Case Report

Relapsing Polychondritis with Laryngotracheal Involvement: A Case Report

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Abstract

Relapsing polychondritis is a progressive multisystemic disease affecting primarily cartilaginous tissue of otorhinolaryngologic structures. Because of its rarity and progressive involvement of multisystem, diagnosing and managing a relapsing polychondritis is a clinical challenge, especially when it involves laryngotracheal cartilages, which can be critical and potentially life threatening. We reported a case of a 56-year-old man who presented with progressive shortness of breath and noisy breathing due to thickening of laryngotracheal cartilages. A diagnosis of relapsing polychondritis was made after considering his clinical, histological and radiological findings. Eventhough relapsing polychondritis with laryngotracheal cartilages involvement was rare and the nature of this disease was progressive, it was still advisable to consider a diagnosis of relapsing polychondritis whenever encountered cases of airway obstruction with evidence of thickened trachea, despite the other systemic manifestations did not reveal yet. Generally, systemic corticosteroid and other immunosuppressive agents are the treatments of choice in most cases however, when laryngotracheal cartilages are involved, emergency tracheostomy will be indicated.

Keywords: Airway obstruction; laryngotracheal; relapsing polychondritis; stridor; tracheal stenosis; tracheostomy

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Introduction

Relapsing polychondritis is an uncommon recurring and progressive multisystemic inflammatory disease affecting primarily cartilaginous tissue of auricle, nasal septum, articular, larynx and tracheobronchial system, as well as proteoglycan-rich tissue in eyes, blood vessels and heart (1-2). Although the exact pathogenesis of relapsing polychondritis is not well known, the disease is suspected to be immunologic-related. As reported by Foidart et al. (3), 5 out of 15 patients were found to have antibodies to Type II collagen at the onset and acute phase of the disease. Because of its rarity and progressive involvement of multisystem, diagnosing and managing a relapsing polychondritis is a clinical challenge, especially when it involves laryngotracheal cartilages, which can be critical and potentially life threatening.

Here we presented a case of relapsing polychondritis, which clarified the challenges in establishing the diagnosis and its treatments.

Case Report

A 56-year-old man presented to our department with progressive worsening of shortness of breath and noisy breathing for 1-month duration. He had no history of fever. On admission, he was in respiratory distress with stridor while flexible laryngoscopy revealed mobile bilateral vocal cords with narrowing of subglottic region.

A computed tomography (CT) scan of the neck (Fig. 1a) showed diffused circumferential thickening of the

larynx maximally at the subglottic region and tracheal wall down until the bronchus. Thus, emergency tracheostomy was performed for airway protection proceeded with direct laryngoscopy, which revealed collapsed of tracheal wall and abnormal, oedematous looking tracheal mucosa with absence of tracheal rings and copious secretion (Fig. 1b). Biopsy of the trachea cartilage was taken twice which was first reported as regenerative appearance with scarring and patchy mild inflammation, whereas the second biopsy showed evidence of cartilage resorption and inflammation with infiltration of fibrous tissue. There were no granulomas, vasculitic changes, evidence of malignancy or amyloid deposits noted.

Apart from that, on examination we noticed that the patient also had saddle nose deformity (Fig. 2), but otherwise, normal in both external auricles. Blood investigations showed leucocytosis with significant raised erythrocyte sedimentation rate. All related autoimmune screening including rheumatoid factor, antinuclear antibody and anti-neutrophil cytoplasmic antibody were negative. The tuberculosis workout was also done with positive results (nil).

Considering these clinical, radiological and histological findings, a diagnosis of relapsing polychondritis was finally made. The patient was then

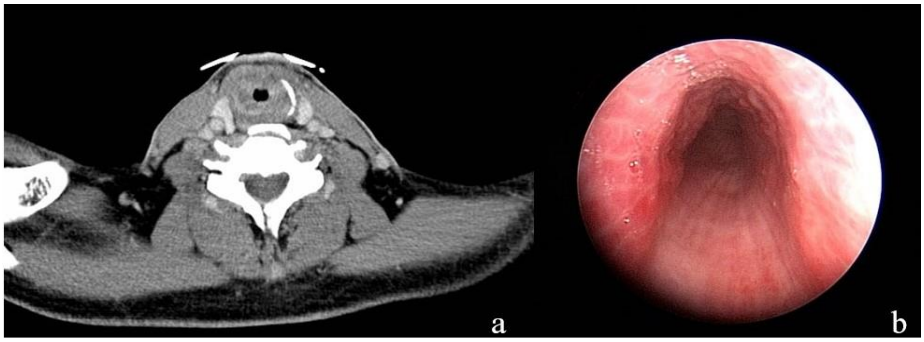

FIGURE 1a (Left): Computed tomography scan of neck showed diffused trachea thickening with luminal narrowing; 1b (Right): Endoscopic examination of trachea demonstrated oedematous mucosa with absence of tracheal rings

underwent a long T-tube tracheal stent insertion and was prescribed with a course of prednisolone by a rheumatologist. However, he developed recurrent severe pneumonia with respiratory failure necessitating change of T-tube to tracheostomy tube for ventilation and treatment with antibiotics. Presently, his condition had improved, despite that he still required long-term prednisolone.

Discussion

Diagnosing a relapsing polychondritis is a clinical challenge in view of multisystemic involvement, which is progressive in nature. There are few schemes of criteria helps in establishing the diagnosis of relapsing polychondritis (Table 1). The first classic diagnostic criteria of Mc Adam et al. in 1976 required at least 3 out of 6 criteria of the following to be fulfilled together with histologic confirmation: recurrent chondritis of both auricles, nonerosive inflammatory polyarthritis, chondritis of nasal cartilages, inflammation of ocular structures, chondritis of the respiratory tract involving laryngeal and/or tracheal cartilages and cochlear and/or vestibular damage (4). These criteria had been modified by Damiani and Levine in 1979 to the

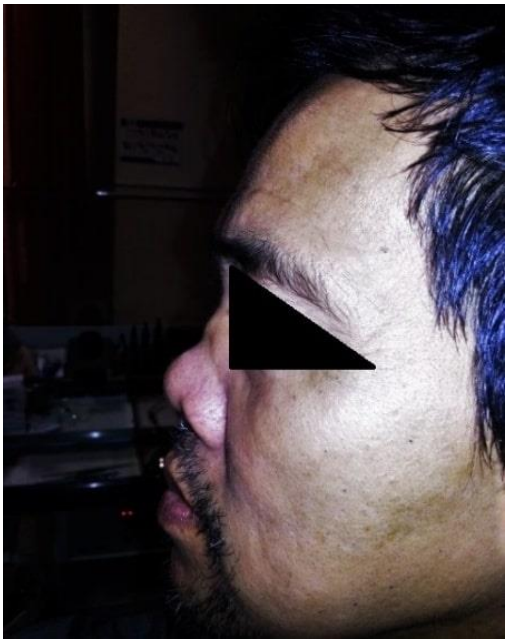

FIGURE 2: Photography of the external nose showed saddle nose deformity

following criteria, which require meeting any of these: fulfilled 3 out of 6 Mc Adam et al.'s criteria, or at least 1 of the Mc Adam et al.'s criteria with a positive histologic confirmation, or 2 of the Mc Adam et al.'s criteria and response to corticosteroid or dapsone (5). Michet et al. had established the latest modified criteria in 1986, which required presence of any of the following: proven inflammation in 2 out of 3 cartilages (auricular, nasal, and Laryngotracheal) or proven inflammation in 1 of these cartilages as well as meeting 2 other signs from ocular inflammation, hearing loss, vestibular dysfunction, or seronegative inflammatory arthritis (6).

Out of those clinical presentations, Sharma et al. (1) reported that auricular chondritis is found to be the most commonly involved in up to 96% of cases, followed by 81% of nasal involvement in which 14% of them complicated with saddle nose deformity and only 11.5% with laryngeal/tracheal involvement. However, other former literature reported by Mc Adam et al. (4) had revealed that nearly 50% out of 23 patients had laryngeal/tracheal chondritis, in which 36% of them required tracheostomy.

On top of that, histological confirmation also plays a part in diagnosing relapsing polychondritis. According to Mc Adam et al., changes in the histological examination of a biopsied cartilage that can be characteristics of relapsing polychondritis include loss of basophilic staining of the cartilage matrix, perichondral inflammation, and cartilage destruction with replacement by fibrous tissue (Table 1) (4).

Table 1: Methods of Diagnosing Relapsing Polychondritis

Diagnostic method	**Criteria**
Mc Adam et al. (1976)	A diagnosis is considered certain when 3 or more of these features are present, confirmed by biopsy. The degree of diagnostic assurance increases with each additional feature: 1. recurrent chondritis on both auricles 2. non-erosive inflammatory polyarthritis 3. chondritis of nasal cartilage 4. inflammation of ocular structures, including conjunctivitis, keratitis, scleritis/episcleritis and/or uveitis 5. chondritis of the respiratory tract involving laryngeal and/or tracheal cartilage 6. cochlear and/or vestibular damage manifest by neurosensory hearing loss, tinnitus, and/or vertigo.
Damiani & Levine (1979)	McAdam et al.'s diagnostic criteria were modified as follows by Damiani and Levine (1979): 1. Fulfilled at least 3 or more of McAdam et al.'s diagnostic criteria, histologic examination is not essential 2. Fulfilled 1 or more of McAdam et al.'s criteria with positive histologic confirmation or 3. Chondritis in 2 or more separate anatomic locations with response to steroids and/or Dapsone
Michet et al. (1986)	Require the presence of any of the following: 1. Proven inflammation in 2 out of 3 cartilages (auricular, nasal, and laryngotracheal) 2. Proven inflammation in 1 of these cartilages as well as meeting 2 other signs from ocular inflammation (conjunctivitis, keratitis, episcleritis, uveitis), hearing loss, vestibular dysfunction, or seronegative inflammatory arthritis.
Histological confirmation by Mc Adam et al. (1976)	Changes are seen in the histological examination of a biopsied cartilage (obtained from the ear, nose or respiratory tract) with light microscopy. The histologic examination of cartilage from a clinically involved site will confirm the underlying chondritis: 1. Loss of basophilic staining of the cartilage matrix due to the loss of matrix acid mucopolysaccharides. This could also lead to focal or diffuse eosinophilic changes 2. Perichondral inflammation 3. Eventual cartilage destruction with replacement by fibrous tissue. The border between cartilage and connective tissue becomes indistinct with increased fibrocytic and endothelial cell activity. The chondrocytes change, becoming vacuolated and pyknotic, while macrophages may phagocytose them, causing dispersion into the surrounding connective tissue and eventual disappearance.
Computed tomography (CT) scan by Faix & Branstetter (2005).	Typical CT findings of relapsing polychondritis includes: 1. subglottic stenosis 2. tracheobronchial luminal narrowing (not specific to relapsing polychondritis, should consider other causes such as tracheopathia osteochondroplastica, trauma, infection, Wegener's granulomatosis, sarcoidosis, and amyloidosis) 3. densely calcified and thickened tracheal cartilage 4. peripheral bronchial narrowing and bronchiectasis 5. calcifications of the pinnae, and nasal cartilage collapse - increased attenuation and smooth thickening of the airway walls (most frequent CT manifestations of relapsing polychondritis)

Computed tomography (CT) scan nowadays has become the most useful and widely used imaging tools as addition to diagnostic instruments of relapsing polychondritis (Table 1) (7). From the CT thorax in particular, both trachea and bronchial wall can be affected as evidence by smooth diffuse thickening of the wall with or without calcification and eventually narrowing of the airway (8).

In this patient, the diagnosis of relapsing polychondritis was made based on the Damiani and Levine criteria, where 2 of the Mc Adam et al.'s criteria had been met including; the chondritis of the respiratory tract which was evidenced by diffused circumferential thickening of the laryngotracheobronchial system and the chondritis of nasal cartilage as showed by saddle nose deformity, plus evidence of inflammation, resorption and regeneration of the tracheal cartilage with fibrous tissue in the histopathological findings. Apart from that, these proven inflammations involving the 2 cartilages: laryngotracheal and nasal also supported the criteria of Michet et al. (6).

Systemic corticosteroid is accepted as the mainstay treatment of choice for relapsing polychondritis (9,10). Other immunosuppressive agents such as methotrexate and cyclophosphomide are also been used in the treatment to decrease the steroid effects (10). In a case of relapsing polychondritis with airway obstruction secondary to laryngotracheal involvement, tracheostomy was indicated. Its prognosis was also reduced compared to those cases involving nasal and auricular alone (10).

Conclusion

Relapsing polychondritis is a rare inflammatory disease affecting primarily cartilaginous tissue of otorhinolaryngologic structures frequently. This paper described a case of relapsing polychondritis with rare involvement of laryngotracheal cartilages which potentially life threatening. In view of its multisystemic involvement, which is progressive in nature, diagnosing a relapsing polychondritis is a clinical challenge. Thus, thorough clinical examinations with evidence of histological confirmation help in diagnosis and subsequently lead to appropriate treatments.

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