fold

Introduction

There is general awareness that myasthenia gravis can cause respiratory complications, particularly ventilatory muscle weakness (Hanson *et al.*, 1996). However, laryngeal stridor due to the paradoxical movement of the vocal folds is a rare presentation of myasthenia gravis. Laryngeal stridor as the first manifestation of myasthenia gravis has been reported after exposure to anaesthesia (Colp *et al.*, 1980) and chlorine gas (Foulks, 1981). Myasthenia gravis presenting with laryngeal stridor is important because it may progress rapidly to respiratory failure. We report a case in which the diagnosis was made promptly and successful treatment instituted thereby preventing the development of respiratory failure.

Case report

A 16-year-old presented to the emergency with acute onset of stridor for five hours. General and neurological examination was normal, apart from inspiratory stridor. Fibre-optic laryngoscopic examination showed paradoxical movement of vocal folds (closing of vocal folds on inspiration) with no structural abnormality. The patient was admitted for observation to rule out any ENT pathology. She was getting attacks mostly during the evening and very mild attacks during the day. Between the attacks she was completely asymptomatic.

Myasthenia gravis was suspected when a careful history and clinical examination revealed fatigability becoming worse at the end of the day, mild dysphagia, dysphonic after prolonged speech, fatigability of eye muscles and diplopia at upward gaze. A physician opinion was sought and a tensilon test (intravenous injection of edrophonium 10 mg) was advised. It produced immediate improvement of stridor, speech and dysphagia, confirming the diagnosis of myasthenia gravis. Routine blood tests, ESR and the thyroid function test were normal. Computed tomography (CT) scan of her thorax did not reveal thymoma. Electromyography testing was suggestive of disordered neuromuscular transmission particularly in the eyelid muscles. Autoantibodies to acetylcholine receptor were negative. After the clinical diagnosis was confirmed by the tensilon test, she was started with pyridostigmine 60 mg t.d.s. This abolished her attacks of stridor, and all other symptoms.

She was followed up regularly and the dose of pyridostigmine was increased to 60 mg five times daily. Initially her response to pyridostigmine was good but now it is variable so she is currently being evaluated for the need of steroids and immuno-suppressive drugs.

Discussion

Myasthenia gravis is an autoimmune disorder as a response to a breakdown in T and B cell tolerance to acetylcholine receptors (Steinman and Mantegazza, 1990). In 90 per cent of patients with myasthenia gravis, IgG autoantibodies to the acetylcholine receptor have been isolated (Davidson *et al.*, 1997). Clinically myasthenia gravis is characterized by weakness and fatigability of skeletal muscle after exercise (Davidson *et al.*, 1997). Three forms of myasthenia gravis has been described: 1) The neonatal form; 2) The juvenile form presenting before puberty and 3) The adult form which is the most common form (Garfinkle and Kimmelman, 1982).

Myasthenia gravis is associated with thymoma in only 10 per cent of patients but 50 per cent of patients with thymoma have myasthenia gravis (Neal and Clarke, 1987). Myasthenia gravis can affect skeletal muscle anywhere in the body, including the intrinsic muscles of the larynx (Hanson *et al.*, 1996). Symptoms of stridor and dysphonia resulting from weakness in vocal fold adduction and abduction are uncommon even though laryngeal muscles have a high density of neuromuscular junctions (Neal and Clarke, 1987). In a review of 147 patients with myasthenia, only four patients had stidor at presentation (Calcaterra *et al.*, 1972). Until now, only six reports of stridor as a presenting feature of myasthenia gravis have been

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published in the literature since 1972 (Colp *et al.*, 1980, 1981; Schmidt-Nowara *et al.*, 1984; Fairley and Hughes, 1992; Job *et al.*, 1992; Hanson *et al.*, 1996). Other rare forms in presentation of myasthenia gravis include facial pain (Lloyd and Mitchell, 1988) and swollen tongue (Davidson *et al.*, 1997).

Myasthenia gravis is important in otolaryngology because bulbar muscles are often involved early in the disease (Dhillon and Brookes, 1984). Laryngeal involvement of the recurrent laryngeal nerve causes dysphonia that is worse with prolonged speech. Poor vocal fold adduction presents as breathy voice and poor vocal fold abduction may present as inspiratory stridor or dyspnoea (Davidson *et al.*, 1997). It has been clinically observed in patients with myasthenia gravis (Schmidt-Nowara *et al.*, 1984) that normally abducted position of the vocal folds is reversed during forced inspiration, bringing the vocal folds together to cause stridor (paradoxical movement) as happened in this case.

This demonstrates the diagnostic dilemma of patients presenting with laryngeal symptoms of myasthenia gravis.

Conclusion

It is important to remember the possible diagnosis of myasthenia gravis in cases of acute inspiratory stridor due to the paradoxical movement of the vocal folds since effective medical treatment is available. As laryngeal stridor is a life-threatening complication of myasthenia gravis, prompt diagnosis and treatment can prevent further complications such as respiratory failure. In doubtful cases the tensilon test is a simple, safe and effective diagnostic procedure (Friedman and Goffin, 1966).

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