International Journal of Medical Case Reports
2025, Vol. 4, No. 4, pp. 63-67
https://doi.org/10.11648/j.ijmcr.20250404.12

Case Report

Purpura Fulminans in an Immunocompromised Patient with Invasive Pneumococcal Disease: A Case Report

Paulo Fernandes[1, *], Ana Carina Baldino[2], Sara Carvalho[1], João Maia Oliveira[1], José Vaz[1]

[1]Intensive Care Unit, Hospital José Joaquim Fernandes, Beja, Portugal
[2]Internal Medicine Department, Hospital José Joaquim Fernandes, Beja, Portugal

Abstract

Background: Streptococcus pneumoniae is a leading cause of community-acquired pneumonia and invasive pneumococcal disease, particularly in patients with impaired immune function or asplenia. Pneumococcal infections remain associated with high morbidity and mortality, especially when complicated by septic shock and multiple organ failure. Purpura fulminans is a rare but catastrophic dermatologic and hematologic manifestation characterized by rapidly evolving purpuric skin lesions with central necrosis, reflecting underlying systemic coagulopathy. Case presentation: We report the case of a 41-year-old man with antisynthetase syndrome and a history of splenic infarction, who was receiving immunosuppressive therapy. He presented with septic shock, respiratory failure with lung infiltrates, and diffuse violaceous skin lesions. Laboratory findings revealed severe disseminated intravascular coagulation, and Streptococcus pneumoniae bacteremia was confirmed by polymerase chain reaction. Despite the immediate initiation of broad-spectrum antibiotics and intensive care admission for supportive care, his condition deteriorated rapidly, developing refractory multiorgan failure, and death occurred within 12 hours of hospital admission. Conclusions: This case highlights the fulminant and often fatal course of pneumococcal sepsis in high-risk patients. The immunosuppressive therapy and functional asplenia likely contributed to impaired bacterial clearance and overwhelming infection. While empiric antibiotic coverage was appropriate, the rapid clinical decline demonstrates the lethal synergy between severe bacterial sepsis and purpura fulminans associated consumptive coagulopathy. Preventive strategies, particularly pneumococcal vaccination, remain central to reducing the burden of invasive pneumococcal disease in immunocompromised and asplenic individuals.

Keywords

Invasive Pneumococcal Disease, Purpura Fulminans, Disseminated Intravascular Coagulation, Septic Shock

1. Introduction

Streptococcus pneumoniae is a Gram-positive extracellular pathogen that typically colonizes the upper respiratory tract mucosa. It remains a significant cause of community-acquired pneumonia (CAP) requiring hospitalization, and despite the availability of effective vaccines, it continues to be a leading cause of morbidity and mortality among adults [1]. The actual

*Corresponding author: paulosmfernandes@gmail.com (Paulo Fernandes)

Received: 1 October 2025; **Accepted:** 14 October 2025; **Published:** 30 October 2025

prevalence of *Streptococcus pneumoniae* in CAP may be underestimated, as more than half of CAP cases lack an identified pathogen. Methodological limitations contribute to this since blood cultures detect pneumococcus in only 20-25% of cases and urine antigen tests show low sensitivity (52%) in non-bacteremia disease [2]. As a result, diagnosis often relies on sputum Gram stainning and culture.

Invasive Pneumococcal Disease (IPD) is defined as isolation of *S. pneumoniae* from normally sterile sites such as blood, cerebrospinal fluid, or other bodily fluids [3]. The risk of severe infection, complications and mortality is higher in patients with bacteremia, representing a substantial global challenge with mortality rates reaching up to 60% in elderly or immunocompromised patients [1, 4, 5].

Purpura fulminans (PF) represents an uncommon but severe cutaneous manifestation of disseminated intravascular coagulation (DIC) characterized by rapidly progressive skin hemorrhage and necrosis, with limb amputation risk [3, 6].

PF can be classified into three main types: neonatal, idiopathic, and acute infectious. Neonatal PF occurs in newborns and is linked to inherited deficiencies in anticoagulants such as protein C, protein S, and antithrombin III. It typically appears very early in life, and treatment focuses on correcting these specific deficiencies. Idiopathic PF is believed to be an autoimmune response that develops approximately 7 to 10 days after an infectious disease and has been associated with the development of anti-protein S antibodies, causing a protein S deficiency and a hypercoagulable state. Acute infectious PF is the most common type and develops in patients with severe sepsis and most often associated with encapsulated bacterial microorganisms, such as *Neisseria meningitides*, *S. pneumoniae*, *Hemophilus influenzae*. The mechanism involved in acute infectious PF is based on the consumption of protein C, S and antithrombin III triggered by bacterial endotoxins, promoting a pro-coagulative state leading to thrombosis of dermal vessels [7].

The classic presentation of PF is diffuse retiform or angular purpuric lesions that rapidly develop central hemorrhagic necrosis [8].

We report a case of an immunocompromised patient with septic shock due to *Streptococcus pneumoniae* infection with multiple organ failure that presented with purpura fulminans leading to death in less than 24 hours.

2. Case Presentation

A 41-year-old man was brought to the Emergency Room (ER) with impaired consciousness and complaints of shortness of breath and generalized pain that had begun the day before. His past medical history was notable for Antisynthetase Syndrome with muscular and lung involvement (myositis and interstitial lung disease) and previous spleen infarct due to thrombosis of the splenic artery. He was on immunosuppressive therapy with Prednisolone 5mg/day and Tacrolimus 3.5mg/day, although with poor therapeutic compliance. He had history of allergic reaction (toxidermia) to Vancomycin and Piperacillin/Tazobactam.

On hospital admission he was normotensive with blood pressure of 106/80mmHg, tachycardic with 130 beats per minute, increased respiratory rate around 30 cycles per minute, febrile (38,9 ℃) and hypoglycemic, with a blood glucose of 50mg/dL. The physical examination revealed a poorly perfused skin and a diffuse reticular violaceous rash, spearing the distal parts of the lower legs (Figure 1). Heart auscultation was unremarkable, and lung auscultation showed crackling rales in the left lower lung. Neurologically he was drowsy but easily arousable, agitated but orientated, without meningeal signs or focal neurologic deficits. The abdomen was painless and without peritoneal signs. He denied cough, chest pain vomiting or diarrhea.

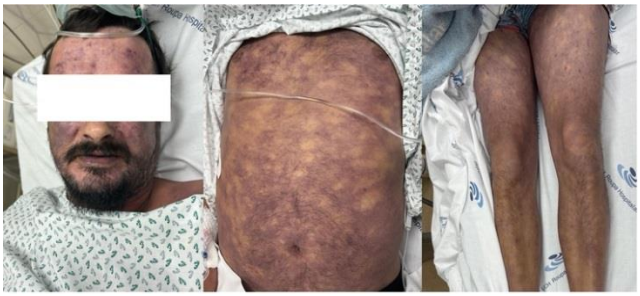

Figure 1. *Difuse reticular violaceous rash spearing the distal parts of the lower legs.*

Abnormalities in initial laboratory findings were polycythemia with hemoglobin 18.1 g/dL, white blood cell count of 17.500 /µL with neutrophil predominance (95%), low platelets of 25.000 /µL, elevated C-Reactive Protein (CRP) of 26.15 mg/dL, prolonged coagulation times with International Normalized Ratio (INR) of 3,89, activated Partial Thromboplastin Time (aPTT) of 116 seconds, low fibrinogen of 69 mg/dL with D-dimer of >30 000 ng/mL, increased creatinine of 2.19 mg/dL with urea 49 mg/dL. The arterial blood analysis showed metabolic acidemia with pH 7.27, hypocapnia with pCO_2 14 mmHg, bicarbonate of 12.7 mmol/L with hyperlactatemia of 12.9 mmol/L, and a pO_2 of 142 mmHg on 15 L/min of oxygen with a non-rebreather mask. Pneumococcal and *Legionella* urinary antigen test was negative.

Table 1. *Relevant laboratory findings on hospital admission.*

Parameters	Results	Reference range
Hemoglobin	18,1g/dL	13,2 – 16,6g/dL
Total leucocyte count	17.500/uL	3.800 – 10.000/uL
Platelets	25.000/uL	> 150.000/uL
INR	3,89	< 1,0
aPTT	116 sec	25 – 35 sec

Parameters	Results	Reference range
Fibrinogen	69mg/dL	200-400mg/dL
D-dimer	>30.000ng/mL	< 500ng/mL
Creatinine	2,19mg/dL	0,7 – 1,3mg/dL
Urea	49mg/dL	20-50mg/dL
Serum pH	7,27	7,35 – 7,45
$PaCO_2$	14mmHg	35 – 45mmHg
Bicarbonate	12,7mmol/L	22 – 28mmol/L
Lactate	12,9mmol/L	< 1,8mmol/L

Computer tomography (CT) of the chest identified small bilateral areas of interstitial lung densification with ill-defined consolidations with poorly defined limits, consistent with foci of pneumonia. No other source of infection was identified on CT of the abdomen and pelvis. Cerebral CT scan was unremarkable.

The patient was diagnosed with septic shock with probable community-acquired pneumonia as source of infection, and he was admitted to Intensive Care Unit (ICU). Crystalloid fluid resuscitation was initiated, blood cultures obtained and empiric therapy with Amoxicillin-Clavulanate, Doxycycline and Clindamycin was started. The patient developed hypotension requiring norepinephrine infusion, and invasive mechanical ventilation was initiated due to respiratory failure. He started hydrocortisone *per* protocol for septic shock as adjuvant treatment.

During the first few hours, the patient developed areas of purpuric rash with central necrosis in the abdomen, face and arms (Figure 2).

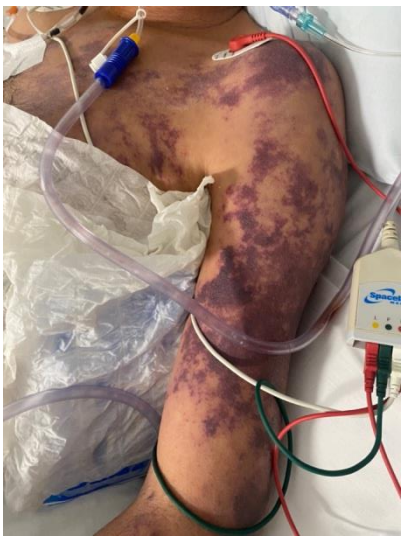

Figure 2. *Purpuric rash developed few hours after hospital admission (left upper arm).*

Despite timely and adequate treatment, the patient evolved with refractory septic shock with worsening hypotension, requiring vasopressin as a second vasopressor. Echocardiogram showed a hyperdynamic heart, without pericardial effusion or signs of hypovolemia, corroborant with a septic shock. The laboratory confirmed the molecular detection of *Streptococcus pneumonia* DNA in the blood cultures by Polymerase Chain Reaction (PCR), confirming the diagnosis of Invasive Pneumococcal Disease and adequate treatment. Despite all efforts to resuscitate the patient, he continued to deteriorate rapidly, and he died in the first 12 hours after hospital admission. *Streptococcus pneumoniae* was later confirmed in cultural examination of blood, and the antibiogram confirmed the adequate antibiotic coverage.

3. Discussion

This case underscores the fulminant nature of IPD in an immunocompromised host succumbing in less than 24 hours due to multiorgan failure with exuberant PF, despite swift and appropriate treatment. The progression exemplifies the lethal synergy between severe bacterial sepsis and the systemic thrombotic coagulopathy that characterizes PF.

Streptococcus pneumoniae is a well-known pathogen implicated in a broad spectrum of diseases, ranging from localized infections such as otitis media and sinusitis to severe systemic illnesses including meningitis, bacteremia, and pneumonia [9]. While IPD can affect immunocompetent individuals, its incidence and associated mortality are significantly increased in patients with compromised immune system, such as patients taking immunosuppressive therapy, with functional or anatomical asplenia, and individuals with chronic systemic illnesses [1]. In this case, the patient had multiple overlapping risk factors, including long-term immunosuppressive treatment for antisynthetase syndrome and previous splenic infarction, which likely contributed to impaired pneumococcal clearance.

The empiric antibiotic regimen was initially selected to provide broad coverage for severe community-acquired pneumonia in an immunocompromised patient with septic shock, while also accounting for his history of allergic reactions to vancomycin and piperacillin-tazobactam. Amoxicillin-clavulanate offers reliable activity against most community-acquired microorganisms, while doxycycline adds coverage for eventual zoonosis (Leptospirosis could explain the acute liver failure), and atypical pathogens such as *Mycoplasma pneumoniae* and *Chlamydophila pneumoniae*, which are also relevant in severe CAP when the pathogen is not yet confirmed. Clindamycin was included for its antitoxin effect, which can be beneficial in toxin-mediated severe infections and in the context of necrotizing soft tissue manifestations.

Functional asplenia is particularly relevant in pneumococcal sepsis. The spleen plays a critical role in filtering encapsulated organisms from the bloodstream. Loss of splenic function results in diminished opsonization and phagocytic

clearance, making patients susceptible to overwhelming infections by encapsulated bacteria such as *S. pneumoniae* [10]. Although the patient did not undergo splenectomy, prior splenic infarction likely rendered his spleen functionally inadequate. This anatomic predisposition, coupled with inadequate vaccine coverage (information on pneumococcal vaccination was not available), may have created the perfect storm for the development of IPD.

Purpura fulminans is a rare but catastrophic complication of severe sepsis and disseminated intravascular coagulation. In acute infectious PF, as seen in this case, bacterial endotoxins, particularly from gram-negative and encapsulated organisms, initiate this consumptive coagulopathy. Although most commonly associated with *Neisseria meningitidis*, *S. pneumoniae* is increasingly recognized as a causative agent, particularly in immunocompromised patients [11].

The diagnosis of PF is primarily clinical, supported by laboratory evidence of DIC, such as elevated D-dimers, low fibrinogen levels, thrombocytopenia, and prolonged coagulation times—all of which were profoundly abnormal in our patient. The skin changes in PF are often accompanied by multiorgan failure, serving as an early visual clue of systemic deterioration. Although a skin biopsy could confirm the diagnosis of PF, it was not performed due to the rapid deterioration of the patient [11].

Management of PF centers on addressing the underlying infection, supportive care for shock and organ dysfunction, and aggressive resuscitation. Early initiation of broad-spectrum antibiotics is critical and should not be delayed pending culture results. In this case, empirical therapy with amoxicillin-clavulanate, doxycycline, and clindamycin was appropriate, particularly in the context of known drug allergies. Molecular diagnostics confirmed *S. pneumoniae* bacteremia within hours, and susceptibility testing later corroborated the appropriateness of the antibiotic regimen. Nevertheless, the patient's clinical trajectory was already irreversibly set.

Adjunctive therapies for PF, such as replacement of natural anticoagulants (e.g., protein C concentrate or fresh frozen plasma), have shown promise in selected cases, particularly in congenital or idiopathic PF. However, in the context of septic PF with multiorgan dysfunction, their role remains controversial and is not supported by high-level evidence [12, 13]. Similarly, the use of corticosteroids in septic shock remains debated but may be justified in patients with adrenal insufficiency or as an adjuvant in catecholamine-refractory hypotension, as was attempted here.

This case also serves as a reminder of the importance of preventive strategies in at-risk populations. Immunization with pneumococcal conjugate and polysaccharide vaccines is a cornerstone in the prevention of IPD, especially in patients with known risk factors such as immunosuppression or asplenia. Current guidelines recommend either 1 dose of the 20-valent pneumococcal conjugate vaccine (PCV20) or 1 dose of the 23-valent pneumococcal polysaccharide vaccine (PPSV23) followed by either 1 dose of PCV20 or PPSV23 for such individuals [14, 15]. Whether this patient had been vaccinated is unclear, but his medical history should have prompted vaccination during outpatient follow-up.

4. Conclusions

This case highlights the rapidly fatal course and exuberant manifestations that IPD, particularly when complicated by PF, can have in immunocompromised individuals. Despite timely medical intervention, including appropriate antibiotic therapy and intensive supportive care, the patient's condition deteriorated swiftly due to overwhelming sepsis and DIC. The presence of multiple predisposing factors (including immunosuppressive therapy, underlying autoimmune disease, and functional asplenia) likely contributed to the severity of the presentation and the unfavorable outcome.

Purpura fulminans remains a rare but critical dermatologic and hematologic emergency that should prompt immediate recognition and intervention. Its characteristic skin lesions may serve as an early warning sign of underlying systemic coagulopathy and multiorgan dysfunction. Clinicians should maintain a high index of suspicion for this diagnosis in septic patients presenting with rapidly evolving purpuric skin lesions.

This case also underscores the essential role of preventive measures, especially pneumococcal vaccination, in high-risk populations. Adherence to vaccination guidelines for immunocompromised and asplenic patients remains a cornerstone in reducing the incidence and severity of invasive pneumococcal infections. Early recognition, aggressive treatment, and preventive strategies are crucial, yet in some cases, as illustrated here, the disease may still follow an inexorably fatal course. Continuous efforts to improve awareness, vaccination coverage, and early identification of complications are imperative to improving outcomes in vulnerable patient populations.

Abbreviations

aPTT	activated Partial Thromboplastin Time
CAP	Community-Acquired Pneumonia
CRP	C-Reactive Protein
CT	Computer Tomography
DIC	Disseminated Intravascular Coagulation
ER	Emergency Room
ICU	Intensive Care Unit
INR	International Normalized Ratio
IPD	Invasive Pneumococcal Disease
PCR	Polymerase Chain Reaction
PCV20	20-valent Pneumococcal Conjugate Vaccine
PF	Purpura Fulminans
PPSV23	23-valent Pneumococcal Polysaccharide Vaccine

Author Contributions

Paulo Fernandes: Conceptualization, Data curation, Formal Analysis, Methodology, Project administration, Validation, Writing – original draft, Writing – review & editing

Ana Carina Baldino: Conceptualization, Data curation, Formal Analysis, Methodology, Project administration, Validation, Writing – original draft, Writing – review & editing

Sara Carvalho: Conceptualization, Data curation, Formal Analysis, Methodology, Project administration, Validation, Writing – original draft, Writing – review & editing

João Maia Oliveira: Conceptualization, Data curation, Formal Analysis, Methodology, Project administration, Resources, Supervision, Validation, Writing – review & editing

José Vaz: Project administration, Supervision, Validation, Visualization, Writing – review & editing

Funding

This work is not supported by any external funding.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Musher, D., & Tuomanen, E. (2023). Pneumococcal pneumonia in patients requiring hospitalization. In UpToDate. https://www.uptodate.com/contents/pneumococcal-pneumonia-in-patients-requiring-hospitalization?search=pneumococcal%20pneumonia&source=search_result&selectedTitle=1~85&usage_type=default&display_rank=1

[2] Said, M. A., Johnson, H. L., Nonyane, B. A., Deloria-Knoll, M., O'Brien, K. L., AGEDD Adult Pneumococcal Burden Study Team, Andreo, F., Beovic, B., Blanco, S., Boersma, W. G., Boulware, D. R., Butler, J. C., Carratalà, J., Chang, F. Y., Charles, P. G., Diaz, A. A., Domínguez, J., Ehara, N., Endeman, H., Falcó, V.,… Watt, J. P. (2013). Estimating the burden of pneumococcal pneumonia among adults: a systematic review and meta-analysis of diagnostic techniques. PloS one, 8(4), e60273. https://doi.org/10.1371/journal.pone.0060273

[3] Djurdjevic, N., Taweesedt, P. T., Paulson, M., LaNou, A., Radovanovic, M., Patel, J. N., Veselinovic, M., McDermott, W. R., & Dumic, I. (2020). Septic Shock and Purpura Fulminans Due to Streptococcus pneumoniae Bacteremia in an Unvaccinated Immunocompetent Adult: Case Report and Review. The American journal of case reports, 21, e923266. https://doi.org/10.12659/AJCR.923266

[4] Centers for Disease Control and Prevention. (2024, February 6). Clinical features of pneumococcal disease. https://www.cdc.gov/pneumococcal/hcp/clinical-signs/index.html

[5] Kahraman, H., Yıldız, P., Yılmaz, Ş., & others. (2025). Impact of invasive and noninvasive pneumococcal diseases on adult populations: Risk factors and vaccination status. *BMC Infectious Diseases*, *25*, Article 172. https://doi.org/10.1186/s12879-025-10579-1

[6] Hamasaki, A., Yumoto, T., Fukushima, S., Hagiya, H., Chang, B., Akeda, Y., Hongo, T., Tsukahara, K., Naito, H., & Nakao, A. (2025). Purpura fulminans caused by Streptococcus pneumoniae serotype 23A in a young post-splenectomy man: A case report. Journal of infection and chemotherapy: official journal of the Japan Society of Chemotherapy, 31(10), 102791. https://doi.org/10.1016/j.jiac.2025.102791

[7] Perera, T. B., & Murphy-Lavoie, H. M. (2023, July 17). Purpura fulminans. In StatPearls. StatPearls Publishing. https://www.ncbi.nlm.nih.gov/books/NBK532865/

[8] Hale, A. J., LaSalvia, M., Kirby, J. E., Kimball, A., & Baden, R. (2016). Fatal purpura fulminans and Waterhouse-Friderichsen syndrome from fulminant Streptococcus pneumoniae sepsis in an asplenic young adult. IDCases, 6, 1–4. https://doi.org/10.1016/j.idcr.2016.08.004

[9] Kruckow, K. L., Zhao, K., Bowdish, D. M., & others. (2023). Acute organ injury and long-term sequelae of severe pneumococcal infections. Pneumonia, 15, Article 5. https://doi.org/10.1186/s41479-023-00110-y

[10] Di Sabatino, A., Carsetti, R., & Corazza, G. R. (2011). Post-splenectomy and hyposplenic states. Lancet (London, England), 378(9785), 86–97. https://doi.org/10.1016/S0140-6736(10)61493-6

[11] Chalmers, E., Cooper, P., Forman, K., Grimley, C., Khair, K., Minford, A., Morgan, M., & Mumford, A. D. (2011). Purpura fulminans: recognition, diagnosis and management. Archives of disease in childhood, 96(11), 1066–1071. https://doi.org/10.1136/adc.2010.199919

[12] Martí-Carvajal AJ, Solà I, Gluud C et al: Human recombinant protein C for severe sepsis and septic shock in adult and paediatric patients. Cochrane Database Syst Rev, 2012; 12: CD004388.

[13] de Kleijn ED, de Groot R, Hack CE et al: Activation of protein C following infusion of protein C concentrate in children with severe meningococcal sepsis and purpura fulminans: A randomized, double-blinded, placebo-controlled, dose-finding study. Crit Care Med, 2003; 31(6): 1839-47.

[14] Centers for Disease Control and Prevention. (2025, May 24). Summary of risk-based pneumococcal vaccination recommendations. U.S. Department of Health and Human Services. https://www.cdc.gov/pneumococcal/hcp/vaccine-recommendations/risk-indications.html

[15] U.S. Pharmacist. (2024, February 15). Updated recommendations for pneumococcal vaccination. Jobson Medical Information LLC. https://www.uspharmacist.com/article/updated-recommendations-for-pneumococcal-vaccination-1