

Cryptogenic Organizing Pneumonia: A Case Report



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ABSTRACT

Cryptogenic organizing pneumonia (COP) is an idiopathic interstitial lung disease affecting the distal airways, characterized by the development of granulation tissue that obstructs the bronchioles and alveoli, leading to respiratory failure. This case report describes a 63-year-old female patient with a history of diabetes, hypertension, and hypothyroidism who presented with persistent productive cough and dyspnea, initially treated as community-acquired pneumonia. Despite empirical antibiotic therapy, the patient's symptoms persisted. Further investigation, including high-resolution CT (HRCT) scans and a CT-guided lung biopsy, revealed fibrotic exudates, interstitial fibrosis with inflammatory infiltrates, and epithelioid granuloma. A diagnosis of COP was made after multidisciplinary discussion, and corticosteroid therapy was initiated, leading to significant clinical improvement and resolution on repeat imaging. This case highlights the importance of considering COP in patients with nonresolving pneumonia and underlines the efficacy of corticosteroids in its management. It also emphasizes the need for a multidisciplinary approach combining clinical, radiological, and histological assessments to reach a definitive diagnosis. Early recognition and appropriate treatment are crucial in preventing complications such as fibrosis and respiratory failure.

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INTRODUCTION

Cryptogenic organizing pneumonia (COP) is a subtype of interstitial lung disease and the idiopathic form of organizing pneumonia (OP) (formerly called bronchiolitis obliterans organizing pneumonia), affecting distal bronchioles, respiratory bronchioles, alveolar ducts, and alveolar walls. It is marked by the development of organized clusters of granulation tissue that block the alveolar spaces and bronchioles, leading to respiratory failure.¹⁻³ The exact etiology of OP is unknown. Several potential causes have been proposed for OP, including viral infections, exposure to toxic gases, certain medications, gastroesophageal reflux, radiation therapy, and connective tissue disorders.⁴ OP is considered a clinicopathological condition, meaning that while clinical and radiological findings can raise suspicion for the diagnosis, histological confirmation through tissue biopsy is necessary for a definitive diagnosis. As such, diagnosing OP requires a combination of clinical presentation, imaging studies, and pathological examination.^{5,6} Here, we outline a case of an elderly female presenting with nonresolving pneumonia diagnosed as COP and treated successfully with corticosteroids.

CASE

A 63-year-old female, known case of type 2 diabetes, systemic hypertension, and hypothyroidism, presented with complaints of productive cough and dyspnea for

5 days and fever for 2 days. Her chest X-ray PA view showed left lower zone alveolar inhomogeneous opacities. HRCT thorax showed left lower lobe consolidation (Fig. 1). Sputum and BAL evaluation were negative for tuberculosis. She was diagnosed with community-acquired pneumonia, treated with empirical antibiotics, and discharged.

The patient revisited our OPD after 4 weeks with persistent cough and occasional fever; hence, an alternative diagnosis was considered for nonresolving pneumonia, including tuberculosis, OP, and malignancy. She was reevaluated with CECT thorax (Fig. 2), which showed persistent, denser consolidation in the left lower lobe with no contrast enhancement. Antinuclear antibody (ANA) profile was negative.

The patient underwent CT-guided biopsy, which showed many fibrotic exudates filling the alveolar sacs and ducts. Dense interstitial fibrosis with mixed inflammatory cells was noted; some alveoli were dilated and showed foamy macrophages. Many well-formed epithelioid cell granulomas with central suppurative inflammation and foci of necrosis were seen. Few Langhans-type giant cells were also noted (Fig. 3). Biopsy specimen GeneXpert was negative. There was a diagnostic dilemma between tuberculosis and OP. After a multidisciplinary discussion involving pulmonology, infectious disease specialists, and pathology, the team decided to initiate corticosteroid therapy tapered over 8 weeks. During follow-up, both the bronchoalveolar lavage (BAL) and biopsy

mycobacteria growth indicator tube (MGIT) cultures remained negative, and repeat imaging showed resolution (Fig. 4).

DISCUSSION

The primary objective of this case study was to evaluate the diagnosis and treatment of a 63-year-old female patient with nonresolving pneumonia, ultimately diagnosed as COP after a thorough clinical, radiological, and histopathological examination. The case emphasizes the importance of considering COP in patients presenting with persistent pneumonia. The patient initially presented with productive cough, dyspnea, and fever, which persisted despite empirical treatment for community-acquired pneumonia. Follow-up imaging revealed persistent lung consolidation. After ruling out tuberculosis and malignancy, a CT-guided biopsy revealed features characteristic of COP, including fibrotic exudates and interstitial fibrosis. The patient was successfully treated with corticosteroids, resulting in clinical and radiological resolution.

Cryptogenic organizing pneumonia generally affects individuals in their 50s or 60s, with the onset of symptoms being gradual and often mistaken for other respiratory conditions. Commonly, patients present with nonspecific symptoms like fever, malaise, persistent cough, and progressive shortness of breath, which can last for several weeks. These symptoms often lead to misdiagnosis as bacterial pneumonia, especially since they do not respond to standard antibiotic treatments.

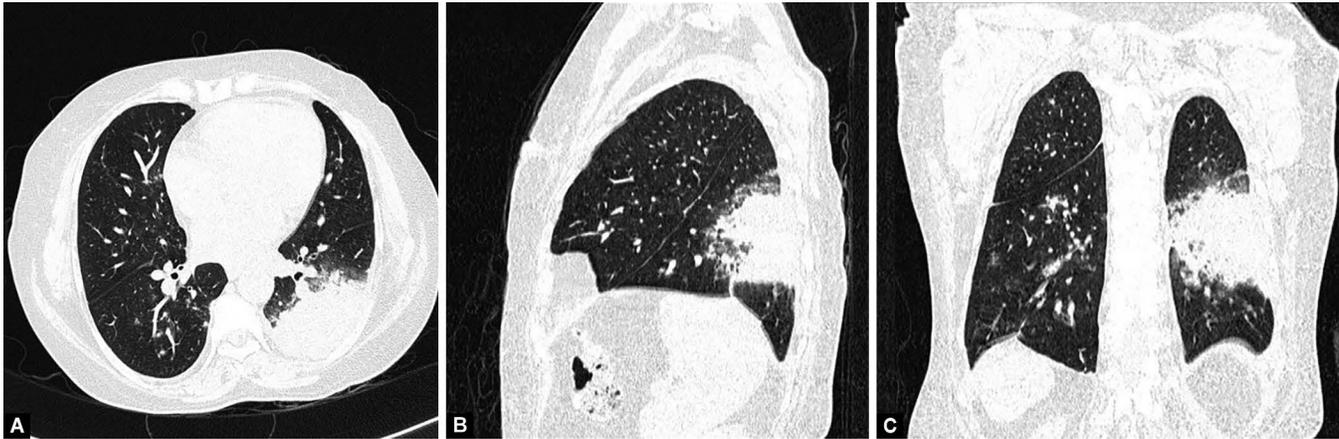
In most cases, the severity of symptoms is mild to moderate, but there are instances where patients may experience more severe

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Figs 1A to C: HRCT thorax (6th April 2024)—left lower lobe consolidation



Figs 2A to C: CECT thorax (4th May 2024)—left lower lobe dense persistent consolidation

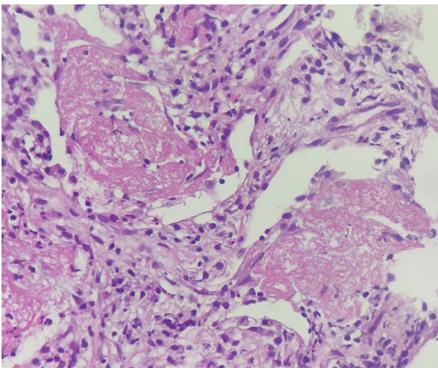


Fig. 3: HPE of CT-guided lung biopsy (4th May 2024)—many polypoidal fibrotic plugs filling the alveolar sacs and ducts (Masson bodies) with dense interstitial fibrosis and mixed inflammatory cells

complications, including significant shortness of breath (dyspnea) and low blood oxygen levels (hypoxemia). These severe cases can become concerning, as they might require more aggressive treatment, including hospitalization and oxygen therapy.

Less commonly, individuals with COP may experience additional symptoms like chest pain, unexplained weight loss, night sweats,

and joint pain (arthralgias). These atypical symptoms can make diagnosis challenging, especially when they mimic other conditions such as tuberculosis, lung cancer, or interstitial lung disease.

The precise cause of COP remains unknown, earning it the term “cryptogenic.” However, it is widely believed that the condition results from injury to the alveolar epithelium, the thin cells lining the air sacs of the lungs. This damage might be triggered by an underlying factor that remains unidentified. Several potential triggers have been proposed, including viral infections (such as those caused by influenza or other respiratory viruses), inhalation of toxic gases, adverse reactions to certain medications, or even gastroesophageal reflux. Other possible triggers include prior radiation therapy and connective tissue disorders like rheumatoid arthritis or lupus.⁴

The pathogenic mechanism involves alveolar epithelial injury causing plasma protein leakage, which recruits inflammatory cells. This process progresses in three stages: fibrin formation and inflammation, fibroblast proliferation with re-epithelialization, and

organization of fibroblasts and connective tissue matrix. Vascular endothelial growth factor (VEGF) and fibroblast growth factors play key roles, while glucocorticoids can inhibit granulation tissue formation in experimental models.^{7–10}

This case is consistent with established studies on COP, where the condition is frequently misdiagnosed as bacterial pneumonia initially. The patient’s presentation aligns with documented symptoms of COP, such as persistent cough, fever, and respiratory difficulty despite treatment. The use of corticosteroids in this case also mirrors standard management protocols found in existing literature, which advocate for their effectiveness in resolving COP-related inflammation.¹¹

Before confirming the COP diagnosis, other potential causes for nonresolving pneumonia, such as tuberculosis and malignancy, were carefully considered and ruled out. Although these conditions can present similarly, the absence of supportive clinical, microbiological, and radiological findings for these alternative diagnoses directed attention toward COP.

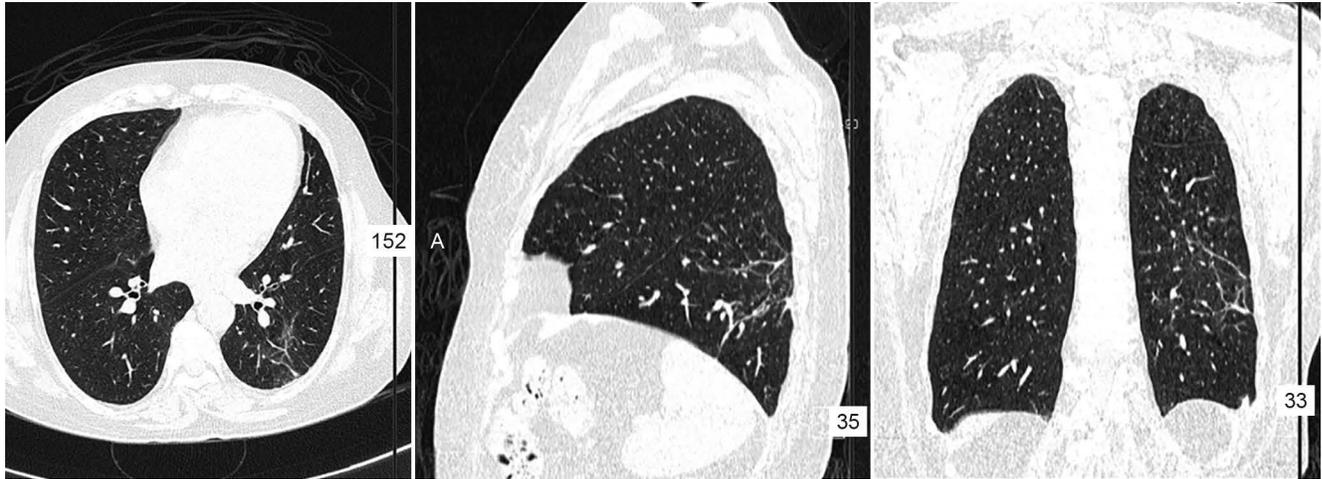


Fig. 4: CECT thorax (26th July 2024)—resolution after corticosteroid treatment

This case highlights the importance of considering COP in patients with nonresolving pneumonia and emphasizes the value of a multidisciplinary approach involving clinical, radiological, and histopathological assessments. Early recognition and treatment with corticosteroids can lead to symptom resolution and prevent complications such as fibrosis and respiratory failure.

A limitation of this case is the inability to identify a definitive etiology for the OP, as the condition remained cryptogenic. Additionally, while corticosteroid treatment was successful, the long-term follow-up to monitor for potential relapse or steroid-related side effects was not detailed in this case. Future research could focus on identifying potential triggers or early biomarkers for COP to aid in earlier diagnosis. Studies on the long-term outcomes of patients treated for COP, including relapse rates and steroid-related complications, would provide valuable insights into optimizing treatment strategies.

CONCLUSION

This case underscores the importance of considering COP in patients with persistent pneumonia-like symptoms that do not

respond to standard treatments. The prompt use of corticosteroids led to a successful outcome, reinforcing their role as the primary treatment for COP. Early and accurate diagnosis remains key to managing the condition effectively and preventing long-term complications.

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