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Acute stridor due to bilateral vocal fold paralysis as a presenting sign of myasthenia gravis

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Abstract
We describe a case of myasthenia gravis in a 46-year-old man presenting as acute stridor with bilateral abductor paralysis of the vocal folds. Prompt diagnosis and medical treatment with pyridostigmine avoided the need for tracheostomy. 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.

Introduction

Stridor due to vocal fold paralysis is an uncommon presentation of myasthenia gravis. It is important to recognize it because it may progress rapidly to respiratory failure. We report a case in which the diagnosis was made promptly and successful treatment instituted, avoiding the necessity of tracheostomy.

Case report

A 46-year-old man was referred to the ENT department with acute episodic stridor, lasting four or five minutes at a time, for one week. Further enquiry revealed a ten-week history of dysphagia with nasal regurgitation, and more recently diplopia and poor phonation. All his symptoms were worse at the end of the day when he was tired.

Examination showed bilateral ptosis and diplopia with poor facial expression. There was no stridor at rest. Spraying the nose and larynx with 4 per cent lignocaine for fibreoptic examination precipitated an acute attack of stridor. Fibreoptic laryngoscopy showed bilateral abductor paralysis of the vocal folds, with no structural abnormality. The attack of stridor resolved after about five minutes. He was admitted and a neurological opinion was sought. A Tensilon test (intravenous injection of edrophonium 10 mg) produced an immediate improvement in muscle power, with return of normal facial expression and phonation, confirming the clinical diagnosis of myasthenia gravis. Further confirmatory tests included autoantibodies to acetylcholine receptor sites, and single fibre stimulation electromyography which showed an increased jitter pattern. A CT scan of his thorax revealed a thymoma.

He was treated with pyridostigmine 60 mg five times a day. This abolished his attacks of stridor and all other symptoms. He later underwent a successful thymectomy and remains well on treatment with pyridostigmine. Histology revealed a thymoma.

Discussion

Myasthenia gravis is an autoimmune disorder resulting from a breakdown in T and B cell tolerance to acetylcholine receptor (Steinman and Mantegazza, 1990). This results in variable muscle weakness which is characteristically made worse by exercise.

Stridor due to vocal fold paralysis is an uncommon presentation of myasthenia gravis. In a review of the otorhinolaryngological manifestations of 147 patients with myasthenia, only four had stridor at presentation (Calcaterra *et al.*, 1972). A further six of these 147 patients developed stridor later in the course of their disease. Only three cases have been reported in the literature since 1972 (Colp *et al.*, 1980; Foulks, 1981; Schmidt-Nowara *et al.*, 1984). In each of these, the diagnosis was delayed until the onset of severe generalized muscle weakness, and either tracheostomy or unnecessarily prolonged medical treatment ensued.

Our patient demonstrates that myasthenia gravis may present as stridor. When a careful history was taken, it became apparent that other bulbar muscle groups were also affected, and that the symptoms were worse when tired at the end of the day. This, together with the physical findings of facial muscle weakness and the bilateral abductor paralysis, was sufficient for a clinical diagnosis of myasthenia gravis. The positive response to edrophonium confirms the diagnosis.

Current medical treatment of myasthenia gravis is generally successful (Fonseca and Havard, 1990). Anticholinesterases are the mainstay of symptomatic treatment. Pyridostigmine acts by inhibiting the breakdown of acetylcholine at the neuromuscular junction by acetylcholinesterase. Therapeutic levels vary widely and have to be optimized to the individual patient (Breyer-Pfaff *et al.*, 1990). A more fundamental therapeutic approach is treatment of the underlying autoimmune state. Steroids and immunosuppressive drugs such as azathioprine and cyclosporin A have been used, but in most cases thymectomy is preferable (Fonseca and Havard, 1990).

Respiratory problems in myasthenia are more often due to respiratory muscle paralysis than vocal fold palsy (Mier *et al.*, 1990). Most of the cases described in the literature in which stridor occurred have needed tracheostomy. This is probably due to delayed diagnosis. It is important to remember the possibility of myasthenia in cases of stridor due to bilateral vocal fold paralysis, since effective medical treatment is available.

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