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A case report of unresolved pneumonia in an adult patient in ICU

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Introduction

Patients with community-acquired pneumonia (CAP) typically show significant improvement within 3-5 days. However, if symptoms persist or radiologic infiltrates remain despite adequate antibiotic treatment, it's considered non-resolving pneumonia. (1) To address this, healthcare providers should first rule out alternative diagnoses such as tuberculosis, cancer, or other conditions that mimic pneumonia. Next, they should assess the effectiveness of treatment and patient compliance. It's also crucial to identify any underlying factors that may be contributing to immunodeficiency, such as chronic obstructive pulmonary disease, diabetes, intra-bronchial construction, alcoholism, smoking, malignancy, or human immunodeficiency virus (HIV) infection, as well as potential complications like necrotizing pneumonia, empyema or superinfection. In some cases, a rare or unexpected cause may be underlying the non-resolving pneumonia.

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Case presentation

A 43-year-old male was transferred to our hospital for unresolved pneumonia due to an endobronchial mass obstructing the left main bronchus, resulting in the collapse of the left lung and a whole opacification of the left hemithorax (**Figure 1**), with hypoxia requiring Intensive Care Unit (ICU) admission.

The patient reported fever, vomiting, a frequent cough mixed with brownish sputum, and increasing shortness of breath. Three months prior, the patient underwent human immunodeficiency virus (HIV) and tuberculosis (TB) tests and bronchoscopy (for bronchoalveolar lavage [BAL], acid-fast bacillus [AFB], and cytology) at another hospital, and the findings were negative for any particular illnesses.

Upon assessment, he was conscious but had respiratory discomfort. His heart rate was 96 beats per minute, respiration rate was 28 breaths per minute, and oxygen saturation was 92% with 2 l oxygen per nasal cannula. His temperature was 39.2 °C.

After being brought to our ICU, the patient experienced a crisis of flushing throughout the face and the belly, in addition to palpitations, stomach discomfort, and inspiratory/expiratory wheezing within all lung fields, both on the left and right sides. Considering that this could be a carcinoid crisis, both serum serotonin as well as urine 5-hydroxyindoleacetic acid were tested and were confirmed to be negative. The patient had a continuous fever and productive cough with brownish sputum. The sputum culture had mixed growth. Screening for the pulmonary tuberculosis panel was again repeated, with negative findings. He was started on Tamiflu 75 mg twice daily and shifted from itraconazole to caspofungin intravenously.

Key words: ICU, pneumonia, Chédiak-Higashi syndrome.

Peripheral antineutrophil cytoplasmic antibodies, cytoplasmic antineutrophil cytoplasmic antibodies, anti-deoxyribonucleic acid, antiphospholipid, and carcinoembryonic antigen tests were negative.
The chest computerized tomography (CT) scan revealed that the left major bronchus was obstructed entirely up to its bifurcation (**Figure 2**). The Intensive Care team, the pulmonary team, and a visiting infectious disease specialist convened a multidisciplinary discussion attended by professionals from numerous fields. The diagnosis made was that it was an instance of Chédiak-Higashi syndrome (CHS) accompanied by invasive lung aspergillosis, as indicated by the individual's medical examination of the patient's sibling. The patient had a repeated bronchoscopy, and a bronchoscopic biopsy from the endobronchial mass showed necrotic granulation tissue containing little atypical squamous epithelium and benign cartilage. No viable invasive malignancy was found in the submitted sections.
As a result, it was strongly suspected that he had invasive pulmonary aspergillosis. Following that, the choice was to start voriconazole IV infusion 400 mg twice a day and the maintenance dose of 200 mg IV infusion twice daily. Significant clinical improvement was noticed in 7 days, and a follow-up chest X-ray after one week showed a significant radiological resolution (**Figure 3**).
The patient was hemodynamically stable on room air and afebrile, with no vomiting. The sputum looked good, so the patient was discharged to the ward.

Discussion

In this case, we will describe a rare occurrence of CHS in the adult male and highlight the complexity of treating CHS and invasive pulmonary aspergillosis.
As we know, CHS is caused mainly by mutations in the lysosomal trafficking regulator (LYST) gene and manifests as immunodeficiency, partial albinism, and neurological disorders. It is extremely rare in adult males, and the coexistence of these diseases highlights the interaction between genetic susceptibility and opportunistic infections, which presents significant challenges for diagnosis and treatment. The rareness of this combination shows the need for further research, focusing on understanding the pathology, diagnostic difficulties, and tailored therapies. Early recognition of symptoms is critical because CHS is rare in adulthood and can overlap with other respiratory diseases, making diagnosis and treatment difficult.
This case report focuses on the complex interaction between CHS, a rare genetic disorder, and invasive pulmonary aspergillosis (IPA). It emphasizes the need for a comprehensive understanding of their pathophysiology and individualized treatment strategies. (2) The malfunctioning of neutrophils and macrophages predisposed him to IPA caused by opportunistic fungal infections, especially in Aspergillus species. (2) This case highlights the challenges associated with CHS and co-morbidities such as IPA. CHS, characterized by albinism, recurrent infections, and neurological disorders, is caused by LYST gene mutations. The diagnosis is based on detecting abnormal granules of leukocytes, and treatment mainly involves allogeneic hematopoietic stem cell transplantation (HSCT). However, this does not prevent neurological progression; a multidisciplinary treatment is essential. Some new treatments, such as gene editing, offer hope, but their clinical utility requires further investigation due to CHS's rarity and complex treatment. (3)
Some initial symptoms, including fever and respiratory distress, have presented a diagnostic challenge despite extensive research, (4) raising concerns about IPA associated with CHS based on family history and clinical presentation. (4) The advanced recognition and prompt treatment, including antifungal therapy with caspofungin, itraconazole, and voriconazole, resulted in clinical improvement. (4) The administration of voriconazole has shown significant efficacy in treating CHS IPA, according to the literature's recommendations. (5,6) Yet, the remaining complications, such as a suspected endobronchial mass, emphasized the need for continuous postoperative monitoring and treatment. (6,7) The main complexity of managing CHS and IPA requires a simultaneous multidisciplinary approach and a thorough risk assessment. (6,7) Hines et al. suggested that chimeric antigen receptor T-lymphocyte cell therapy may exacerbate complications in patients with CHS, highlighting the importance of tailored treatment plans and scoring systems for hyperinflammatory disorders and life-threatening complications such as IPA. (7)
Some emerging treatments, such as gene editing and gene therapy, offer promising treatments to correct the genetic defects underlying CHS and related immunodeficiencies, as discussed in Kang's study. (8) However, their clinical use requires further investigation of feasibility and appropriateness, acknowledging limitations such as the rarity of adult-onset CHS and the complexity of simultaneous management of CHS and IPA, which may require multidisciplinary involvement and close medication monitoring.

Conclusion

The present case illustrates difficulties in treating and spotting IPA in a male adult with CHS. This stresses the importance of being watchful for infectious diseases among people with weakened immune systems, quickly recognizing atypical clinical pictures, conducting timely diagnosis tests, and administering selective antifungal drugs against these infections.

Source of support
Nil.

Conflict of interest
None declared.

Figure 1. Chest radiography on admission to the Intensive Care Unit

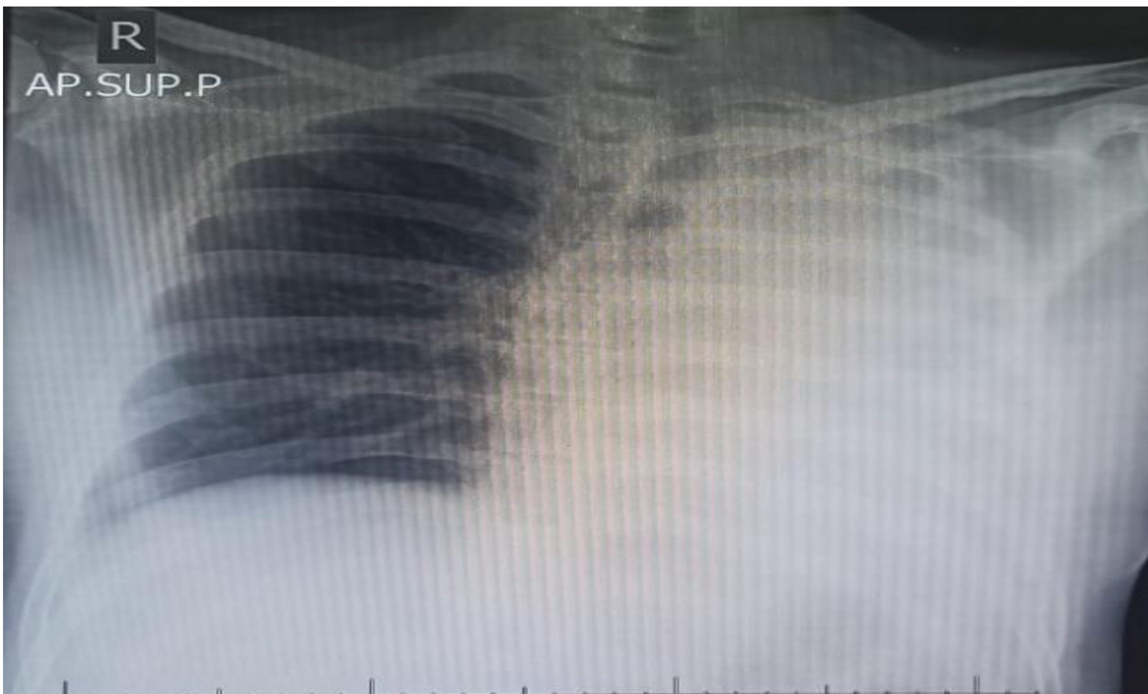


Figure 2. Computerized tomography scan on admission to the Intensive Care Unit

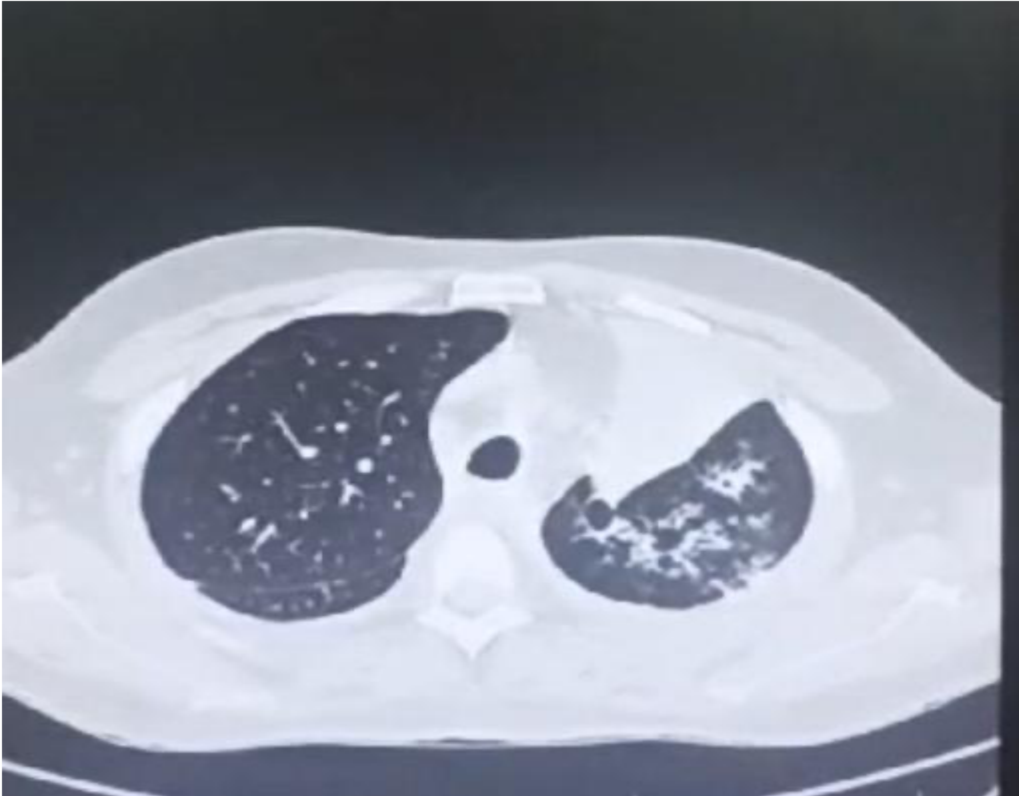
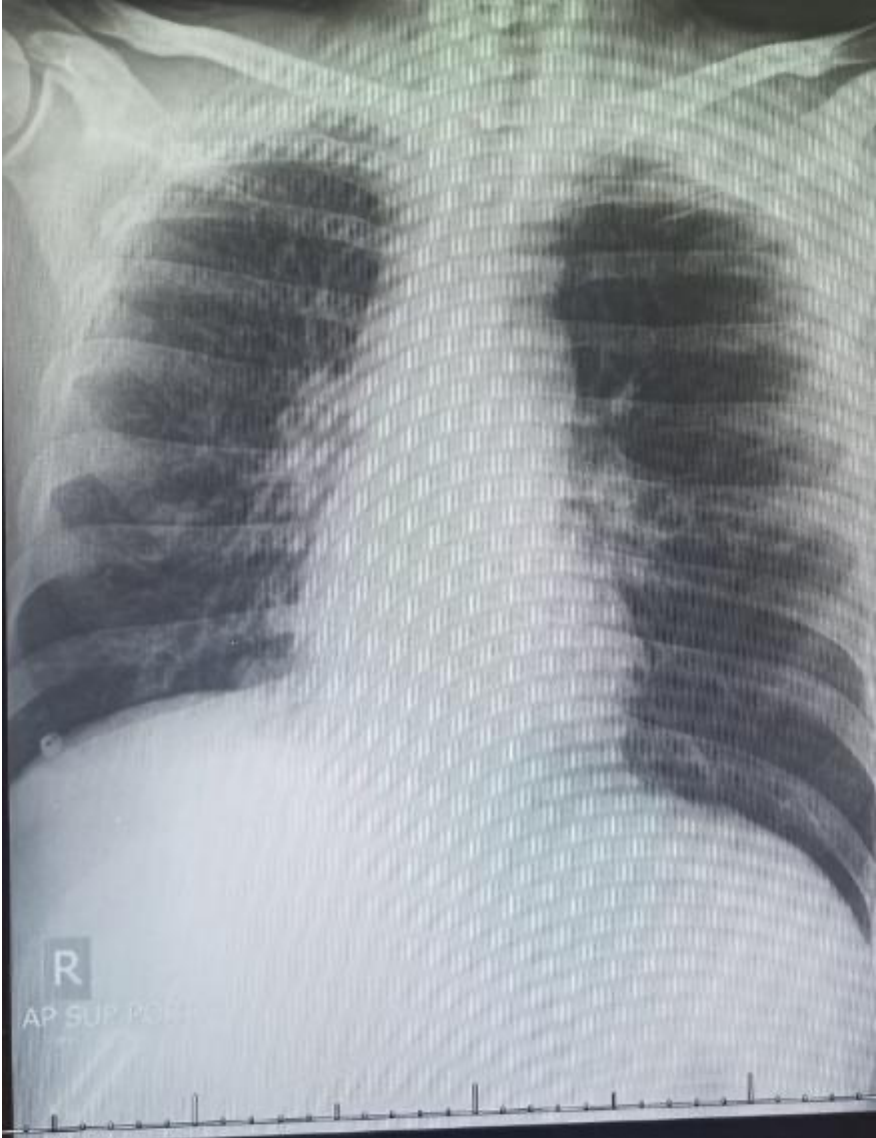


Figure 3. Chest radiography on discharge from Intensive Care Unit



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