Acute infectious purpura fulminans caused by group A β-hemolytic *Streptococcus*: An uncommon organism

Sir,

Acute infectious purpura fulminans (AIPF) occurs most commonly in children in the setting of sepsis due to *Neisseria meningitidis*, *Streptococcus pneumoniae*, *Haemophilus influenzae*, Group B β-hemolytic *Streptococci*, *Staphylococcus aureus*, and rickettsiae.[1] Herein we describe a case of acute infectious purpura fulminans (PF) in an adult male caused by Group A β-hemolytic *Streptococci* (GABHS), an unusual organism.

A 50-year-old man, without any known comorbidities, was brought with high-grade fever, decreased urine output, shortness of breath, and dusky painful skin patches over the extremities for 3 days. He was febrile (101°F) with tachycardia (120/min), tachypnea (28/min), and hypotension (90/60 mmHg). Hemorrhagic bullae over necrotic skin were present over left arm, right thigh, and right calf (15 × 15 cm, 12 × 11 cm, and 10 × 9 cm, respectively) [Figure 1]. A rim of erythema, edema, and tenderness was present around these well-demarcated necrotic patches. Blood sugar was 110 mg/dL, urea was 128 mg/dL, creatinine 2.5 mg/dL, and sodium 145 mEq/L. Thrombocytopenia (20,000 cells/mm^3) and prolonged PT-INR were present. The level of fibrin degradation products was raised. Blood culture grew GABHS [Figure 2]. Anaerobic culture did not reveal any growth and Weil–Felix test was negative. Histopathology revealed blood vessels with intramural fibrinoid necrosis and transmural inflammation composed of lymphocytes and neutrophils [Figure 3]. Hemorrhagic necrosis of epidermis, dermis, and subcutis was seen. Protein C and protein S levels were within normal range. Based on these findings, a diagnosis of GABHS sepsis with sepsis-induced PF was made. The patient was started on vancomycin and piperacillin–tazobactam with intravenous fluids and platelet transfusion. He showed a marked improvement and the eschar eventually sloughed off to reveal underlying necrotic tissue, which was managed by surgical debridement and eventually skin grafting.

PF can occur in the neonatal period secondary to inherited/acquired deficiency of protein C or S, after an infection such as varicella or scarlet fever or during acute bacterial sepsis (acute infectious PF).[2] Bacterial toxins as well as host inflammatory factors and proteolytic enzymes damage the endothelium resulting in loss of antithrombotic molecules and upregulation of adhesion molecules leading to thrombosis of postcapillary venules in papillary dermis. Acute infectious PF differs from other forms of PF in that circulatory collapse, shock, and multiorgan failure are commonly seen, which also contribute to a higher mortality rate. Also, vasculitis, including a perivascular neutrophilic infiltrate is a unique feature of AIPF.[2] Specific antimicrobial therapy along with treatment of shock and multiorgan failure is the cornerstone of treatment. Antithrombin III, protein C concentrates, and fibrinolytic agents have been used successfully.

This case is unusual in that GABHS is rarely implicated as a cause of AIPF, and to our knowledge, there are only three previous reports of the same in English literature, all in the pediatric age group.[3-5] Acute infectious or postinfectious PF is more common in children and is reported in adults only in the setting of asplenia/other inherited or acquired immunodeficiencies where capsulated organisms such as *Pneumococcus* are usually responsible. Our patient was a healthy adult male without any underlying comorbidities.

To conclude, PF is rarely described as a cutaneous manifestation of GABHS and this report highlights the importance of maintaining a high degree of suspicion when confronted with sepsis with cutaneous necrosis.

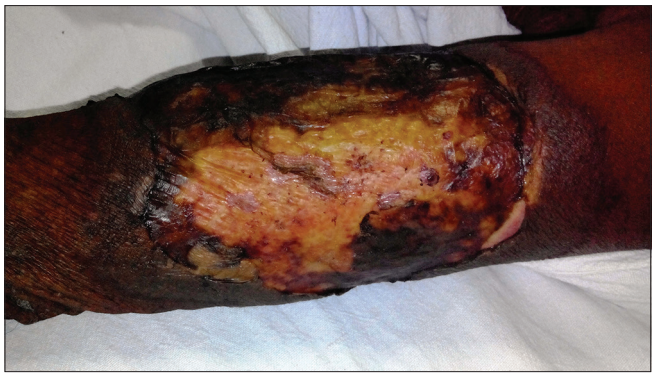

Figure 1: Hemorrhagic bullae overlying well-demarcated necrotic skin patches over the left arm

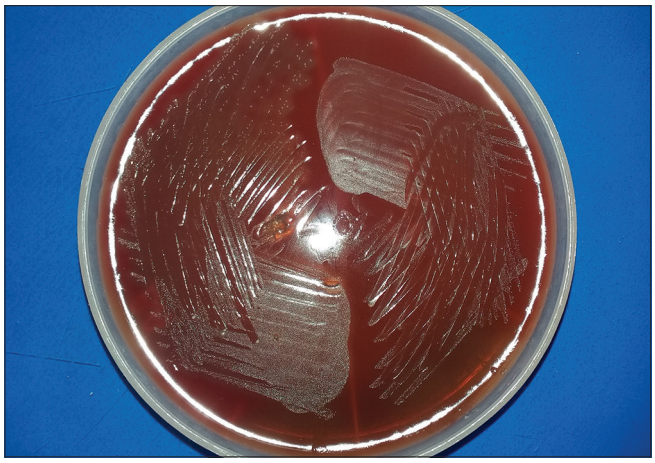

Figure 2: Blood culture showing positive growth of Group A β-hemolytic *Streptococci*

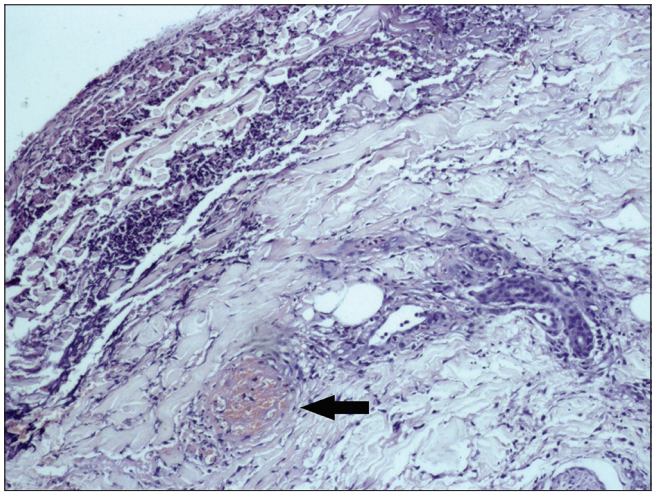

Figure 3: Hemorrhagic necrosis of skin with blood vessels showing intramural fibrinoid necrosis and transmural inflammation (hematoxylin and eosin stain, original magnification ×100)

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Conflicts of interest

There are no conflicts of interest.

Divya Gupta, Laxmisha Chandrashekar, Bheemanathi Hanuman Srinivas[1], Devinder Mohan Thappa

Departments of Dermatology and [1]Pathology, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Puducherry, India

Address for correspondence:
Dr. Laxmisha Chandrashekar,
Departments of Dermatology, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Puducherry - 605 006, India.
E-mail: laxmishac@gmail.com

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