Chronic sclerosing tracheobronchitis

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SUMMARY We report a child with wheezing and respiratory distress of 53 months duration which was undiagnosed and unresponsive to treatment. Necropsy examination of trachea and bronchi showed narrowing by chronic inflammation and fibrosis of mucosa and submucosa. Despite intensive investigations, the aetiology and pathogenesis remained undetermined.

Case history

The child, who weighed 3940 g at birth, had otitis media and pharyngitis accompanied by watery rhinorrhea and a ‘chronic’ cough from age 3 months. At 17 months she began to have episodes of laryngitis, coughing, and wheezing which recurred almost weekly. At 25 months of age bronchoscopy showed appreciable inflammation of the hypopharynx and larynx.

She was admitted to hospital at age 31 months because of the rapid onset of fatigue and breathlessness with exertion. Wheezing was reportedly provoked by irritants, exercise, and emotions. Radiographs of the chest before admission showed ‘increased markings’ but were otherwise normal. Treatment with bronchodilators, corticosteroids, and antibiotics had not altered the course of illness.

Her height was at the 10th centile and weight was less than the third centile for age. Abnormal physical findings were shortness of breath, the presence of thick mucus in the pharynx, diffusely decreased breath sounds, and rhonchi.

The patient did not have typical reversible bronchospasm; she always had some degree of respiratory distress characterised by intercostal recession, continuous rhonchi, periodic wheezing, decreased breath sounds, and a productive cough. There were three episodes of respiratory arrest in hospital, apparently caused by obstruction from secretions.

Cultures of tracheal and bronchial contents and tissues (viral, fungal, aerobic, and anaerobic) showed *Staphylococcus aureus*. Alpha, antitrypsin was 200 mg/dl; cold agglutinins were negative; IgE mediated skin tests were negative; cell mediated immunity skin tests showed a positive reaction to tetanus and candida antigens; and sweat chloride was normal. Fungal serology was negative. Immunoglobulins and parotid IgA were normal for age. Nitro blue tetrazolium reduction by neutrophils was normal and the results of chemotaxis assays and tests for lymphocyte number and function were within the normal range.

Bronchoscopies were performed on 8 occasions, initially in an attempt to determine a diagnosis but later in order to remove secretions which the patient could not eliminate herself. The earlier bronchoscopies showed erythema and friability of the distal trachea and mainstem bronchi with near total occlusion of the left mainstem bronchus. Biopsy of the stenotic area showed granulation tissue. *Staphylococcus aureus* was treated with dicloxicillin sodium. On day 306 after admission to hospital and at age 4 years 8 months an episode of respiratory arrest was quickly followed by a cardiac arrest and attempts at resuscitation failed.

Pathological findings

The necropsy was limited to examination of the pulmonary system. The larynx and lungs were unremarkable but the lumens of the upper airway were reduced to as little as 3 mm in diameter by concentric mural thickening. Sections of the larynx showed squamous metaplasia and submucosal, chronic inflammation. Step sections of trachea and mainstem bronchi were examined; the lining was squamous metaplasia or denuded epithelium. The lamina propria was considerably thickened and was composed superficially of granulation tissue moderately to densely infiltrated by lymphocytes and plasma cells, scattered mast cells, and rare eosinophils and neutrophils; beneath was a thick layer of densely collagenised fibrous connective tissue, extending to the underlying cartilage. Mucosal ducts and glands were reduced in number. Smooth muscle and elastic fibres were reduced in number or absent. The underlying hyaline cartilage was intact. The adventitia was unremarkable.

Review of the tracheal and bronchial biopsies before death showed the same type of granulation tissue as that found in the lower trachea at necropsy. The lungs were unremarkable with the exception of...
The anatomic lesion was an unusual diffuse, chronic fibrosing tracheobronchitis which, strikingly, stopped short of the intrapulmonary airways. Review of the many biopsy specimens obtained during life showed essentially the same inflammatory process indicating that this had been established early in life. It is easy to understand why this child had difficulty with stridor and clearing respiratory tract secretions and it is surprising that there was no evidence of pneumonia, since the respiratory tract's normal mucosal defenses were inoperative.

The cause of the inflammation is unknown. No foreign objects were found and it seems unlikely that any localised irritant could generate such diffuse inflammation and scarring over such a prolonged period of time. It is possible that this lesion was the consequence of especially severe or repeated episodes of infectious tracheobronchitis. During life, Staphylococcus aureus was the only pathogen cultured from the respiratory tract; this organism probably caused a 'super' infection and antibacterial treatment in large doses did not improve the patient's course or ameliorate the pathological findings.

The possibility of a primary deficiency of tracheobronchial cartilage as seen in Williams-Campbell syndrome was considered. Any effect on the patient's cartilage seemed, however, to be secondary to inflammatory changes rather than primary.

Other conditions considered included laryngotraceobronchitis with a predominantly mononuclear infiltration as has been described at necropsy in sudden infant death syndrome; tuberculosis and non-tuberculous mycobacterial infection as a cause of chronic endobronchitis in children; non-specific infiltration of bronchi by lymphocytes referred to as 'mural bronchitis' by Sinclair-Smith et al; and gastroesophageal reflux and aspiration. Neither the clinical nor pathological descriptions of any of these illnesses were, however, compatible with those in this child.

Henry et al recently reported 7 children with pseudomembranous croup who were acutely ill. Cultures of airway secretion in five of the 7 children yielded Staphylococcus aureus. The major difference between the case reported here and the patients of Henry et al is chronicity.

An immunological defect in this child was always an attractive explanation. Every available immunological test for humoral and cellular immunity was performed but results were within the normal range. None of the physiological, immunological, microbiological, or biochemical data collected showed the aetiology and we have thus been left with a description of a progressive lesion which did not respond to any specific or empirical treatment.

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References


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