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Idiopathic tracheal stenoses

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Abstract
The case histories are presented of three patients with circumferential subglottic stenoses who presented, over a six-month period, to a teaching hospital's Otolaryngology department. No recognisable cause for their subglottic stenoses was found. Traumatic, iatrogenic, infectious and specific inflammatory processes were excluded. The histopathological evidence suggested a chronic inflammatory process. All patients were treated by resecting the stenosis with a carbon dioxide laser. One patient required an emergency tracheostomy for airway obstruction. A review of the published literature on this rare condition is given.

Key words: Tracheal stenosis; Laser surgery

Introduction

Fibrotic stenosis can occur in the subglottic larynx as a result of external trauma or from endotracheal intubation. However, idiopathic subglottic stenosis is extremely rare and is generally thought to be the result of an inflammatory process.

Over a three-month period three cases of circumferential subglottic stenosis presented to our department and despite extensive investigation no cause has been found for these stenoses. All were treated by endoscopic excision of the stenotic area employing a carbon dioxide (CO_2) laser. In one patient where there was a rapid recurrence of the stenosis a Montgomery T-tube has been inserted. All three patients are still receiving treatment.

Case studies

Case 1

The patient was a 50-year-old woman with increasing stridor of four to five year duration. Her past medical history included possible Crohns disease treated by sulphasalazine 1 g t.d.s. The finding was circumferential subglottic stenosis. She was treated by numerous laser resections of the stenotic area. The patient had been offered tracheostomy with the insertion of a Montgomery T-tube but prefers to have the stenosis reduced at intervals of two to three months.

Case 2

A 35-year-old woman presented with increasing inspiratory stridor over three years. Marked subglottic stenosis was evident on X-ray and fibreoptic examination. The airway was severely compromised at presentation and a tracheostomy was performed. At direct laryngoscopy, extensive subglottic stenosis was found and this was removed with the CO_2 laser but the recurrence of the stenosis was rapid and after further laser excision and dilatation a Montgomery T-tube was inserted and left in place, currently for 12 months.

Case 3

A 46-year-old woman presented with increasing strido for one month. She had angioneurotic oedema when aged 27 years which resulted in moderate tongue swelling. Marked subglottic stenosis was evident at direct laryngoscopy. She underwent excision and vaporisation of the stenosis with the CO_2 laser and has been maintained with a satisfactory airway by further removal of the stenosis at three monthly intervals for the last 15 months.

Histopathology

This was essentially the same in all three patients and no diagnostic features were evident.

All three showed similar features which consisted of dense fibrous tissue with scattered groups of chronic inflammatory cells. The fibrous tissue showed some vascularity and contained active proliferating fibroblasts set in a collagenous stroma.

The inflammatory infiltrate was composed mainly of reactive plasma cells together with lymphocytes, macrophages and scattered eosinophils. Some of the latter cells showed degranulation. The inflammatory cells were mainly perivascular in location; neutrophils were conspicuously absent.

In *Case 1* four biopsies were performed over a nine month period. Although the features of the initial biopsies resembled those of the other cases, in later specimens there was granulation tissue formation at the surface, evidence of haemorrhage and some deposits of foreign material within the tissues. These changes were all considered to be related to the surgical procedures.

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Discussion

To be termed idiopathic subglottic stenosis requires the careful exclusion of all known and documented causes of tracheal stenosis.

Although it may be possible that a single cause may be responsible for this group, it seems unlikely as similarities are so few that a single origin seems unlikely. It is possible that over time symptoms consistent with conditions causing tracheal stenosis such as relapsing polychondritis or Wegener's granulomatosis will develop.

Perhaps the commonest cause of subglottic stenosis in adults is as a result of prolonged intubation for ventilation purposes and this has been well documented throughout the world literature. Other causes include external trauma to the trachea, inhalation burns and irradiation. Tracheal stenosis may also result from infective causes including bacterial tracheitis, tuberculosis, histoplasmosis, diphtheria and scleroma.

Specific inflammatory conditions which have been associated with tracheal stenosis include Wegener's granulomatosis, relapsing polychondritis, systemic lupus erythematosus and amyloidosis.

Wegener's granulomatosis is a condition of unknown aetiology characterised by a necrotizing granulomatous inflammation and vasculitis. It classically affects the upper airways, lung and kidneys. It is thought to be immunologically mediated.

Subglottic stenoses are found in patients suffering from Wegener's granulomatosis and often in patients 20 years of age or younger. The overall incidence of subglottic stenoses in this condition has been estimated and reported as 8.5 per cent (Waxman and Bose, 1986).

Measurement of circulating antineutrophil cytoplasmic antibodies (ANCA) has been shown to be useful in diagnosing Wegener's granulomatosis in this group of patients. Gans *et al.* (1991) identified positive ANCA tests in five patients with subglottic stenosis, in whom two had no known involvement of other organs. This test was negative in all three patients.

Pathological interpretations of biopsy specimens from the trachea can be difficult since diagnostic features of granulomatous inflammation, parenchymal necrosis and vasculitis may be inconspicuous and are found in the minority of specimens (Lebovics *et al.*, 1992).

Otherwise when a patient has undergone a previous surgical procedure on the trachea, the possibility of a foreign body giant cell reaction should be considered especially if granulomas are observed on biopsy (Devaney *et al.*, 1990).

Relapsing polychondritis is a rare disease which often presents initially to Otolaryngological departments. It is characterised by recurrent inflammation and degeneration of cartilage, commonly the pinnae, nose, trachea, larynx and joints. Involvement of major airways is more common in young patients and can be more serious than in adults (Michet *et al.*, 1986). When relapsing polychondritis affects the tracheal cartilages, total collapse can occur often resulting in the rapid onset of stridor.

Histology of a tracheal stenosis resulting from relapsing polychondritis depends on the stage of the disease, but in the active phase consists of a loss of basophilic staining of the cartilage matrix, perichondrial inflammation and later cartilage destruction with fibrous replacement (Clark *et al.*, 1992).

Systemic lupus eythematosus (SLE) is another disease of uncertain aetiology and is characterised by inflammatory lesions involving multiple organ systems, the skin, kidneys, and mucous membranes.

Smith and Ferguson (1974) describe a case of subglottic stenosis in 63-year-old female with SLE. They concluded that the stenosis was best managed by tracheostomy if warranted and immunosuppressive drugs. They felt that the stenosis was incurable, but was never severe enough to cause the death of the patient with SLE.

Once all the other possible causes of tracheal stenosis have been excluded, then a diagnosis of a truly idiopathic tracheal stenosis can be made. Grillo *et al* (1993) have published their series of 49 patients with idiopathic tracheal stenoses, this group included both subglottic and tracheal stenoses. We have found histopathological findings essentially similar to those reported in this paper although they found that the surface epithelium of their specimens often showed evidence of squamous metaplasia and areas of epithelial ulceration.

Grillo *et al.* (1993) found only 14 of their 49 patients were unsuitable for surgical resection and reconstruction and were treated along the same lines as our smaller group of patients. Thirty-five of their 49 patients were treated by a single stage resection and reconstruction. Of these, one required a permanent tracheostomy, one patient required the use of a Montgomery *t*-tube and two needed regular dilatation.

Brandenburg (1972) described three cases of idiopathic subglottic stenoses seen over a 10-year period. Two cases were complicated by retro-orbital pseudotumour. Further case reports of idiopathic tracheal stenosis have been reported (Mikaelian, 1974; Jazbi *et al.*, 1977; Havas *et al.*, 1984). Treatment regimes in these reports consisted of tracheostomy followed by laryngofissure and stenting or repeated laser treatment.

There may exist a relationship between idiopathic tracheal stenosis and other forms of idiopathic fibrous proliferation or to retroperitoneal fibrosis. This relationship may be as a result of immunological hyperreactivity (Hanley *et al.*, 1984; Osborne *et al.*, 1987; Kelly and Hwang, 1989).

Adult subglottic stenosis appears to be a rare condition and recognition of the possible causative factors will help in its diagnosis and treatment, it is only after the failure of these exhaustive efforts to find a cause for the stenosis, can it justifiably be labelled an idiopathic tracheal stenosis.

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