Progressive tracheobronchial polychondritis: need for early diagnosis

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Tracheobronchial chondritis is an unusual presenting feature of relapsing polychondritis carrying a poor prognosis.¹ We report a case of rapidly progressive tracheobronchial chondritis leading to tracheal collapse within two months of presentation that emphasises the need for early diagnosis and treatment.

Case report

A 71 year old woman, an ex-smoker and previously well, presented in late June 1983 with a six week history of productive cough, hoarseness, and arthritis of the right knee. Examination showed a harsh expiratory wheeze when she coughed and laryngobronchoscopy showed supraglottic mucosal swelling and a cobblestone appearance of the trachea and major bronchi. Histological examination of the tracheal mucosa showed nothing abnormal and cultures of bronchial aspirate were negative.

At the end of July 1983 she developed breathlessness, wheeze, inspiratory stridor, and dysphagia, with a weight loss of 1 kg. A barium swallow was normal. Tinnitus was present and she was found to have bilateral mixed conductive and high tone (8000 Hz) nerve deafness. A perforation of the nasal septum was noted and shortly after this she developed painless collapse of the nasal bridge. Biopsy of the nasal septum showed the presence of active chondritis and confirmed the diagnosis of relapsing polychondritis. Tenderness and swelling of the proximal interphalangeal joints of both hands were noted. There was no clinical evidence that the rib cage was affected.

The erythrocyte sedimentation rate was 68 mm in one hour, haemoglobin concentration 10.4 g/dl, and white blood count $9.0 \times 10^9/l$ (with normal differential count). The serum urea and electrolyte concentrations were normal; total serum protein was 63 g/l and albumin 31 g/l. The results of the Rose Waaler, latex, antinuclear factor tests were negative and antibodies to gastric parietal cells, pancreatic islets, smooth muscle, and mitochondria were absent. Venereal disease research laboratory and *Treponema pallidum* haemagglutination assay tests gave negative results. IgG antibodies to human fetal cartilage matrix were detected by immunofluorescence (titre 1/16). A chest radiograph showed normal lung fields with tracheal narrowing evident on tomography.

There was a greatly reduced FEV₁ of 0.6 litre (40% predicted) and a normal forced vital capacity (FVC) of 1.7 l (80%), with a low airways conductance of $0.66 \text{ s}^{-1} \text{ kPa}^{-1}$

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Flow-volume loop. $\dot{V}_{E_{\max}}$ and $\dot{V}_{I_{\max}}$ —maximum expiratory and inspiratory flow; TLC—total lung capacity.

(31%), indicating severe airflow obstruction. Both the residual volume (2.23 l—211%) and the total lung capacity (TLC) (4.03 l—131%) were increased. The single breath diffusing capacity and resting arterial blood gases were normal. A flow-volume loop (fig) showed considerable reduction in flow rates, particularly on expiration, suggesting that the major airflow limitation was intrathoracic. An abrupt decrease in flow on expiration near TLC was thought to represent dynamic intrathoracic airways compression. Repeat laryngobronchoscopy in August 1983 confirmed the presence of intrinsic narrowing of the trachea throughout its length and dynamic tracheal collapse on expiration with prolapse of the posterior wall.

Treatment was started in early August with prednisolone 50 mg and dapsone 200 mg. The patient remained well during the first three weeks of treatment but after this her clinical course deteriorated with increasingly frequent, severe, and prolonged episodes of dyspnoea. Surgical splinting of the major airways was attempted in September 1983 but death occurred several days after the operation. Permission for necropsy was refused. A specimen of trachea obtained at surgery showed intense chondritis with dissolution of cartilage rings.

Discussion

Laryngeal and tracheobronchial complications of relapsing polychondritis have been adequately described,²⁻⁴ although little is known about peripheral airways disease.

Progressive tracheobronchial polychondritis

Patients presenting with disease of the respiratory tract have a worse prognosis than those who develop it later in the course of their disease, and progression of such disease may not be influenced by corticosteroids.¹

An autoimmune basis for relapsing polychondritis seems likely. Antibodies to cartilage and to type II collagen have been demonstrated⁵ and the observation that antibodies to the latter occur mainly in patients with active disease suggests an important role for antibody in this disease. Furthermore, transient neonatal polychondritis has been described in a child born to a woman with polychondritis, suggesting that factors which cross the placenta may play a part in the pathogenesis of the disease.⁶ The ability of cartilage antigens to transform lymphocytes from these patients suggests that cell mediated immunity may be important.⁷

Our patient's disease had an unusually aggressive course, with rapid progression to tracheal collapse within two months of presentation despite treatment with corticosteroids. One similar case has been reported,³ although abrupt deterioration in this instance followed tracheal biopsy.

Reports of the use of immunosuppressive agents in relapsing polychondritis is limited to a few cases.^{1,8} If this is an autoimmune disease early use of immunosuppressive agents in the more aggressive cases might prevent rapid cartilage dissolution. The detection of cartilage antibody in active disease⁹—in our case as early as 10 weeks from the onset of symptoms—suggests that measurement of this antibody in a patient showing possible signs of relapsing polychondritis may lead to swift recognition. Furthermore,

since antibodies seem to be important in the pathogenesis of this disease, plasmapheresis may have a role in the management of these patients.

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