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Resection and Anastomosis for Laryngotracheal Stenosis in Wegener's Granulomatosis: Case Report

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ABSTRACT

Management of the airway stenosis due to Wegener's Granulomatosis Disease (WGD) is controversial. A 37-year-old woman with WGD and severe subglottic stenosis was treated successfully by resection and anastomosis of the subglottic area. Twelve months after the operation, she enjoys normal breathing and near normal voice. ***(Tanaffos 2002; 1(4): 73-76)***

Key words*: Wegener's granulomatosis, Subglottic stenosis, Laryngotracheal resection*

INTRODUCTION

Wegener's Granulomatosis Disease (WGD) is an inflammatory disease with unknown etiology in which multiple organs may be involved.

Vasculitis, granuloma formation and necrosis may be found in the microscopic examination of the tissues of the involved areas.

Major airway stenosis in WGD is part of the systemic manifestation, which would usually respond to medical treatments. In some cases, despite remission of systemic signs and symptoms, the stenosis will persist. In such cases, surgical interventions may be indicated. Usually conservative approaches such as bronchoscopic dilatation and use of Neodymium Yttrium- Aluminum- Garnet (Nd-YAG) laser are preferred methods to relieve these persistent stenoses.

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Some authors believe that in WGD, any reconstructive procedure in trachea and subglottic area is impossible due to the probability of recurrence and exacerbation of disease induced by surgical trauma. The possibility for disruption of anastomosis in a patient under immunosuppressive therapy is another prohibitive factor (1,2).

CLINICAL SUMMARY

A 37-year-old woman, known case of WGD, was admitted with severe dyspnea due to tracheal and subglottic stenosis. Her illness began 11 years earlier with severe pain in her wrists. She was treated with various medications without clinical improvement. One year later, fever, malaise, and weakness ensued; moreover, her chest x-ray showed nodular opacities in both lungs. Open lung biopsy was performed. Histopathologic report was nonspecific granulomatous inflammation. Then empirical

antituberculous therapy was prescribed without any clinical improvement.

The patient has been symptomatic periodically during the following six years for which symptomatic therapy was performed for each course. Five years before admission, lesions such as sinusitis, otitis, and defects in nasal septum appeared. ANCA (Anti Neutrophil Cytoplasmic Antibody) became positive, and the patient was treated with cyclophosphamide 100 mg daily and prednisolone 40 mg daily (tapered down to 5 mg daily dose) with diagnosis of WGD. By this regimen, the patient remained asymptomatic for about 3 years. Then progressive dyspnea developed, and bronchoscopy showed severe subglottic stenosis, although the other systemic manifestations were in remission. Biopsy of laryngeal mucosa showed granulomatous lesion in favor of WGD. Initially, bronchoscopic dilatation was carried out and resulted in partial improvement, but symptoms relapsed one month later.

Frequent dilatations were performed but each time returned with more severe stenosis. Finally, we decided to perform surgical resection. She was operated upon about 9 months after initial admission in our service. Surgical findings were: fibrotic stenosis of subglottic area, thickened cricoid cartilage, fibrotic stenosis of tracheal mucosa, and narrowing of tracheal cartilagenous rings.

The surgical procedure was: "resection of the subglottic stenosis and 2cm of proximal trachea with anastomosis of trachea of the thyroid cartilage anteriorly and to the mucosa of subglottic area posteriorly".

Furthermore, a T-tube was also inserted as a stent through a longitudinal incision on the tracheal wall 1cm below the anastomosis. Upper arm of the T-tube was conducted between the vocal cords while the lower arm was in the trachea. On the third post-operative day, severe dyspnea developed due to obstruction of lumen of the T-tube by mucus plug, not removable by suctioning and the patient was taken to the operating room emergently. Operation site was re-opened and the T-tube was drawn out. Anastomosis was intact at that time, and disruption of suture line was seen. A small tracheostomy tube was inserted through the same stoma. Thereafter, the postoperative course was uneventful, and two weeks later, tracheostomy tube was drawn out and the patient was discharged with a normal airway and near normal voice.

After 12 months of operation, the patient became well with normal breathing and the only remaining problem was mild hoarseness.

DISCUSSION

Treatment of major airway stenosis due to WGD is primarily systemic, with cyclophosphamide and prednisolone. However, in some cases, in spite of remission of systemic disease, stenosis persists and local treatment becomes necessary. Biopsy of these stenotic sites usually shows nonspecific findings. Reports of similar cases show that subglottic stenosis may have an independent course from the other manifestation. Conservative non-surgical methods have been used for the treatment, and common belief has been that these stenoses can not be managed by resection and anastomosis (3,4).

A report of successful surgery in 3 patients by Herridge and coworkers (5) was a good stimulus to dispute this belief and our case is another proven one. One important difference exists between Herridge's patients and ours: In the Herridge's patients, biopsy of stenotic sites revealed nonspecific lesions, while in our patient, granulomatous lesions were seen. The post surgical specimen in our patient showed vasculitis in addition to granulomatous lesions. Thus, we think that our patient had a locally active disease in the stenosis site at the time of operation while active disease process had ceased in Herridge's patients.

Although we have no doubt that subglottic and tracheal stenosis in WGD are primarily a local process that must be treated with local procedures initially; however, we insist on surgical resection as an option with effective and appropriate results for some of these patients in whom systemic therapy has failed to relieve local stenosis in major airways.

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