New approaches to the management of subglottic stenosis in Wegener's granulomatosis

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OBJECTIVES

1. Describe new modifications in the management of laryngeal manifestations of Wegener's granulomatosis (WG).
2. Present a conservative yet aggressive method for long-term management of subglottic stenosis.
3. Present and demonstrate how tube-free speech-ready tracheostomy can be applied to the more challenging cases of WG.

The specific etiology and pathophysiology of subglottic stenosis in Wegener's granulomatosis is not clearly understood. It is postulated that this segment of the airway is particularly vulnerable to pathological processes affecting the microcirculation, as it is a junction between two embryological growth centers. Support for this argument is found in the anatomy of the stenosis being localized to the subglottic region at the junction of two microcirculations. The true vocal cords and the trachea above and below the stenotic ring are almost completely spared, which further reinforces the microcirculation hypothesis. This ring-like inflammatory process develops approximately 1.5 to 2 cm below the true vocal cords and above the trachea resulting in progressive airway obstruction. The normal mucociliary transfer system is disrupted and tracheobronchial secretions are trapped at the level of the stenosis and below it, resulting in secondary infections. The forced airflow through this narrow segment creates turbulence, which is aggravated by coughing and attempts to clear secretions. The overall effect is to further irritate the involved tissue, precipitating a vicious cycle that may progress to life-threatening airway obstruction.

The subglottic stenotic lesion is membranous in most cases and is limited to a short or narrow segment of the lower larynx. Similar stenotic lesions may appear at the junction between the trachea and the right or main bronchi and sometimes even further down the tracheobronchial tree, at other areas where different microcirculations meet. In some instances, the stenotic process partially or completely resolves with systemic treatment such as steroids and immunosuppressant medications. In more stubborn cases, a local surgical intervention can reestablish a stable and secure airway and promote local healing that may control and possibly prevent persistent or recurrent stenosing processes. Contrary to treatment of laryngotracheal stenosis caused by different etiologies such as trauma, local treatment of subglottic stenosis in WG must also focus upon subduing or reversing the manifestations of the systemic disease. The surgical method of treatment must also take into account the unique nature of the pathological process, which is predominantly caused by vasculitis. Certain surgical modalities such as laser surgery, stents, and reconstructive procedures have higher tendencies to aggravate circulation disorders and may be contraindicated. Given their rheumatologic diagnosis and the systemic medications taken for it, many of these patients are not candidates for complex reconstruction or resection in any case.

Every possible measure should be taken to restrict and avoid potential damage to the vocal cords, the trachea, the bronchial tree, and the lungs in the course of the surgery and anesthesia. We present the current methods of treatment conjointly adopted by the Departments of Rheumatology and Otolaryngology at The Cleveland Clinic Foundation (CCF). The principles follow the original guidelines laid by Drs. Lebovics and Hoffman during their tenure at the NIH in Washington, DC. Certain technical modifications have been adopted over time.

Patient population

The patients differ according to their degree of subglottic involvement by the systemic disease, the stage of the disease, and, when applicable, previous treatments. Fiberoptic laryngotracheal bronchoscopy determines the degree and extent of the involvement at the level of the subglottic segment of the airway. Computed tomography

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performed in the axial and tangential planes is performed, especially in those chronic and previously treated cases that are more complicated. Consultation and coordination with the treating rheumatologist determines the scheduling and planning of surgical interventions. On the rare occasions when patients present with acute life-threatening airway obstruction even before the final diagnostic process is completed, emergent intervention is performed. Otherwise, we usually coordinate a thorough and prompt effort to establish the diagnosis and detailed clinical findings while the patient is prepared for surgery. These efforts include physical examinations, radiologic exam as described above, and appropriate laboratory studies. Prior planning with the anesthesiologists is imperative to ensure uncomplicated success in these sometimes very challenging cases.

■ ENDOSCOPIC TREATMENT

Using a specially designed articulating laryngoscope with ports for jet ventilation, the larynx is exposed and the patient is maintained under general anesthesia utilizing jet ventilation. Photographic documentation of the larynx and trachea is routinely achieved. In initial or virgin cases, a biopsy is taken to verify the diagnosis. Telescopic or flexible fiberoptic endoscopy is performed to assess the full extent of the tracheobronchial tree and rule out additional lesions. The stenotic lesion is injected submucosally with a long-acting corticosteroid suspension such as methylprednisolone. The solution is infiltrated along the submucosal-perichondrial plane. A total of 80 mg is injected. Under microscopic magnification, four to six lysing longitudinal incisions are made in a star-like fashion, employing sharp metal microlaryngeal blades. These incisions release the constricting stenotic ring and break it up, widening the diameter of the airway and simultaneously preserving islands of intact mucous membrane between the incisions. This epithelium is intended to regenerate and re-surface the expanded lumen. Progressive serial dilatations are performed using semi-rigid, flexible, smooth Malloney dilators lubricated by topical steroid cream. If bleeding occurs, it is controlled by topical application of 1:1,000 adrenaline, and the lesion and tracheobronchial tree are irrigated. The next stage of the procedure involves repeated topical applications of mitomycin-C over a 6-minute period with the intent to further inhibit fibrosis and re-stenosis. The airway is suctioned out and cleared of reactive secretions and blood. Finally, 4% Lidocaine is squirted down the tracheobronchial tree, to act topically to prevent postsurgical reactive laryngospasm as the patient is awakened. A bolus of systemic steroids is administered intravenously during the procedure. The procedure is performed in full coordination with the anesthesiologists throughout each stage. Postoperatively, the patients receive respiratory treatments in aerosol form for the twelve hours after the procedure and are discharged the following morning.

Throughout this group's experience, we have never used laser surgery on subglottic stenosis caused by WG. Incidentally, the remote cases in which patients were treated with laser surgery in other institutions prior to their referral to the CCF developed complicating secondary stenosis that required more extensive surgical intervention to overcome the severe secondary superimposed damage, most probably induced by management with laser and other surgical methods. These patients required laryngotracheal reconstructive procedures, or had to undergo establishment of permanent tracheostomies. In our experience, then, laser therapy is contraindicated is this patient population.

Results

To date, over fifty patients have been managed with this endoscopic dilatation procedure. Twenty-six have rheumatologic diagnoses, twenty-five with WG and one with pemphigoid. Thirty patients have needed repeated similar procedures. Only six patients suffering from WG with severe and complicated subglottic stenosis, all complicated or aggravated by unsuccessful surgical treatments including laser surgery, have had to undergo the surgical establishment of a long-term tube-free tracheostomy or laryngotracheal reconstruction. The only complication encountered during or following the endoscopic dilatation was secondary to the jet ventilation mode of providing anesthesia. Pneumothorax was encountered in one patient with known pulmonary involvement of her WG with history of past episodes of pneumothorax. This particular patient underwent three additional dilatation procedures since the pneumothorax without repetition or recurrence of this complication.

■ PERMANENT TUBE-FREE SPEECH-READY TRACHEOSTOMY

This procedure is a radically new approach to the management of chronic airway obstruction. When indicated, it provides safe, secure airflow through a bypass of the glottic or subglottic narrowing with preservation of the patient's voice, cough mechanism, and the swallowing process. Once the tracheostoma has healed, usually three weeks after the surgical procedure, the patient is free of the pain, discomfort, and complications of the indwelling tracheotomy tube which is otherwise a prerequisite for the maintenance of conventional tracheotomy. Within two months postoperatively, 70% of the patients with long-term tube-free tracheostomy develop the ability to constrict their tracheostomal opening for unaided production of normal voice and effective cough. A supplementary sling procedure is available to the minority group that fails to achieve effective constriction and sealing of the stoma for the purpose of producing speech and normal cough.

■ SUMMARY

Further development of conservative endoscopic procedures at the CCF has provided patients with dependable surgical adjuncts to the systemic medications of WG. In most instances, a single surgical dilatation procedure stabilizes the patients at an airway diameter that exceeds 50% of the norm, thereby rendering the patient almost asymptomatic at rest and minimally restricted during exercise. The more chronic patients presenting with history of previous surgical procedures performed on their larynx

and trachea, usually in other institutions, have been observed to require consecutive treatments after the original treatment, enjoying shorter symptom-free intervals. We have not encountered any local complications such as damage to the vocal cords, altered voice, or compromised structural integrity of the larynx and the trachea. The procedure has been found to be effective and well-tolerated as a means for treating, maintaining, and rehabilitating patients with chronic airway obstruction, particularly in those that have been initially treated by our service, and those that were managed from the very beginning of their disease. However, even the more difficult and complicated cases clearly demonstrate an improvement in their condition through the above treatment protocol, and the interval between treatments gradually increases in this group as well.

The more aggressive long-term tube-free tracheostomy procedure, usually performed only on difficult and select patients with severe complications, has proven itself to be a highly gratifying procedure, achieving a permanent mode of management for these patients which safely allows for almost complete freedom from symptoms combined with good tolerance and functional rehabilitation.

Video documentation will serve to further demonstrate the beneficial effects of both these modes of treatment.