Iatrogenic Membranous Tracheal Tear in the Setting of Relapsing Polychondritis

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Abstract

Importance: Relapsing polychondritis is a rare immune mediated disease disorder characterized by episodic inflammation of cartilaginous tissues with sequelae affecting virtually every major organ system. Recurrent destruction of these cartilaginous tissues leads to eventual weakening, and may predispose the patient to complications requiring airway intervention.

Observations: This case report details the management of an iatrogenic 10 cm membranous tracheal tear in 27-year-old female suffering from relapsing polychondritis.

Conclusions and Relevance: Conservative management coupled with starplasty and cervical approach tracheopexy was successful in complete closure of a complex tracheal tear. To our knowledge this is the first case report of a tracheal perforation in the setting of active relapsing polychondritis.

Keywords: Polychondritis; Iatrogenic; Tracheal Tear

Introduction

Relapsing Polychondritis (RP) is a rare systemic inflammatory disease that primarily affects hyaline cartilage [1]. This Th1-immune mediated disease is thought to have a worldwide prevalence of approximately 3.5 per million and is characterized by episodic inflammation of cartilaginous tissues leading to eventual degradation and scarring of the involved structures [1,2]. The target antigens for this disease continue to remain unknown; however type II collagen is currently under investigation with promising results. These patients often present with recurrent episodes of respiratory distress, biphasic stridor, auricular deformity, saddle-nose deformity, and sensorineural hearing loss. Airway manifestations of this disease process include laryngomalacia, subglottic stenosis (SGS), and tracheobronchomalacia [2]. Recurrent inflammation of the cartilaginous trachea leads to decreased overall support and increased propensity for collapse due to negative intrathoracic pressures [4]. When severe, these sequelae can be managed with repeated dilation, stenting, laryngotracheal reconstruction, and rarely tracheostomy [2-5]. Prospective data regarding medical management is sparse and RP is currently treated with a combination of corticosteroids, methotrexate, azathioprine, and newer biologic agents [6].

Perforation of the membranous tracheal wall is a rare complication of traumatic orotracheal intubation, percutaneous tracheostomy, or rarely, surgical tracheostomy. Common presenting signs include shortness of breath, bleeding, and subcutaneous emphysema [7,8]. In these cases pneumothorax, pneumomediastinum, and tracheoesophageal injury must be ruled out. Due to the infrequency of this complication, universally accepted guidelines for management have not been developed and therapeutic options range from conservative observation to open reconstruction.

Case Report

On arrival, the patient was found to be septic. Significant physical findings included subcutaneous emphysema and a sizable air leak around the 6.5 endotracheal tube. She was immediately co-boarded with cardiothoracic surgery for direct laryngoscopy, bronchoscopy, esophagoscopy, and PEG (Percutaneous Endoscopic Gastrostomy) tube placement. Direct laryngoscopy revealed grade III subglottic stenosis with 99% airway occlusion. Serial rigid laryngeal dilation was utilized to establish an upper airway access point to the trachea and flexible bronchoscopy was performed. A large 10 cm posterior tracheal perforation was noted originating immediately distal to the tracheostomy stoma and extending to within 1 cm of the carina (Figure 1: a,b). The esophagus was noted to be prolapsing through the tracheal perforation and occluding 75% of the airway (Figure 1b) and severe bronchomalacia was also appreciated. There was no sign of a tracheo-

![Figure 1](image_url)

**Figure 1:** Parts a,b: Initial endoscopic evaluation of posterior tracheal perforation. Flexible bronchoscopy on presentation reveals a 10 cm posterior tracheal tear. The tracheal mucosa is inflamed and friable. a) The false lumen is evident (arrow) with positive pressure. b) Esophageal prolapse is seen anteriorly (arrow) with spontaneous respiration.
esophageal fistula (TEF) on esophagoscopy. Due to instability of the airway the ETT was replaced with a cuffed Shiley® distal long XLT. This was done in several steps to prevent cannulation of the false passage to the mediastinum. The patient was intubated from above using the flexible bronchoscope. The bronchoscope was then withdrawn and inserted through the tracheostomy stoma. Once this was in satisfactory position distal to the perforation, the tracheostomy tube was advanced over the bronchoscope and placement was confirmed. A salivary diversion tube was placed in order to prevent persistent aspiration of oral secretions.

Throughout this hospital stay the Otolaryngology, Cardiothoracic, and Rheumatology services were in frequent communication regarding therapeutic options as well as prognosis. Once stable from an infectious and cardiopulmonary perspective, consideration was given to primary repair via trans-thoracic approach with a muscle flap, trans-tracheal repair, endoscopic repair, as well as stenting with a Montgomery® T-tube [9]. Ultimately it was felt that the chronic inflammation associated with active relapsing polychondritis would make surgical repair and approach to the perforation very difficult, and could lead to diffuse tracheal stenosis. Conservative therapy versus surgical repair was discussed with the patient and her family.

Once the patient further improved from a cardiopulmonary perspective, a total of three weeks following the injury, the tracheostomy tube was reconsidered as it proved to be uncomfortable for the patient and exerted pressure on the anterior esophageal wall. In order to provide a more permanent option, the tracheostomy stoma was revised, and a starplasty was performed [10]. A DL with rigid laryngeal dilation and flexible bronchoscopy was again performed to inspect the trachea. Findings at this time included a substantial improvement in the membranous tracheal tear, with only a 1 cm residual perforation identified (Figure 2). The healthy anterior esophageal wall appeared to have served as scaffolding for re-epithelialization of the posterior tracheal wall. Additional findings included severe tracheobronchomalacia without any sign of tracheoesophageal fistula or distal tracheal stenosis. The cardiothoracic team used our 4 cm horizontal incision to perform a tracheoectomy to address the patient's severe tracheomalacia. The dissection was carried down to the level of the anterior clavicular periosteum in a pretracheal plane. The trachea was mobilized superiorly and secured to the clavicular perichondrium using 5-0 polypropylene. Superiorly, similar sutures were secured from the trachea to the sternocleidomastoid muscle. Following this intervention, bronchoscopy revealed significant improvement of tracheal collapse. Following completion of the starplasty, a soft silastic sternal stent was placed to maintain patency, decrease pressure on the anterior esophageal wall and reduce the possibility of erosion.

Over the following two weeks routine surveillance was performed via tracheoscopy. The tear subsequently closed without significant narrowing of the tracheal lumen (Figure 3). Following closure of the perforation, we were able to visualize the salivary diversion tube through a sub-centimeter tracheoesophageal fistula. This was likely due to the silastic sternal stent and salivary diversion tubes lying in close proximity and exerting pressure on the anterior esophageal wall. The diversion tube was downsized and tracheal stent was trimmed at an oblique angle to avoid any contact with the posterior tracheal wall. This tracheoesophageal fistula resolved over the course of two weeks. The patient was discharged and has had no additional respiratory events at 1, 2, 3, 6-month follow-ups.

**Discussion**

To our knowledge, this is the first case of an iatrogenic membranous tracheal tear occurring in the setting of relapsing polychondritis and the largest tracheal tear managed with conservative therapy reported.

The incidence of tracheal injury has been described as 1/20,000 with single lumen intubations and 0.5-1% in double lumen ETT [7,9]. Previously described case series recommend that perforations greater than 2-4cm, as well as those with serious respiratory compromise or progressive subcutaneous emphysema should be managed with surgical repair [7-9]. It should be noted, however, that mortality in patients undergoing open repair following tracheal tears sustained during emergency intubation is substantially higher than those managed conservatively [8]. Guidelines initially introduced by Ross et al. [11] have been advocated to guide selection of patients for non-operative management of tracheal lacerations. These guidelines include: 1) Vital sign stability, 2) No difficulty ventilating the patient while intubated or respiratory distress while extubated, 3) No evidence of esophageal injury, 4) Minimal mediastinal fluid collection, 5) Non-progressive

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**Figure 2:** Endoscopic surveillance 2 weeks following membranous tracheal injury. Flexible bronchoscopy 2 weeks following the injury reveals interval improvement in the size of the tracheal perforation. The anterior esophageal wall is still visible through posterior tracheal perforation (arrow) without significant prolapse.

**Figure 3:** Office based endoscopic surveillance following discharge. Flexible tracheoscopy performed in the office six weeks after the injury reveals interval resolution of the tracheal perforation. The posterior wall of the mid-trachea demonstrates minimal granulation tissue (arrow) without significant obstruction. There is no evidence of tracheo-esophageal fistula.
and an interdisciplinary approach allowed us to avert a potentially lethal disease. Careful airway management, conservative treatment, and an interdisciplinary approach allowed us to avert a potentially catastrophic outcome.

**Conclusion**

While iatrogenic membranous tracheal perforations remain a rarity, the literature concerning the management of these complications is growing. Attempts have been made at developing uniform guidelines for surgical versus conservative based on extent of the tear and associated symptoms. It is important however to use appropriate clinical judgment, as well as to enlist the expertise of our colleagues in other related fields when managing a complex perforation in the setting of an underlying collagen disease. Careful airway management, conservative treatment, and an interdisciplinary approach allowed us to avert a potentially catastrophic outcome.

**References**


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