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Case Presentation

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Relapsing Polychondritis with Non-Cardiac Pulmonary Edema Presenting as Acute Breathlessness in a Postpartum Female: A Case Report

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Abstract

Introduction: Relapsing polychondritis is a rare autoimmune disorder characterized by progressive inflammation of cartilaginous structures. This case report aims to present the unique presentation of RP as acute breathlessness in a postpartum female.

Case report: A 29-year-old female, one day post-elective lower segment cesarean section, presented with sudden shortness of breath, weakness, and sweating. Examination revealed bilateral lung

abnormalities, and diagnostic assessments, including chest CT and PET-CT scans, confirmed RP. Treatment with CPAP (continuous positive airway pressure) and Prednisolone initiated a positive response, with subsequent methotrexate introduction in follow-ups. The patient's clinical course demonstrated improvement in symptoms and inflammatory markers.

Discussion: This case highlights the importance of considering RP in the differential diagnosis of acute breathlessness. Early intervention with corticosteroids and immunosuppressive therapy proved effective, aligning with existing literature on RP management.

Conclusion: Timely diagnosis and treatment are important in managing RP, as illustrated by this case. The positive response to CPAP (continuous positive airway pressure), corticosteroids, and methotrexate highlight the potential efficacy of this therapeutic approach. This report contributes valuable insights into the condition's clinical spectrum, emphasizing the significance of a multidisciplinary approach in the care of patients with autoimmune disorders.

Keywords*:* Relapsing polychondritis; Postpartum; Breathlessness; Case report

Introduction

Relapsing Polychondritis (RP) is a rare autoimmune disorder affecting multiple systems including the connective tissue. Characterized by severe, episodic, and progressive inflammation, RP involves various cartilaginous structures, including the nose, ears, larynx, trachea, blood vessels, heart, cornea, sclera, kidney, and joints **[1]**. Jaksch-Wartenhorst documented the first case in 1923, and the nomenclature "relapsing polychondritis' ' was coined in the 1960s by Pearson et al **[2]**. With an onset possible at any age, RP typically has a peak incidence between 40 to 50 years, with approximately 50% of patients experiencing laryngeal and tracheal involvement. This case report contributes to the understanding of RP by highlighting its unique first presentation as postpartum acute breathlessness, offering valuable insights into the diverse clinical manifestations of this complex autoimmune disorder. This case report has been reported in accordance with the CARE checklist **[3]**.

Case Presentation

A 29-year-old female, one day post-elective lower segment cesarean section for oligohydramnios at 38+ weeks of gestation, presented with sudden shortness of breath, sweating, weakness, and difficulty expectorating. The patient had a history of childhood asthma, pneumonia, and underwent septoplasty for atrophic rhinitis 10 years back. On examination, the patient appeared lethargic, with a respiratory rate of 22 breaths per minute and oxygen saturation of 92% on 2 liters of oxygen via nasal prongs. Bilateral rhonchi and crepitations were noted, more on the right side. Her heart rate was 94 bpm, blood pressure 120/70 mmHg and her temperature was 99°F. Abdominal examination revealed a soft abdomen with mild tenderness over the lower abdomen. Bowel sounds were present. A chest x-ray **(Figure 1)** and laboratory investigation was sent. Laboratory values are tabulated in **Table 1**.

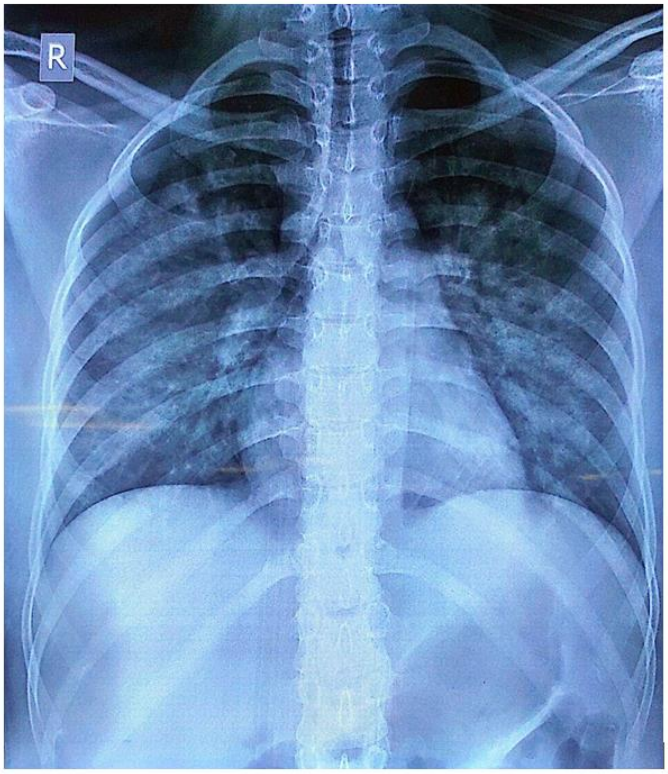

Figure 1: CXR PA view showing prominent interstitial lung markings, suggestive of pulmonary edema.

Table 1: Clinical Laboratory Values.

Lab reference	Reference range	Laboratory results
Hemoglobin	11-15 gm/dl	11.5 gm/dl
Platelet count	150000-450000/cu.mm	180000 /cu.mm
White Blood Cell	4000-11000/cu.mm	21,840 /cu.mm
Neutrophils	40-75%	89%
CRP	0-6 mg/mL	75.6 mg/dl
ESR	0-20 mm/hr	35 mm/hr
Rheumatic factor	<8	<8
Antinuclear cytoplasmic antibodies **(**C-ANCA**)**	<20.0 RU/ml	2.02 RU/Ml
Myeloperoxidase	<20.00 Ru/ml	4.03 RU/Ml

We suspected postpartum cardiomyopathy due to pulmonary edema. Echocardiography, NT-pro BNP, and ECG were performed. However, these tests showed normal results. Subsequent chest Computed Tomography (CT) with inspiratory and expiratory phase was done and revealed consolidation and ground glass opacities, predominantly in bilateral perihilar distribution. Additionally, there was bilateral smooth interlobular septal thickening and minimal pleural effusion with fissural extension

suggestive of non-cardiac negative pressure pulmonary edema. The examination also indicated thickening of the anterior and lateral walls of the trachea and bilateral main bronchi with areas of calcification, sparing the posterior wall. Tracheal narrowing with luminal irregularities was observed, consistent with features supporting relapsing polychondritis **(Figure 2)**.

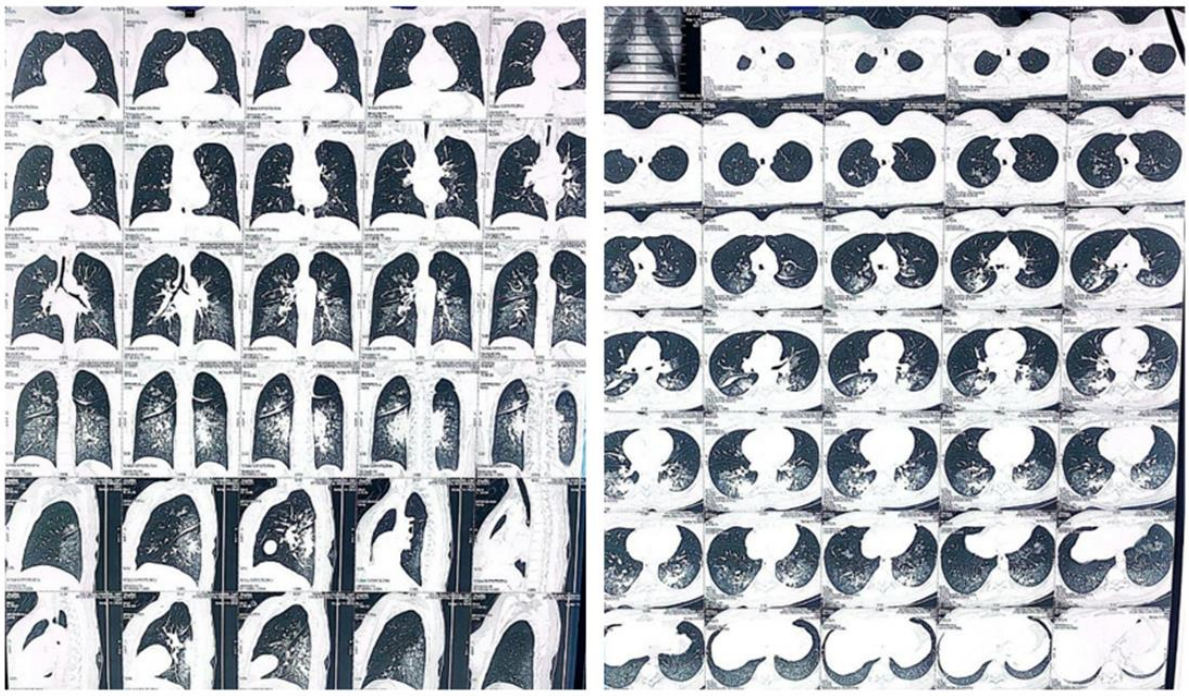

Figure 2: CT scan of the chest.

Positron emission tomography (PET) CT scan of the chest, abdomen, and musculoskeletal was conducted to confirm the diagnosis **(Figure 3)**. PET CT revealed evidence of mild hypermetabolism in the bilateral nasal ala and external auditory canal, along with luminal narrowing without calcification of the trachea. No abnormalities were detected in the abdomen and musculoskeletal system.

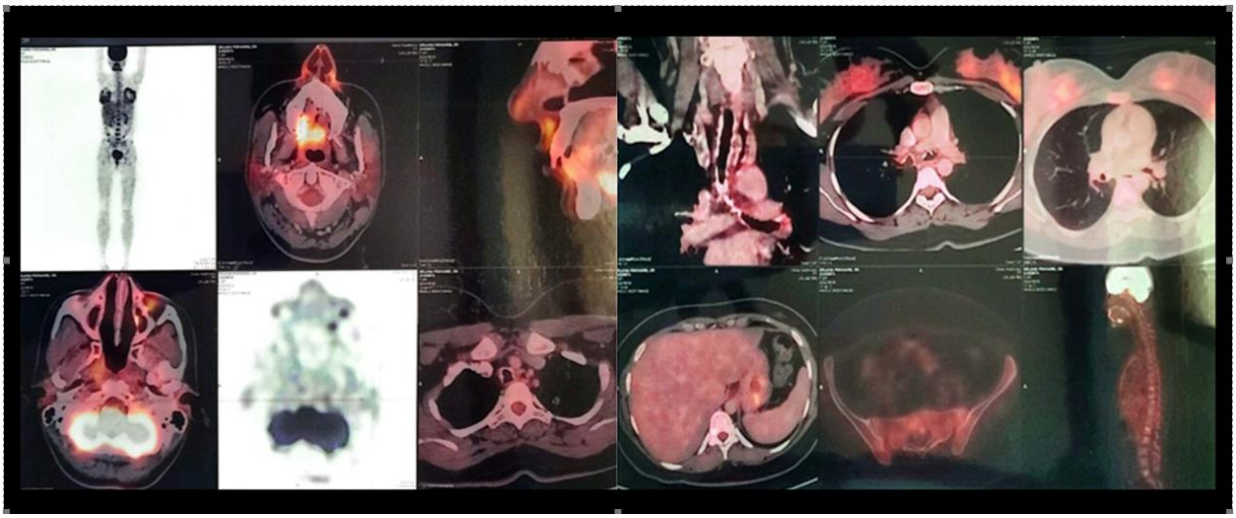

Figure 3: PET CT scan of head and neck, abdomen, axial and appendicular skeleton.

The patient was diagnosed with relapsing polychondritis with non-cardiac negative pressure pulmonary edema. Treatment consisted of Continuous Positive Airway Pressure (CPAP) for four hours, N-acetylcysteine 100 mg twice daily and nebulization with salbutamol solution for pulmonary edema. Patient was discharged on prednisone 20 mg once daily for one month, Vitamin D and calcium supplements were prescribed as well. The patient also received a dose of pneumococcal and influenza vaccine. The prednisone dosage was gradually tapered, reaching 2.5 mg once daily. Subsequently, methotrexate at 10 mg weekly, along with folic acid at 5 mg on alternate days, was started. The patient's acute phase response, indicated by sedimentation rate and C-reactive protein, recovered (35 mm/h and 2.82 mg/dL, respectively). The patient remains under regular follow-up, displaying stability in their clinical condition.

Discussion

Table 2: Patient Evaluation under McAdam et al. (1976), Damiani and Levine, 1979, and Michet et al, 1986 criteria of relapsing polychondritis **[4]**.

Clinical criteria	Patient Presentation
McAdam et al, 1976 **[5]** 1. Recurrent chondritis of both auricles 2. Non Erosiveive inflammatory polyarthritis. 3. Chondritis of nasal cartilages 4. Inflammation of ocular structures 5. Chondritis of respiratory tract 6. Cochlear and/or vestibular damage.	• • • •

Three out of six criteria must be fulfilled for the diagnosis to be established.	
Damiani and Levine, 1979 **[6]** 1. One out of six McAdam et al.'s criteria and a positive histologic confirmation 2. Two out of six MAdam et al.'s criteria and response to corticosteroid or dapsone Any of these requirements be met	• (biopsy was not done) +
Michet et al. 1986 **[7]** 3. Proven inflammation in two out of three cartilages: auricular, nasal, and laryngotracheal 4. Proven inflammation in one of the above and meeting two other signs from ocular inflammation, hearing loss, vestibular dysfunction, or seronegative inflammatory arthritis Any of these requirements be met	+ -

Relapsing Polychondritis is diagnosed using the criteria's listed above **(Table 2)**. The diverse clinical presentations of RP, ranging from intermittent painful ear cartilage episodes to life-threatening airway collapse, shows the variability of this immune-mediated disorder **[8,9]**. For this reason, RP can be challenging to diagnose. There is a mean diagnostic delay of 2.9 years **[10]**. Relapsing polychondritis commonly presents with auricular chondritis in 80-90% of patients, followed by ocular manifestations in 50-60% of cases **[11]**. Notably, the typical symptoms of auricular chondritis were absent in this patient. The patient exhibited large airway involvement, with postpartum shortness of breath. Only two other cases of postpartum onset of RP have been reported. Both presented as bilateral auricular perichondritis, bilateral conjunctivitis, bilateral sensorineural hearing loss, and bilateral tinnitus. Pregnancy as a triggering factor for RP remains unexplored **[12]**. The potential lethality of RP shows the critical importance of timely diagnosis and management. Mortality results from complications such as pneumonia or systemic vasculitis-related issues like aneurysm, renal failure, and valvular involvement. The etiology of Relapsing Polychondritis (RP) remains complex and elusive, involving a combination of genetic, environmental, and immunological factors **[1]**. RP shows a significant association with HLA DR4 **[13]**. An autoimmune reaction to type II, IX, and XI collagen, which is abundant in sclera and cartilage, is implicated in the pathogenesis of Relapsing Polychondritis **[4]**. Triggering factors for RP may include trauma, infectious agents, and toxic agents. However, the precise mechanisms triggering the inflammatory response in cartilaginous structures remain unknown **[13]**.

Management of RP involves NSAIDs, systemic glucocorticoids and dapsone; alternative options include methotrexate, azathioprine, leflunomide, colchicine, and cyclosporine, if the initial response is inadequate **[4]**. In this case, the patient was initially treated with oral glucocorticoid and methotrexate was added on a later date. Strengths of this case report lie in its comprehensive detailing of the diagnostic process, treatment regimen, and the patient's response to therapy. However, limitations include the rarity of RP, making it challenging to generalize findings to a broader population. Additionally, the long-term

outcomes of the therapeutic approach require ongoing evaluation.

Conclusion

This case report highlights relapsing polychondritis (RP), an autoimmune disorder affecting cartilaginous structures. The comprehensive diagnosis and treatment, including prednisone, and vaccination, followed by methotrexate, resulted in successful management. Positive patient response, marked by the resolution of symptoms and acute phase markers, emphasizes therapeutic efficacy. RP's complex etiology necessitates ongoing exploration of genetic, environmental, and immunological factors. Recognizing RP's variable presentations and employing personalized therapeutic approaches are crucial for optimal outcomes. This case highlights the need for continued patient monitoring and research to refine treatment protocols in the evolving landscape of RP management.

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