https://doi.org/ 10.33472/AFJBS.6.5.2024. 6500-6505

African Journal of Biological Sciences

Purpura fulminans: A rare disease with menacing outcome

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Article History
Volume 6, Issue 5, 2024
Received: 15 May 2024
Accepted: 22 May 2024
doi: *10.33472/AFJBS.6.5.2024.*
6500-6505

Abstract:

Introduction: Purpura fulminans is a rare, life-threatening condition characterized by sudden onset of intravascular thrombosis and hemorrhagic skin necrosis, often leading to vascular collapse and disseminated intravascular coagulation. This case study highlights the rapid progression of the disease and the comprehensive treatment approach employed.

Case Presentation: A 40-year-old male presented to the emergency department with a two-day history of swelling and a rapidly progressing bluish-black rash over his body, following a recent treatment for a foot wound. The patient exhibited symptoms including fever, tachycardia, hypotension, and a high respiratory rate. Initial laboratory investigations showed elevated leukocytes, fluctuating platelet counts, and markers indicative of severe infection and coagulopathy.

Interventions: The treatment strategy included aggressive supportive care, hydration, broad-spectrum antibiotics targeting suspected pathogens, and anticoagulation therapy. Early surgical intervention for debridement of necrotic tissue was conducted to manage extensive skin damage. Additional treatments included the use of recombinant tissue plasminogen activator, plasmapheresis, and intravenous immunoglobulin [IVIg] to combat the severe inflammatory and thrombotic processes.

Outcomes: The interventions stabilized the patient's condition, mitigated the spread of necrosis, and managed the underlying infectious and coagulative complications. This case underlines the importance of prompt diagnosis and multifaceted treatment approaches in managing purpura fulminans.

Conclusion: This case study emphasizes the critical nature of early diagnosis and aggressive treatment in managing purpura fulminans. It underscores the necessity for a multidisciplinary approach to care, including early pharmacological intervention, surgical management, and vigilant monitoring of the patient's hemodynamic status and coagulation parameters.

Keywords: Purpura fulminans, disseminated intravascular coagulation, necrosis, anticoagulation, surgical debridement, case study.

Introduction:

Purpura fulminans is a life-threatening condition that manifests as a severe, rapidly progressing purpuric rash, characterized by the coagulation within the microvasculature leading to extensive skin necrosis and purpuric lesions. This condition is recognized as a critical dermatological emergency due to its rapid progression and the severe systemic symptoms it presents. Patients with purpura fulminans often experience high fevers, bleeding from multiple sites, hypotension, and signs of systemic shock [1]. As the disease progresses, it can lead to disseminated intravascular coagulation [DIC] and significant circulatory collapse, making immediate and effective management crucial to patient outcomes.

Purpura fulminans can affect individuals at any age, including neonates, children, and adults. It is classified into three distinct types based on its underlying causes:

1. **Neonatal Purpura Fulminans**: This form typically presents shortly after birth and is often linked to inherited deficiencies in natural anticoagulants such as protein C, protein S, and antithrombin III. These deficiencies contribute to the thrombotic environment conducive to the development of purpura fulminans. Management strategies focus on replacing the deficient proteins to control and prevent the progression of symptoms [2].
2. **Idiopathic or Acquired Purpura Fulminans**: Usually occurring as a sequel to a febrile, infectious illness, this form of purpura fulminans manifests approximately 7 to 10 days following the onset of the primary infection. Common preceding infections include skin-involved illnesses such as varicella or scarlet fever. The pathogenesis is believed to involve an autoimmune component, where the body's immune response to the infection results in widespread inflammation and coagulation [3,4].
3. **Acute Infectious Purpura Fulminans**: This is the most common type and occurs in the context of severe infections, particularly those caused by endotoxin-producing gram-negative bacteria, leading to a severe systemic response and sepsis. The presence of purpura fulminans in these patients is a poor prognostic indicator and often accompanies more severe manifestations like necrotizing fasciitis [5].

Despite advances in supportive care and targeted treatments that have improved survival rates and reduced complications, purpura fulminans remains a debilitating condition. Survivors often face significant morbidity, including potential major amputations due to extensive tissue necrosis. The management of purpura fulminans requires a multi-disciplinary approach, emphasizing rapid diagnosis, supportive care, and addressing the underlying cause to improve outcomes and reduce mortality rates.

The work enhances understanding of Purpura Fulminans by detailing its classification, pathogenesis, and tailored treatment approaches based on etiology. It emphasizes a multi-disciplinary, patient-specific strategy, reflecting advances in early diagnosis and management. This synthesis of current knowledge not only informs clinical practice but also highlights areas for further research, particularly in improving long-term outcomes and reducing morbidity. This targeted approach represents a significant advancement in the field, guiding future studies and innovations in treatment.

1. **Discussion**

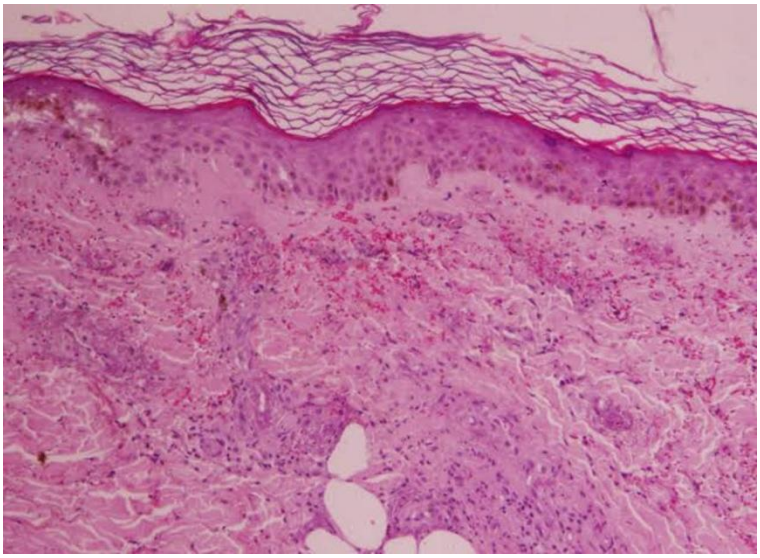
Purpura fulminans is a rare but severe condition characterized by intravascular thrombosis and hemorrhagic infarction of the skin, which rapidly progresses and often leads to vascular collapse and disseminated intravascular coagulation. The lesions initially appear as well-demarcated erythematous macules, which quickly evolve into irregular central areas of blue-black hemorrhagic necrosis [6].

The management of purpura fulminans begins with supportive care and ensuring adequate hydration due to the extensive thrombosis that can damage multiple end organs. Concurrently, identifying and addressing the underlying cause is crucial. Anticoagulation therapy may be initiated to prevent further thrombotic events. Treatment often requires the replacement of blood, clotting factors, and platelets due to the pro-coagulant state and the presence of DIC. Early surgical debridement of necrotic areas has been demonstrated to reduce mortality [7].

In cases of acute infectious purpura fulminans, treatment should include broad-spectrum antibiotics to cover key pathogens such as Neisseria meningitidis, Streptococcus, Staphylococcus, and Clostridia species. Regimens typically include carbapenems or vancomycin paired with beta-lactam/beta-lactamase inhibitor combinations. Clindamycin is frequently added for its ability to inhibit toxins that contribute to the progression of the disease [8]. Intravenous immunoglobulin [IVIg] therapy is employed to provide antibodies against these toxins. Administering activated protein C can help mitigate the inflammatory cascade and restore coagulation balance, potentially reducing the progression of skin injuries.

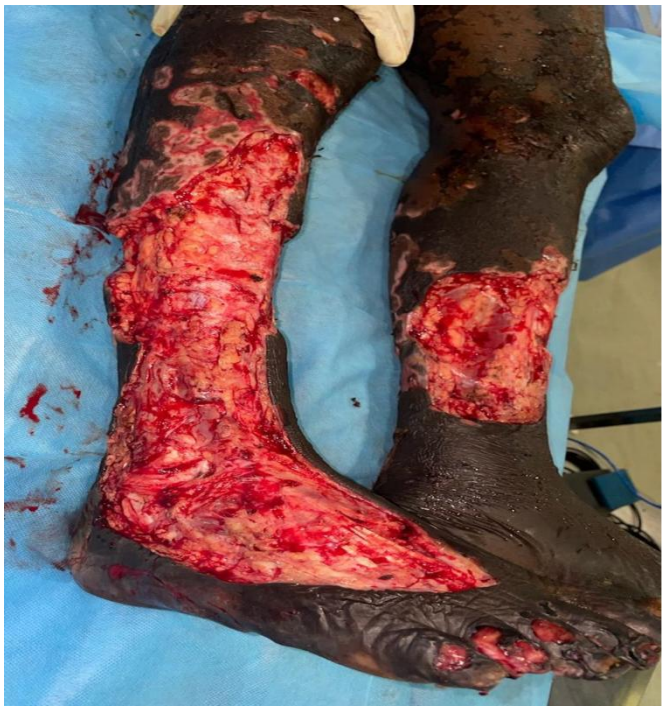
Supportive therapy forms the cornerstone of management, supplemented by high-level antibiotics, replacement of blood products, heparin, recombinant tissue plasminogen activator, and plasmapheresis. In neonatal cases, management includes hydration and platelet transfusions, assessment of protein C and S levels, followed by fresh frozen plasma transfusions [9]. Heparin and warfarin are utilized as anticoagulants, with the addition of protein C concentrate if a deficiency is identified. In acute infectious forms, the decision to initiate anticoagulation is influenced by the presence and severity of concurrent DIC [10].

Figure 1: Histopathological Examination of Purpura Fulminans



The Fig 1 depicts a histopathological slide that is likely stained with Hematoxylin and Eosin [H&E]. This type of staining is used to visualize the details of tissue structure and pathology. In the context of the provided clinical case, the slide shows changes consistent with Purpura Fulminans. At the top layer, the epidermis shows preservation of the normal stratified structure but there is a hint of alteration in the underlying dermis. The dermal layer demonstrates widespread hemorrhage, as indicated by the pink and red areas representing extravasated red blood cells. There is evidence of tissue necrosis and destruction of the normal architecture, possibly due to thrombosis in the small blood vessels, which is characteristic of Purpura Fulminans. This occlusive vascular pathology can lead to ischemia and subsequent necrosis of the affected tissue.

The sample also shows a cellular infiltrate that is likely composed of neutrophils, suggesting an acute inflammatory response. However, the specific type of cells present would require further immunohistochemical staining to accurately identify. The presence of these histological features supports the clinical diagnosis of Purpura Fulminans, illustrating the severe vascular and tissue damage that occurs in this condition.



Extensive Surgical Debridement of Lower Limbs for Necrotizing Fasciitis to Avert Progression of Sepsis

This image captures the postoperative condition of a patient's lower limbs following urgent surgical debridement to remove necrotic tissue caused by rapidly progressing necrotizing fasciitis. The procedure is a critical intervention to prevent the further spread of infection and systemic sepsis.

In conclusion, Purpura Fulminans is a critical and rapidly progressing condition characterized by severe vascular damage and skin necrosis due to intravascular thrombosis. The histopathological findings, including widespread hemorrhage and tissue necrosis, highlight the devastating impact of this disease on the dermal structures. Effective management requires a multi-faceted approach that addresses both the symptoms and the underlying causes. Early and aggressive treatments such as anticoagulation, surgical debridement, and broad-spectrum antibiotics are essential to mitigate the severe complications associated with Purpura Fulminans. Supportive care, including fluid management and replacement of blood products, remains fundamental in managing the extensive thrombosis and potential multi-organ failure. The use of advanced therapies like IVIg and activated protein C further underscores the complexity of treatment needed to counteract the rapid progression of this life-threatening condition. Overall, timely and comprehensive medical intervention is crucial to improving outcomes in patients suffering from Purpura Fulminans.

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