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A case of tracheobronchomalacia due to relapsing polychondritis treated with Montgomery T-tube

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Abstract

Relapsing polychondritis is a rare and multi-system autoimmune disease of unknown etiology characterized by inflammation and destruction of cartilaginous structures. Its clinical manifestations include recurrent chondritis of the ears, nose, pinna, peripheral joints, and laryngotracheobronchial tree and can be life-threatening in advanced cases of laryngotracheal stenosis. Because of the rarity of relapsing polychondritis and lack of understanding of its pathogenesis, there is no standard medical therapy, and treatment is tailored according to disease activity and site of organ involvement. In respiratory failure due to laryngotracheal involvement, which has been reported in up to 50% of relapsing polychondritis patients and is a major cause of death, immediate procedures such as stenting and tracheostomy are very important. This report describes a 70-year-old male patient suffering from tracheobronchomalacia due to relapsing polychondritis who was treated with Montgomery T-tube insertion.

Keywords

Relapsing polychondritis, tracheobronchomalacia, tracheostomy

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Introduction

Relapsing polychondritis (RP) is a rare multi-system autoimmune disease characterized by recurrent, destructive, inflammatory lesions of the auricular, nasal, and laryngotracheobronchial cartilages.[1] The annual incidence is estimated to be 3.5 cases per million, and the peak age of disease onset is in the fifth decade of life.[2–4] The etiology of RP is not clearly defined, but approximately 30% of RP cases are associated with autoimmune disease.[1,5,6] The diagnosis of RP is usually based on characteristic clinical manifestations and several clinical diagnostic criteria.[3,5,7] However, initial symptoms are often diverse, and the diagnosis is often missed or delayed in the early stages of the disease.[8] Cases in which the first manifestations of RP are dyspnea and acute respiratory distress secondary to tracheobronchial involvement are especially difficult, and diagnosis and adequate treatment (including immediate airway protection) are essential in the emergency department.[9] In this report, we describe the early diagnosis and successful airway protection by Montgomery T-tube application in a patient with acute respiratory failure.

Case report

A 70-year-old man complaining of worsened dyspnea was evaluated in the emergency department. His past medical history included diabetes. In addition, several years prior to presentation, he had visited dermatology clinics for intermittent erythema and swelling in the right auricle and deformity of the nose, but no specific disease was diagnosed. The patient complained of intermittent breathing difficulties for the past year, which was getting worse. His vital signs upon arrival to the emergency department were as follows: body temperature 36.9°C, heart rate 125 beats/min, blood pressure 165/90 mmHg, and respiratory rate 35 breaths/min. Physical examination revealed loud inspiratory stridor on auscultation and inharmonic movement of the thorax and abdomen on respiration, similar to

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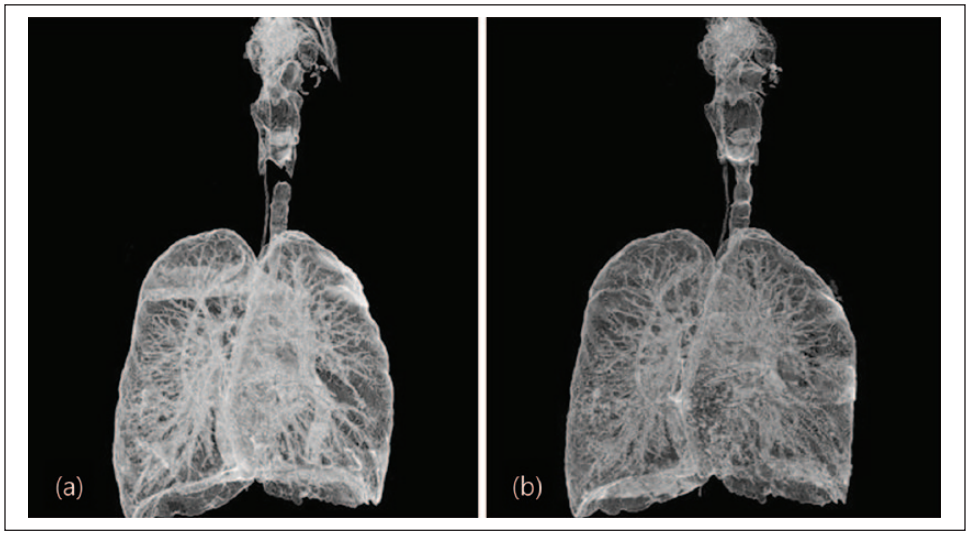

Figure 1. CT scan with 3D reconstruction of the trachea and bronchus. Luminal narrowing between expiration (a) and inspiration (b) at the level of the aortic arch.

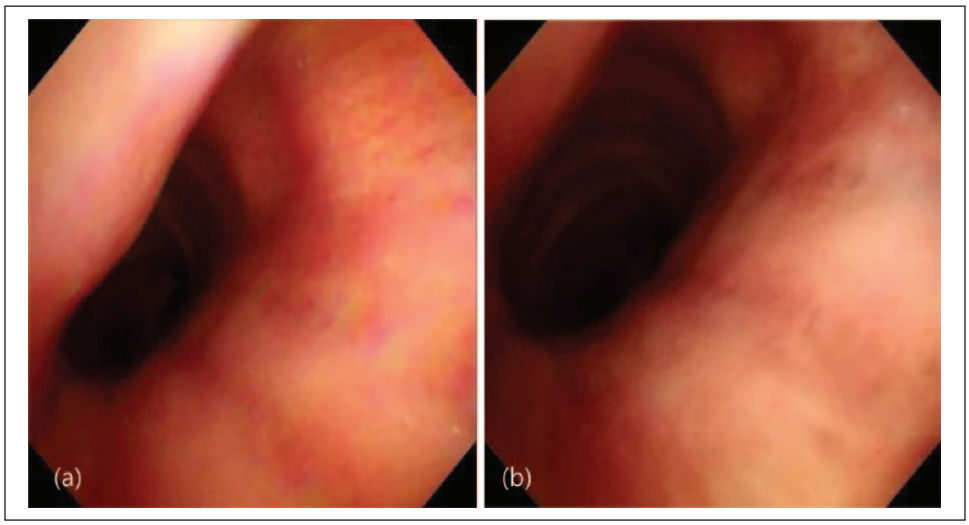

Figure 2. Flexible bronchoscopy showed dynamic luminal narrowing between expiration (a) and inspiration (b) at the upper trachea.

paradoxical respiration. There was no abnormality in the laboratory findings, and plain chest radiography was normal. Because of acute respiratory failure in which oxygen saturation was not maintained, the patient was intubated and moved to the Intensive Care Unit (ICU). Flexible bronchoscopy was immediately performed, and it showed dynamic luminal narrowing of the middle to distal trachea, especially on expiration. On ICU day 3, the patient was stabilized and extubated. A computed tomography (CT) scan with 3D reconstruction of the trachea and bronchus and repeated flexible bronchoscopy were obtained after extubation to confirm the upper trachea lesion. The CT scan showed luminal narrowing between inspiration and expiration at the level of the aortic arch without stricture (Figure 1). The repeated flexible bronchoscopy revealed significant narrowing of the upper trachea that worsened during expiration without abnormality of vocal cord movement or larynx. In addition, we could not find the cartilage ring in the upper trachea (Figure 2). A few hours following extubation, the patient experienced severe respiratory distress with stridor, and endotracheal intubation was performed again. On physical examination, the auricular cartilage was floppy and misshapen, and a saddle nose deformity was observed (Figure 3). Based on the presence of bilateral auricular chondritis, saddle nose deformity, and respiratory tract chondritis, a diagnosis of RP was made by clinical diagnostic criteria. The patient was treated with an intravenous systemic steroid, methylprednisolone 1 mg/kg, for 1 week. In order to evaluate the patient's response to the systemic steroid, a follow-up flexible bronchoscopy was performed and showed sustained bronchomalacia, especially in the upper trachea around the aortic arch. Finally, we decided to perform a

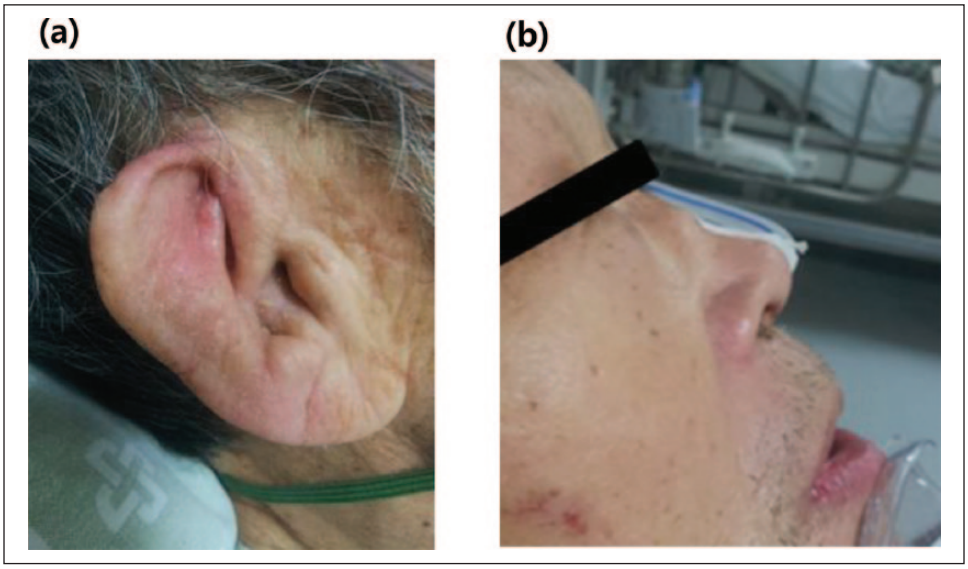

Figure 3. (a) The auricular cartilage was floppy and misshapen, and the pinna was swollen and reddish. (b) Saddle-nose deformity was observed at the nasal septum.

tracheostomy because obliteration of the cartilaginous framework in the trachea is irreversible. A Montgomery T-tube was utilized to protect a longer length of airway and enable the patient to speak. After Montgomery T-tube training for several days in the general ward, the patient was able to breath and speak without inconvenience. To this date, the patient is living well and is being followed in the outpatient clinic with low dose methylprednisolone and methotrexate.

Discussion

We described a rare case of RP that presented as rapidly worsening dyspnea as the first significant manifestation of the disease. As mentioned above, RP is a systemic autoimmune disease characterized by inflammation and destruction of various cartilaginous structures, and the spectrum of clinical presentations is very diverse. Therefore, the diagnosis of RP is difficult, and the rate of misdiagnosis has been reported up to 73%.[10] There are no standardized therapeutic guidelines, but non-steroidal anti-inflammatory drugs, steroids, and immunosuppressant agents have been used according to disease activity and organ involvement. However, patients with respiratory involvement require immediate non-pharmacologic management with airway protection because of the high mortality rate. There are various therapeutic options, such as stenting, airway dilatation, tracheostomy, and laryngotracheal reconstruction. Careful and individualized selection of management with consideration of the patient's condition is very important. Insertion of a Montgomery T-tube is non-invasive and not associated with severe complications. In addition, patients can live well while speaking without great inconvenience for a relatively long period of time. In conclusion, RP is a rare disease that is difficult to diagnose, and treatment in the emergency department is very important. We successfully treated a patient who presented with acute respiratory failure as the first significant manifestation of RP by application of a Montgomery T-tube.

Conclusion

Clinical manifestations of RP include recurrent chondritis of the laryngotracheobronchial tree and can be life-threatening in advanced cases of laryngotracheal stenosis. In respiratory failure due to laryngotracheal involvement, immediate procedures such as stenting or tracheostomy are very important. We describe successful airway protection by Montgomery T-tube application in a patient with acute respiratory failure.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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