Paradoxical vocal cord adduction in an adolescent with cystic fibrosis

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Abstract

Many patients with cystic fibrosis have symptoms of dyspnoea and wheeze which are responsive to treatment with bronchodilators. An adolescent woman with cystic fibrosis is described who presented with inspiratory stridor and in whom the classical features of paradoxical vocal cord adduction were found.

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Paradoxical vocal cord adduction is a very rare functional disorder caused by inappropriate adduction of otherwise normal vocal cords. Typically, it affects women under 40 years of age who often have a background of employment in health care. The upper airway obstruction associated with this condition may be mistaken for asthma and symptoms may persist in spite of varying therapeutic interventions. We describe here an adolescent woman with cystic fibrosis with the classical features of paradoxical vocal cord adduction—an association, to our knowledge, not previously described.

Case report

A 17 year old adolescent woman was admitted in July 1993 for assessment of chronic, predominantly dry, daytime and nocturnal cough with mucus production. Cystic fibrosis was diagnosed at birth and she attended the Adult Cystic Fibrosis Unit at St Vincent's Hospital, Dublin from 1991. Features of her illness included chronic bronchiectasis, multiple nasal polyps, pancreatic insufficiency, and recurrent meconium ileus equivalent. Her sputum was
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In 1992 she was admitted with newly acquired symptoms of wheeze and cough. At that time she was found to have a significant reduction in forced expiratory volume in one second (FEV₁) of 1.77 l (50% pred) with a vital capacity (FVC) of 2.71 l (73% pred) and an 18% improvement in FEV₁ after inhalation of salbutamol, consistent with a diagnosis of asthma. She was commenced on inhaled corticosteroids, inhaled bronchodilators, and oral theophylline. This resulted in considerable improvement in her symptoms of wheeze but the cough persisted. In the summer of 1992 she developed prominent hoarseness and noisy sounds in her throat. There was no evidence of oropharyngeal candidiasis or urticaria. The addition of oral corticosteroids did not result in any clinical improvement and she was admitted for assessment. Following admission she was noted to have considerable inspiratory stridor with minimal expiratory wheeze. Lung function measurements of expiratory volumes and flow rates were surprisingly normal with an FEV₁ of 3.05 l (95% pred), FVC 3.65 l (101% pred), and a carbon monoxide transfer factor (TLCO) of 23.18 (78% pred). However, examination of the flow–volume loop showed significant persistent inspiratory upper airways obstruction (figure). Considerable paradoxical adduction of the vocal cords was found on bronchoscopy carried out under local anaesthesia. She was prescribed codeine phosphate linctus with symptomatic relief. Initial assessment by the clinical psychologist attached to the unit confirmed the patient's desire to pursue a career in health care but did not reveal any psychological abnormality. Long term psychological assessment was declined by the patient's family.

Discussion
Paradoxical vocal cord adduction is an unusually rare condition due to inappropriate adduction of the vocal cords.¹ It usually affects young adult women working in health care-related jobs. While functional airways obstruction is most often the result of inappropriate adduction of the vocal cords, it may also be caused by inappropriate movement of the pharyngeal wall muscles.² It is considered to be an hysterical conversion reaction which may be responsive to a wide range of psychological therapies.³ Although initial reports suggested that it was a discrete isolated disorder, we have recently described a number of patients in whom paradoxical vocal cord adduction co-existed with documented asthma, and in whom objective evidence of refractory symptomatic paradoxical vocal cord adduction persisted for over a decade in spite of a wide range of psychotherapeutic interventions.⁴ This present report further extends the spectrum of pulmonary disorders in which paradoxical vocal cord adduction may be found.

Many patients with cystic fibrosis have symptoms of wheeze and shortness of breath with a positive response to inhaled bronchodilators which may vary with pulmonary exacerbations of the disease.⁵ To our knowledge there have been no previous reports of paradoxical vocal cord adduction in patients with cystic fibrosis. One of the puzzling psychodynamic features of this disease is its virtually exclusive occurrence in young women, the majority of whom have been involved in health care-related occupations. Our patient, like many individuals with cystic fibrosis, had been introduced to the hospital milieu from an early age and had many hospital admissions. Interestingly, the patient reported here had been heavily involved in life-saving courses, had spent considerable time as a beach lifeguard, and had expressed a desire to be a medical laboratory technician. This functional disorder is obviously rare, but it is clear that it could be easily overlooked in patients with cystic fibrosis who may present with various disorders of the upper and lower airways including recurrent respiratory infections, asthma, oropharyngeal candidiasis, and dysphonia due to inhaled steroid. We believe that the diagnosis of paradoxical vocal cord adduction should be considered in patients with cystic fibrosis in whom atypical chest symptoms, hoarseness, or stridor persist, and we suggest that appropriate physiological and endoscopic assessments are carried out.

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