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Case Report

# Subglottic Stenosis as an Isolated Clinical Manifestation in Relapsing Polychondritis – A Case Report

**José Antonio Pinto[1,2], Heloisa dos Santos Sobreira Nunes[2], Gabriella Spinola Jahic[2], Andréia Natalia Azevedo Ferreira de Vasconcelos[2*], Marina Cançado Passarelli Scott[2], Caio Alonso Orlandini[2], Cassia Paloma da Cunha Onofre[2]**

[1]Chief of Department of Otolaryngology from Núcleo de Otorrinolaringologia, Cirurgia de Cabeça e Pescoço e Medicina do Sono de São Paulo.

[2]Medical Doctor of Department of Otolaryngology, Núcleo de Otorrinolaringologia e Medicina do Sono de Sao Paulo, Brazil.

***Corresponding Author:** Ferreira de Vasconcelos, Medical Doctor of Department of Otolaryngology, Núcleo de Otorrinolaringologia e Medicina do Sono de Sao Paulo, Brazil.

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## Abstract

Relapsing Polychondritis is a rare, multisystem autoimmune disease still of unknown origin, characterized by recurrent inflammation of the hyaline cartilages.

The involvement of Laryngotracheal cartilages in polychondritis is one of the most serious known complications, observed in 50% of patients with worse prognosis. Among the causes of death, laryngotracheal stenosis associated with pneumonia or severe respiratory failure stands out, and may be found in 10% to 50% of patients with the disorder.

We describe a case of relapsing polychondritis in a female patient, with dyspnea as an initial manifestation, in which Grade III Cotton-Myer Laryngotracheal Stenosis was later confirmed. During the 10 years of investigation, clinical treatment stabilized the disease, but the respiratory distress persisted, which prompted the patient to be submitted to surgical correction of laryngotracheal stenosis.

**Keywords:** Relapsing polychondritis, laryngotracheal stenosis, larynx, trachea, dyspnea

## Introduction

Relapsing Polychondritis (RP) is a rare connective tissue inflammatory disease with autoimmune characteristics [1], [2] characterized by recurrent inflammation of the hyaline cartilages. The term "polychondropathy" was first employed in 1923 by Jaksch-Wartenhorst. The incidence of RP is approximately three in a million, affecting mainly patients between 20 and 60 years old, with a peak incidence at 40 years, with only 5% of cases seen in childhood. Other concomitant autoimmune, rheumatological and hematological diseases are observed in 30% of patients [3]. It is believed that specific antibodies and immune responses against type 2 collagen, that develop in up to two thirds of patients with the disorder, are responsible for the pathophysiology of RP [1],[4]. In addition to the uncertainty of the etiology, the duration and severity of the disease and the developed symptoms can vary significantly. Laryngotracheal and bronchial involvement is observed in only 10% of cases[5]. Laryngotracheal stenosis may develop in some of these cases, worsening the prognosis, fatal in the most severe cases [1],[4].

## Case Report

The study was approved by the National Research Ethics Committee, CAAE: 38664820.0.0000.8114 – Comissão Nacional de Ética em Pesquisa (CONEP, in the Portuguese acronym) and was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki.

Female patient, 59 years old, presented intermittent dyspnea which worsened progressively for 10 years. Patient showed improvement after initial therapy with oral corticosteroids, which at first, led to a diagnosis of Bronchial Asthma. Bronchodilators and inhaled corticosteroids were included in her treatment, without improvement. A spirometry test was performed, with results within normal standard values, ruling out the initial diagnosis. A bronchoscopy revealed laryngotracheal stenosis. A biopsy of glottic region was carried out, which demonstrated edema and a nonspecific lymph- plasmocytic infiltrate of the chorion, with no signs of malignancy.

The computed tomography (CT) of the neck (Figure 1), presented minimal calcification amidst a circumferential thickening of the cartilage which lines the infraglottic region of the trachea – minimal transverse area of air column measuring 32mm$^2$.

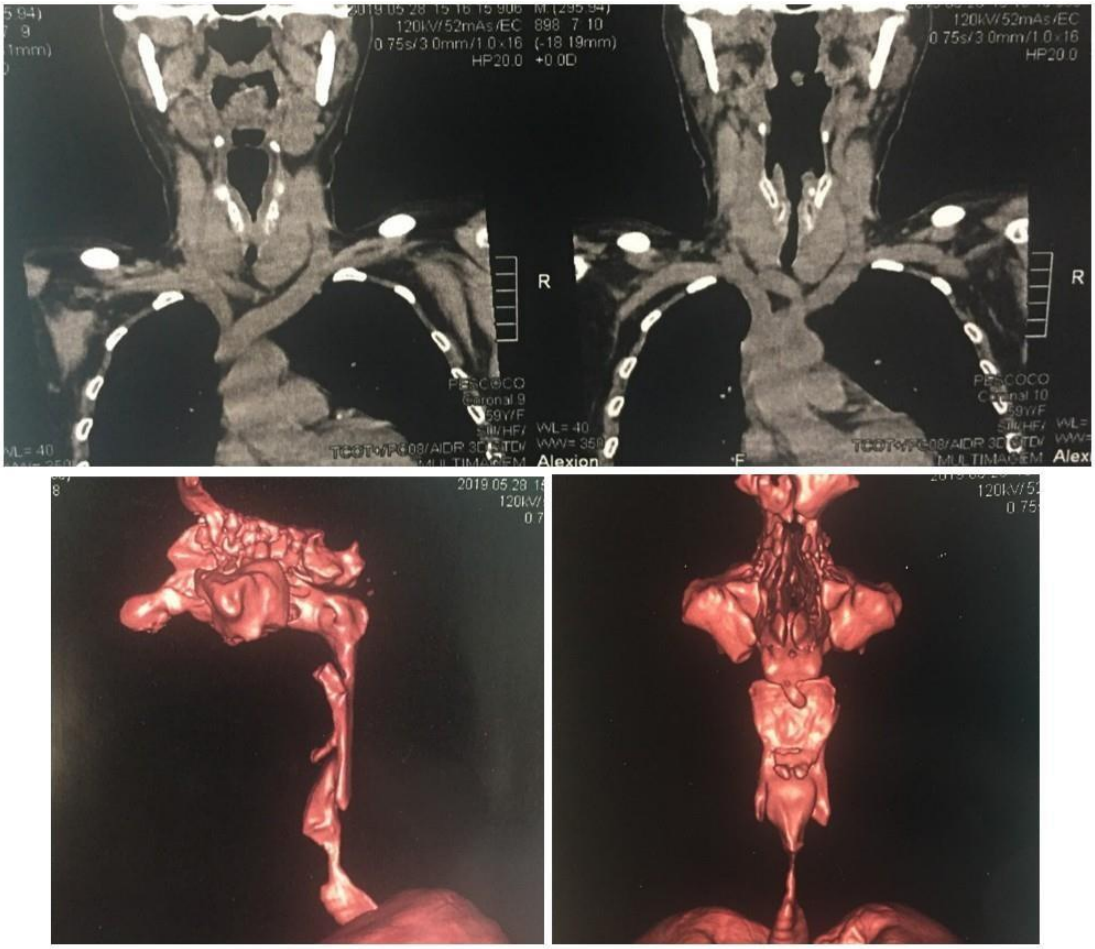


**Figure 01 -** *Computed tomography (CT) of the neck shows small calcification areas in the infraglottic region (proximal cervical trachea). Minimal transverse area of air column measuring 32mm$^2$.*

These findings were supplemented with nasal endoscopy (Figure 2), revealing a Grade III Cotton-Myer laryngotracheal stenosis, with an 80% lumen obstruction and extent to the first tracheal ring, in association with right vocal fold dysmotility. Finally, the patient underwent Magnetic Resonance Imaging (MRI) (Figure 3) which demonstrated stenosis of the proximal trachea, in the infraglottic region, with an area of 10mm$^2$.

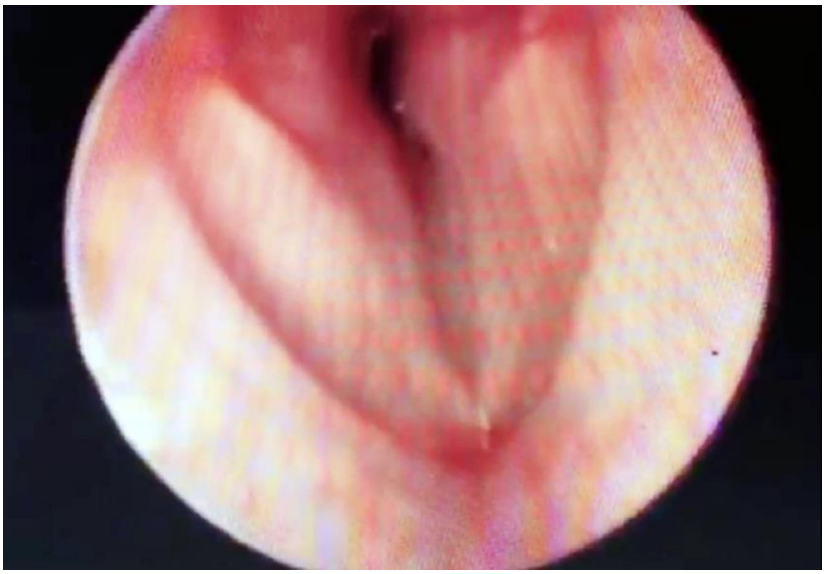

**Figure 02 -** *An 80% laryngeal lumen obstruction observed in the nasal endoscopy*

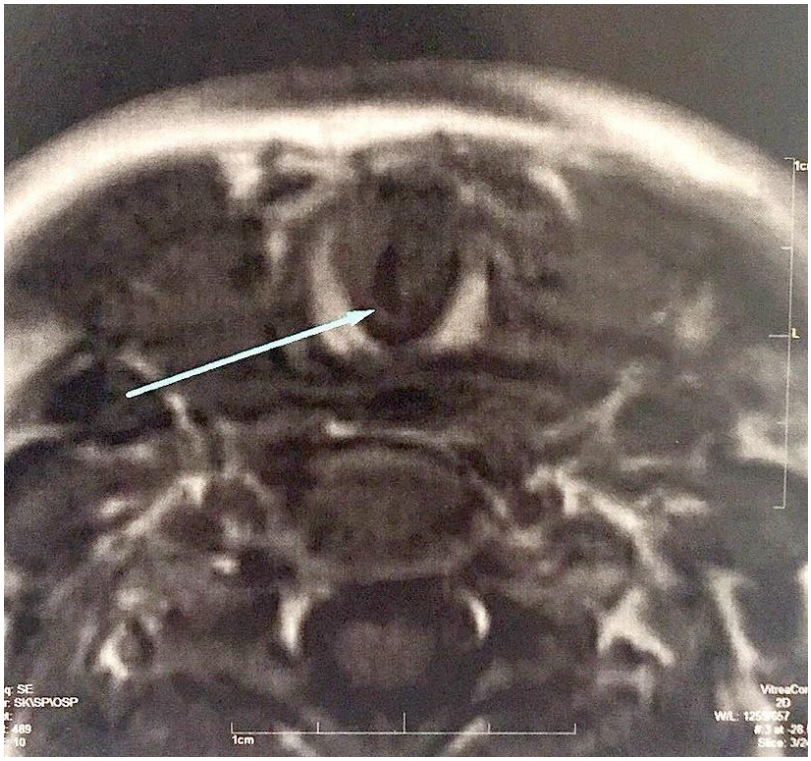


**Figure 03 -** *Proximal trachea, in the infraglottic region, with an area of about 10mm².*

The diagnostic hypotheses suggested included: Leishmaniasis, Wegener's granulomatosis and RP. The patient presented during following investigation Montenegro test with negative results and P- ANCA with positive results. Ultimately, the histological and laboratorial finding, in association with the patient's clinical symptoms, helped determine the diagnosis as RP.

The patient underwent clinical treatment with methotrexate and oral corticosteroids surveilled by a rheumatologist for years, with disease stabilization, despite maintaining dyspnea on small efforts. The patient was then reassessed and underwent surgical correction of the laryngotracheal stenosis with end-to-end anastomosis as well as tracheostomy with size 6 cuffless tube.

## Discussion

RP is a rare connective tissue disease characterized by recurrent episodes of inflammation that affect cartilage in the ears, nose, larynx, tracheobronchial tree, joints and cardiovascular system, with the potential for permanent destruction the affected structures up to fatal outcome [2]. It affects the population between the ages of 20 to 60 years, with no predominance of sex, race[4] and a strong association with HLA-DR4 [2].

The main pathophysiology is considered to be autoimmune [2]. This hypothesis is supported by the finding of antibodies against type II collagen in 2/3 of patients with RP [4].

The diagnosis of the disease is essentially clinical, and biopsy of the compromised structures is recommended only in cases where the clinical picture is atypical [2]. There are currently no specific or conclusive diagnostic tests for RP [2],[3],[4] and histopathological findings are not pathognomonic [2] - however, inflammatory parameters are useful to guide the diagnosis.

The histological findings normally associated with RP are loss of the basophilic characteristic of the cartilaginous matrix, the formation of round perichondral cells and the destruction of the cartilage, with the replacement by fibrous tissue [4].

In early stages, CT shows thickening of the airway wall and attenuation of cartilage, with focal or diffuse narrowing of the lumen in advanced stages, resulting from cartilage destruction and fibrosis1[1].

The therapeutic approach is based on the use of non-hormonal anti-inflammatory drugs, corticosteroids and immunosuppressants [3].

Early onset of primary therapy with oral corticosteroids is associated with a better prognosis [4]. It has been observed that corticosteroid therapy has a positive impact on acute laryngeal inflammation, however it does not affect the prognosis when there is loss of cartilage support [4],5.

Surgery might be necessary to correct stenosis due to airway obstruction, or placing of stents to maintain permeability of the tracheobronchial tree[2].

Reconstruction surgery alone is also associated with greater morbidity and mortality, due to risk of collapse of inflamed tissue during surgical approach, and postoperative complications such as infection, tracheal obstruction[4].

In cases of subglottic involvement, a tracheostomy is recommended. However, this procedure presents a high risk of fatal airway obstruction, as it makes it difficult to perform anesthesia when facing a difficult airway, on account of glottal narrowing due to cartilage destruction [4].

## Conclusion

Autoimmune diseases are of extremely importance amongst the differential diagnosis of larynx's obstructive diseases, due to severe respiratory complications that must be early recognized by the

otolaryngologist.

The diagnosis of RP is challenging, as it is a pathology whose initial symptoms are usually mild and lead to differential diagnoses such as asthma. ormally, respiratory tract involvement is uncommon, being a late manifestation with worse prognosis, among which we highlight laryngotracheal structures.

It is never enough to emphasize the importance of strict monitoring, with a multidisciplinary team in the differential diagnosis and treatment, with experience in postoperative care of laryngotracheal stenosis surgery.

## Declarations

**Funding: Not applicable**

**Conflicts of interest/Competing interests:** The authors whose names are listed immediately below certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

José Antonio Pinto, Heloisa dos Santos Sobreira Nunes, Gabriella Spinola Jahic, Andréia Natalia Azevedo Ferreira de Vasconcelos, Marina Cançado Passarelli Scott, Caio Alonso Orlandini, Cassia Paloma da CunhaOnofre

**Ethics approval:** The study was approved by the National Research Ethics Committee, CAAE: 38664820.0.0000.8114 – Comissão Nacional de Ética em Pesquisa (CONEP, in the Portuguese acronym) and was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki

**Consent to participate:** the patient agreed to surgery and filled out the surgical consent form provided by the Brazilian society of Otorhinolaryngology and Cervico- Facial surgery (ABORL-CCF) – available on their website.

**Consent for publication:** Our team made several attempts to locate the patient's family members, without success or return - the patient in question passed away a few days after the surgical intervention.

We insisted on the case report due to the rarity of the disease and its relevance in early diagnosis to avoid laryngeal complications such as the one that led to the surgical indication of the patient and later to her death. The researcher is committed to maintaining data confidentiality and using the information only for this study and to forward the results of the research for dissemination, thus returning the study to society, with due credit to the authors, through presentation in congress and publication in a journal.
The study was waived from the Informed Consent Form and approved by the National Research EthicsCommittee, CAAE: 38664820.0.0000.8114 –Comissão Nacional de Ética em Pesquisa (CONEP, in the Portuguese acronym).

**Availability of data and material:** Raw data were generated at Núcleo de Otorrinolaringologia e Medicina do Sono de São Paulo. Derived data supporting the findings of this study are available from the corresponding author on request

**Code availability:** CAAE - 38664820.0.0000.8114

**Authors' contributions:** All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by José Antonio Pinto, Heloisa dos Santos Sobreira Nunes, Gabriella Spinola Jahic, Andréia Natalia Azevedo Ferreira de Vasconcelos, Marina Cançado Passarelli Scott, Caio Alonso Orlandini, Cassia Paloma da Cunha Onofre. The first draft of the manuscript was written by Andréia N. A. F. de Vasconcelos, Marina C. P. Scott and Gabriella S. Jahic and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

## Compliance with Ethical Standards

**Disclosure of potential conflicts of interest:** The authors whose names are listed immediately below certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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**Research involving human participants and/or animals:**

This study was a retrospective analysis of medical record data, approved by theNational Research Ethics Committee, CAAE: 38664820.0.0000.8114 –Comissão Nacional de Ética em Pesquisa (CONEP, in the Portuguese acronym) and was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki. The paper does not report on primary research. All data analyzed were collected as part of routine diagnosis and treatment.

Patient was diagnosed and treated according to national guidelines and agreements..

**Informed consent:**

Although this is a Case Report and does not pose any risk to the patient or family members, a waiver was requested regarding the application of the Informed Consent Form to the National Research Ethics Committee (Comitê Nacional de Ética e Pesquisa - CONEP).

Our team made several attempts to locate the patient's family members, without success or return - the patient in question passed away a few days after the surgical intervention.

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