Case report

Multiple endobronchial airway stents in a case of relapsing polychondritis - A rare entity

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ABSTRACT

Relapsing polychondritis (RP) is a rare multisystem condition. Nearly 50% of patients are suffering from airway involvement in RP and it can be fatal. Besides immunotherapies, endobronchial stenting has been found to be useful in the treatment. Insertion of endobronchial stents in patients with RP has its own complications and has been associated with increasing morbidity and mortality.

We describe placement of multiple endobronchial stents to prevent airway closure in a 76-year-old man with RP due to recurrent dyspnea. Insertion of multiple stents (6th stent) in the left main bronchus was necessary due to severe narrowing of the left main bronchus.

Recurrence of airway involvement in RP is common. Early diagnosis and prompt treatment are essential to reduce the risk of life-threatening airway collapse. The insertion of multiple stents in this patient has resulted in improving symptoms, spirometry, and a return to daily activities.

Abbreviations

RP Relapsing Polychondritis
CT Computed Tomography
Nd YAG Neodymium Yag Laser
APC Argon plasma Coagulation
USA United States of America
SEMS Self Expandable Metallic Stents
FEV\textsubscript{1} Forced Expiratory volume in 1 second
FVC Forced vital Capacity

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1. Introduction

RP was first described as early as 1923 by Jaksch-Wartenhorst. It is a relapsing–remitting multisystem disease of unknown aetiology with episodes of recurrent inflammation of cartilaginous structures.

Literature search showed around 600 cases of RP that were reported worldwide. It often goes misdiagnosed due to its diagnostic arduousness. We report a case of RP who presented atypically with a great therapeutic challenge.

2. Case report

A 76-year-old-man presented with gradual worsening of dyspnoea and chest tightness 4 months ago. He was referred from the Hospital where he used to attend follow up 10 years ago. The condition was diagnosed as scleritis when he presented with red eyes. A year later, his voice turned hoarse, saddle nose deformity was seen alongside with sensory neural hearing loss. Nasal septal cartilage biopsy revealed RP. He was commenced on prednisolone.

In the same year, he started complaining of SOB which surfaced due to stenosis of the left main bronchus as shown on CT Thorax (Fig. 1). Thus, he was stented for the 1st time in 2012 with uncovered Ultraflex stent measuring 12mm × 20 mm. In the second year of presentation, he was stented for the 2nd time by uncovered Ultraflex stent measuring 12mm × 20 mm due to granulation tissue formation.

He developed granulation tissue in the proximal part of the second stent. Due to this, a 3rd uncovered Ultraflex stent measuring 12mm × 40 mm was placed. He suffered from a recurrent stenosis due to formation of granulation tissue and was treated with balloon dilatation a year later. However, it rapidly recurred in the same year and was treated by YAG LASER,APC, balloon dilatation and the 4th stent covered ultraflex measuring 14mm × 40 mm was placed 2 years later. A year later, a 5th stent covered ultraflex measuring 14mm × 30 mm was placed.

On his latest admission, he was clinically distressed. Oxygen saturation was 96% on room air. Respiratory examination revealed expiratory rhonchi. High-resolution CT of the chest currently revealed narrowing in the left main bronchus (Fig. 2 a).

He was planned for the insertion of the 6th stent. After an informed consent was obtained, and with the patient under general anaesthesia, a rigid bronchoscopy (size 13.2 mm in outer diameter) was passed through the trachea. A covered ultraflex with thick sputum on it was seen on the left main bronchus. The left upper lobe orifice was marginally seen from the bare metallic distal end (Fig. 3) and lower lobe bronchi were easily collapsible. A cryo probe (2.4 mm) was used for the removal of granulation tissue at the orifice of the left main bronchus. Subsequently, CRE balloon was used to dilate the bronchus and via fluoroscopic guidance, a fully covered metallic stent (AERO, Merit Medical Systems, South Jordan, UT, USA, 10 mm diameter, 30 mm in length) was deployed into the left main bronchus (Fig. 4). A repeat spirometry on the following day demonstrates FVC improved mildly from 2360ml to 2370ml, FEV1 from 970ml to 990ml and patients’ symptoms improved dramatically. Post procedure CT scan showed patent left main bronchus (Fig. 2 b).

3. Discussion

Literature on RP is rare. The incidence is 3.5 per 1 000 000,[1] and it commonly occurs in the 40–60 years age group. [2]. RP affects both genders equally. However, severe airway involvements are more common in women.2RP is an immune-mediated disease targeting mainly type II collagen in cartilaginous structures.

There are few available criterias to diagnose RP [3]. The McAdam’s criteria were the initial diagnostic criteria of RP and required three out of six criterias of the following: bilateral auricular chondritis, nonerosive seronegative inflammatory arthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, and audiovestibular damage [4]. Damiani and Levine proposed modified criteria which include meeting one McAdam’s criteria plus histopathological confirmation or two McAdam’s criteria plus response to corticosteroids or dapsone [5]. In recent times, the diagnosis of RP relies mostly on the criteria established by Michet et al. which

![Fig. 1. (a) & (b) CT Thorax of the patient before stenting in December 2011.](image-url)
require the presence of a proven inflammation in at least two of three of the auricular, nasal, or laryngotracheal cartilages or the proven inflammation in one of these cartilages plus two other signs, including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis, or hearing loss [6]. (Table 1). In our case the patient has ocular, nasal and respiratory tract chondritis with sensory neural hearing loss. He has done biopsy of his nasal cartilage, however our team unable to trace the histopathological slides from the referred center as it was done almost 10 years ago.

Almost all patients presented with auricular chondritis [2]. Airway involvement in RP manifests due to the abnormal softening of the trachea and bronchi as a result of chronic chondritis. Eventually, half of the patients experience airway involvement in RP and it can be fatal [7]. Airway involvement is characterized by dyspnoea, cough, wheeze, stridor, hoarseness and a phonia. Tracheomalacia, bronchomalacia or tracheobronchomalacia is due to recurrent airway chondritis and often associated with poor prognosis [1,8].

There is no standard treatment for RP. No clinical trials have been carried out due to its rarity. Therefore, current medical therapy for RP is largely empirical and are based on case reports. However, RP generally responds to corticosteroids and immunosuppressive drugs.

Late diagnosis of airway involvement in RP leads to irreversible damage. Corticosteroids or other immunosuppressive drugs are often ineffective in these severe cases [9]. Thus, treatment strategy has shifted from medical therapy to positive airway pressure by mechanical ventilation and stent placement. Stent placement has significantly improved the patient outcome. Other airway interventions which can be used in RP are balloon dilation, tracheostomy or tracheal and laryngotracheal reconstructions. Although there are several reports of the usage of expandable metallic stents for relapsing polychondritis, we believe that there are no published reports on the usage of multiple SEM stents in a patient.

Tracheobronchial stents in patients with a major airway stenosis or collapse is a life-saving option. Stents maintain airway patency
and provide immediate symptomatic relief [8]. Many case studies reported immediate improvement of dyspnoea following stent insertion, with patients even returning to their normal active lifestyle [7]. [8, 9].

Complications of airway stenting are aspiration pneumonia, tracheobronchial wall erosion, granulation tissue formation, and recurrent infection. Therefore, stents are only inserted in patients who are unresponsive to medical therapy. This case demonstrates that the placement of multiple metallic bronchial stents can successfully alleviate recurrent airflow limitation caused by RP.

In conclusion, airway involvement in RP can be fatal. Interventional pulmonology techniques can successfully alleviate the symptoms of airway involvement due to RP. More studies need to be conducted in the future to consolidate treatment guidelines of RP.

Author contribution statement

Arvindran Alaga is a Corresponding Author. Arvindran Alaga and Hatem Mohammed are responsible for draft the manuscript. Akane Ishida is responsible for collection of patient’s data and history. She also involved in patients’ management. Masahide Oki is involved in patients’ management and supervise the draft of manuscript. Hideo Saka is involved in patients’ management and

Fig. 3. Covered Ultraflex with cream colored sputum on the left main bronchus.

Fig. 4. Proximal end of Aero stent was just at the tracheal bifurcation by rigid forceps retraction.
supervise the draft of manuscript. All authors read and approved the final manuscript.

4. Ethics statement

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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Declaration of competing interest

☒ The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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