SCIENTIFIC JOURNAL
CEREM-GO

DOI 10.37951/2675-5009.2024v5i14.150
ISSN 2675-5009
SCIENTIFIC ARTICLE - ORIGINAL

RELAPSING POLYCHONDRITIS AND THE CHALLENGE OF DIAGNOSIS: AN EXPERIENCE REPORT

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ABSTRACT

Relapsing polychondritis (RP) is a rare and chronic inflammatory disease characterized by recurrent inflammation of cartilage, primarily affecting the joints, ears, and respiratory airways. Diagnosis is challenging as its symptoms can be confused with other autoimmune conditions. The clinical presentation is variable, including joint pain, hearing loss, skin rashes, and respiratory difficulties. Treatment for RP involves the use of immunosuppressive medications such as corticosteroids, and in more severe cases, methotrexate and cyclophosphamide. Biological therapies have also been successfully employed. This case report discusses a patient with a previous diagnosis of severe refractory asthma, who, after further investigation, was diagnosed with relapsing polychondritis. The patient presented persistent symptoms, such as cough, respiratory difficulty, and nasal changes, which led to the change in diagnosis. Treatment with pulse therapy and immunosuppressive medications was initiated, resulting in clinical improvement. The report highlights the importance of early diagnosis, multidisciplinary follow-up, and personalized treatment, emphasizing the challenges in managing the disease, the need for rigorous monitoring, and the relevance of patient education for treatment adherence and quality of life improvement.

Keywords: Relapsing polychondritis, Early diagnosis, Immunosuppressive treatment.

INTRODUCTION

Relapsing Polychondritis (RP) is a rare and chronic inflammatory disease characterized by recurrent inflammation of the cartilage, especially in the joints, ears, and respiratory airways. The condition is difficult to diagnose, as its symptoms can be mistaken for other autoimmune and rheumatological disorders. The relapsing nature and variable course of the disease require careful and individualized management, aiming to control inflammation and prevent irreversible damage to the affected structures. The clinical presentation is broad and can range

from joint symptoms to more severe complications, such as respiratory problems, making diagnosis and treatment challenging.[1]

Early recognition of RP is crucial for improving patient prognosis, as the disease tends to cause permanent deformities and complications that affect quality of life. Among the most common early manifestations are joint pain, fever, skin rashes, and otolaryngological symptoms such as hearing loss and ear pain, as well as respiratory changes, such as difficulty breathing and airway stenosis. Because it is a disease with symptoms similar to various inflammatory conditions, careful differential diagnosis is required, involving clinical evaluation, laboratory tests, and often biopsies of the affected areas.[2]

The treatment of RP is based on the use of immunosuppressive medications, aiming to control inflammation and prevent further relapses. Corticosteroids, such as prednisone, are frequently used in high doses, but disease management may require the use of more potent immunosuppressive drugs, such as methotrexate or cyclophosphamide, depending on the severity and recurrence of episodes. In some cases, biological therapies have been successfully employed, offering new perspectives for disease control. However, treatment is often challenging, as patients may have varied responses to medications and experience significant adverse effects.[3]

This experience report aims to share the lived experiences of patients with relapsing polychondritis, highlighting the challenges faced in diagnosis and the therapeutic options adopted. Based on this clinical experience, the goal is to provide a comprehensive view of the disease's progression, its implications for treatment, and the importance of multidisciplinary follow-up in managing this condition. The report also seeks to contribute to the understanding of the clinical variability of RP and the need for personalized treatment.

The report also discusses the importance of early therapeutic strategies to prevent permanent sequelae and ensure a better quality of life for the patient. By addressing the challenges faced by both the medical team and the patient, the aim is to offer a reflection on the importance of early diagnosis, appropriate treatment, and continuous patient guidance—key aspects for successful management of relapsing polychondritis.

EXPERIENCE REPORT

A young patient, with a prior diagnosis of severe asthma, had multiple visits to the emergency room and frequent episodes of wheezing and dyspnea since 2019, with a significant worsening over the last 12 months and daily asthma attacks. The condition was exacerbated after a COVID-19 infection in 2020. The daily therapeutic regimen included Alenia, Clenil, and Aerolin, but the patient reported worsening cough with Alenia. A recent spirometry revealed severe obstructive ventilatory disorder. After hospitalization, bronchodilator therapy was initiated, along with treatment for bacterial tracheobronchitis with Tazocin. However, the patient showed refractoriness to bronchodilator measures for severe asthma, with continuous episodes of severe bronchospasm, coarse crackles, diffuse rhonchi, and wheezing.

Due to the persistence of symptoms, a triple therapy was initiated with Trimbow, Clenil, Tiotropium, Salbutamol, and Prednisone. A chest computed tomography (CT) with 3D reconstruction revealed a significant reduction in the caliber of the lower airways, suggesting tracheobronchial chondritis. The patient presented with a saddle nose, persistent cough,

thickening, and calcification of the tracheal wall. The suspicion of relapsing polychondritis was confirmed based on the clinical criteria of McAdam and Damiani and Levine, in addition to the exclusion of other autoimmune conditions with a negative ANCA.

According to McAdam's criteria, the patient met two criteria (nasal and respiratory chondritis). The criteria of Damiani and Levine also confirmed the diagnosis, as they showed chondritis in two distinct anatomical regions with a response to corticosteroids.

Based on the diagnosis, pulse therapy with methylprednisolone was initiated for three days. During hospitalization, the patient developed an Influenza A infection, which was treated with oseltamivir. Later, a multisensitive Pseudomonas aeruginosa infection was diagnosed and treated with levofloxacin. The immunosuppressive therapy included methotrexate, folic acid, prednisone, Trimbow, and Clenil, resulting in gradual improvement. The patient was advised on tracheostomy in case of refractoriness and was discharged for outpatient follow-up with Pulmonology and Rheumatology.

DISCUSSION

There is diagnostic complexity in patients with relapsing polychondritis (RP) due to symptom overlap with other conditions, such as severe asthma. The diagnosis was made more difficult by multiple visits to the emergency room and lack of consistency in the doctor-patient relationship. The presence of multiple complications, such as nasal and tracheobronchial chondritis and structural changes detected on the CT scan, raised the suspicion of RP.[4,5]

RP is a rare autoimmune disease, and its confirmation requires access to advanced tests and a multidisciplinary team.[6] This case highlights the need for a robust differential diagnosis in patients with chronic respiratory symptoms and a history of refractoriness to standard treatment. The involvement of distinct anatomical areas, such as the airways and nasal cartilage, was essential for confirmation based on the McAdam and Damiani and Levine criteria.[7,8]

Proper management requires vigilance for infectious complications, as immunosuppressive treatments, such as methotrexate and corticosteroids, increase susceptibility to infections.[9] A multidisciplinary approach with Pulmonology and Rheumatology, along with outpatient follow-up, is essential to prevent recurrences and adjust therapeutic management.[10]

Casos como este reforçam a importância de investigar associações entre PR e outras manifestações sistêmicas, como oftalmológicas e neurológicas, destacando a relevância do diagnóstico precoce para evitar complicações graves e otimizar a qualidade de vida do paciente.

CONCLUSIONS

This experience report highlights the importance of early diagnosis and proper management of relapsing polychondritis, a rare and challenging condition for both healthcare professionals and patients. This case underscores the importance of thorough investigation in patients diagnosed with severe asthma refractory to optimized treatment, especially in cases where symptoms persist despite appropriate behavioral and environmental changes and optimized therapy.

Relapsing polychondritis, an autoimmune and progressive disease, if not diagnosed and treated appropriately, can lead to severe complications. A differentiated approach and careful review of the diagnosis are essential to ensure symptom control and the patient's quality of life.

Effective treatment requires a personalized approach, considering the relapsing nature and the diversity of clinical manifestations of the disease. The use of immunosuppressive medications, along with strict follow-up, is crucial to control inflammatory episodes and minimize irreversible damage to the affected structures.

Despite advances in treatment, the clinical variability of relapsing polychondritis makes disease control complex, with many patients facing difficulties with medication side effects and frequent relapses. Therefore, the need for a multidisciplinary approach, involving rheumatologists, otolaryngologists, pulmonologists, and other specialists, is essential to ensure effective treatment and the best possible prognosis for the patient. Furthermore, psychological support and patient education about the disease are crucial to improve treatment adherence and quality of life.

The control of inflammation and the prevention of new episodes are the pillars of managing relapsing polychondritis, and the combination of conventional and biological therapies offers new perspectives in the treatment of the disease. Finally, this report contributes to the understanding of the nuances of relapsing polychondritis and serves as a reference for healthcare professionals dealing with similar cases. The constant evolution in the understanding of the disease and therapeutic options is essential to provide patients with a more balanced life and reduced impact from complications. Ongoing education about the disease and advances in treatment are crucial to achieving better clinical outcomes and promoting patients' quality of life.

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Library Review - Romulo Arantes
Spell Check: Dario Alvares
Received: 11/12/24. Accepted: 12/12/24. Published in: 15/01/25.