A Case of Purpura Fulminans in an Elderly Patient

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ABSTRACT

A 72-year-old man, known case of eczema presented with blackish discoloration with multiple bullae with surrounding erythema over extremities, abdomen and buttocks associated with fever. He was diagnosed to have purpurafulminanas (PF). PF is a rare syndrome of hemorrhagic infarctions of the skin with intravascular thrombosis, seen along with disseminated intravascular coagulation and vascular collapse.

Introduction :

Purpurafulminans (PF) is a rapidly evolving, rare syndrome of hemorrhagic infarction of the skin due to intravascular thrombosis.[1,2] This is often secondary to infections or deficiency of endogenous protein-C which leads to localized or disseminated intravascular coagulation (DIC). It typically arises in children mostly due to infections. However, purpurafulmimans is uncommonly seen in adults. We report a case of purpurafulminans in elderly male possibly due secondarily infected eczema.

Case Report :

A 72-year-old man presented to us with history of fever since 2 days; blackening of hands and fluid filled lesions over both the legs since 5 days. He was a known case of eczema, had gone to a private practitioner due to its aggravation. He was given Tab Amoxicillin + Clavulanic acid orally and Fusidic cream for local application along with injectable analgesics.As there was no response to treatment patient was refered to our hospital.

On examination, patients' general condition was moderate and vitals were stable. General examination revealed bilateral non-tender inguinal lymphadenopathy and generalized edema. On cutaneous examination, there were multiple painful hemorrhagic bullae with surrounding erythema over thighs, abdomen, buttocks and back. There were multiple geographic areas of cutaneous infarction or gangrene covered with necrotic slough. Necrotic plaques were also present over scrotum and the prepuce. Oral mucosa was normal.

Figure 1 : **Skin lesions over feet**

Figure 2 : **Skin lesions over abdomen**

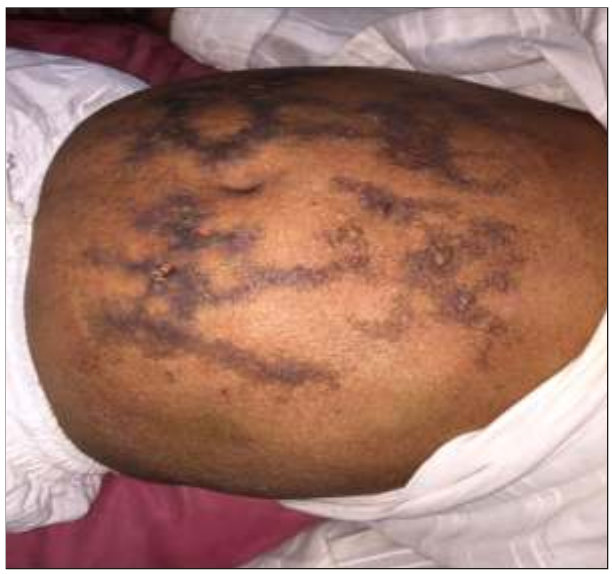

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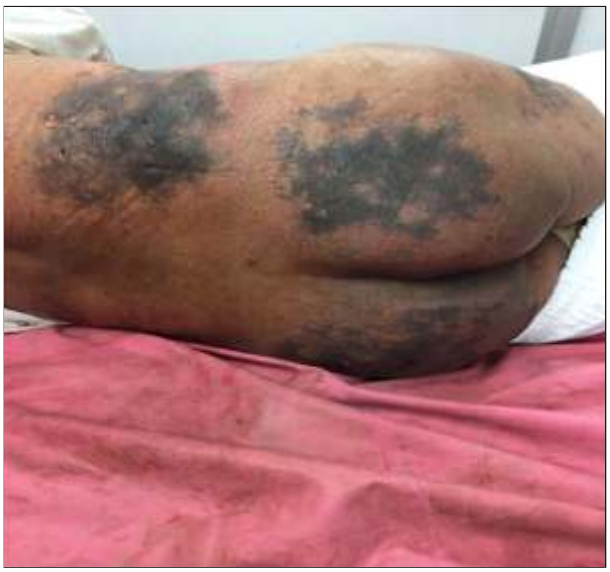
Figure 3 : **Skin lesions over hand**

Figure 4 : **Skin lesions over buttocks and back**



Investigations :

Laboratory analysis showed deranged kidney function tests (KFTs) with serum creatinine 2.38 mg/dl and blood urea - 130 mg/dl. Complete blood count showed mild leucocytosis. D-dimer was positive (1724) with fibrinogen levels of 199ng/ml. Serum albumin levels were 1.2g/dl and total proteins were 4.3g/dl. Other investigations were normal. ELISA-HIV was non-reactive. Skin biopsy showed thrombotic occlusion of dermal blood vessels with extravasation of red blood cells in the dermis and subcutaneous region. This was suggestive of PF. Based on clinical examination, history and investigations the patient was diagnosed as a case of PF.

Patient was treated with intravenous (IV) Piperacillin-tazobactum 2.25g 8 hourly for 7 days and increased to 4.5g 12 hourly for next 7 days, along with IV Metronidazole 500 mg 12 hourly for 14 days, tablet Levofloxacin 500mg alternate day for 7 doses then switched to ceftriaxone-sulbactum 1.5g i.v. 12 hourly. Pus from necrotic areas of the skin showed growth of P. aueriginosa. Patient was given enoxaparin 0.6 mg subcutaneously and oral Cilostazole 100 mg BD and topical Nitroglycerine. Due to hypoalbuminemia, patient was infused with intravenous albumin.

Supportive care was given in the form of dressing and removal of necrotic tissue wherever possible, fluid management, and nutritional support.

During the course of the disease, serial KFTs were done which showed a deteriorating trend. There was a serial decrease in platelet count, hemoglobin and total leucocyte count. Patient developed sepsis with acute kidney injury and metabolic acidosis. Metabolic acidosis was managed with infusion of sodium bicarbonates. However patient developed peripheral circulatory failure and sudden cardiac arrest and succumbed to death.

Discussion :

Exact mechanism of development of PF is not known. The Shwartzman and Arthrus reactions are supposed to cause PF.[3] PF manifests in three clinical settings : Neonatal PF in the neonatal period due to inherited, homozygous protein C or, rarely, protein S deficiency;[4] idiopathic PF occurring approximately 7 to 10 days after an infection, such as varicella or scarlet fever;[3] and acute infectious PF in conjunction with an infectious illness particularly sepsis with endotoxin-producing gram-negative bacteria.

Lesions are similar irrespective of the precipitating condition. Its cardinal manifestations are presence of circumscribed ecchymosis of skin and symmetrical gangrene of the extremities with coagulation abnormalities suggestive of DIC.

Primary lesions are tender, indurated areas of purpura surrounded by an advancing erythematous border. Late findings are formation of bullae, which signal development of hemorrhagic necrosis, and firm eschar, which ultimately sloughs.

Hematologically, there is low fibrinogen, clotting factors and platelets, with prolonged prothrombin and partial thromboplastin times (PT and PTT). Fibrinogen degradation products tend to be raised and concentrations of proteins C, S, and antithrombin III reduced.

The histopathologic hallmarks of PF are dermal vascular thrombosis and secondary hemorrhagic necrosis.[5]

Initial management of the patient with acute infectious PF must be focused on preserving life through respiratory and hemodynamic support, correction of acid-base imbalance and prompt intravenous antibiotic coverage. Circulatory collapse with tissue hypoperfusion and ischemia directly damages endothelial cells and predisposes to thrombosis.

Many therapies have been used to arrest the progression of disease. Heparin bonds with antithrombin III to inhibit thrombus formation, protein C has anticoagulant and anti-inflammatory properties. In homozygous protein C deficiency, fresh frozen plasma can give effective replacement therapy. Antithrombin III replacement can reverse DIC. Recombinant tissue plasminogen activator improves peripheral perfusion by fibrinolysis.[6] i.v.epoprostenol, a vasodilator, has been used. Plasmapheresis removes circulating endotoxin and assists in control of fluid balance. Other therapies used include topical nitroglycerine, i.v.Dextran, and leech saliva.

Conclusion :

It seems doubtful that a single therapeutic agent will be able to reverse the complex derangements that occur in PF. However, as we understand the pathophysiologic mechanisms of PF, rapidly diagnose infection and the inflammatory state of the patient, our ability to preserve life and prevent disfigurement due to acute infectious PF is likely to improve.

References :

1. J. Nolan, R. Sinclair. Review of management of purpurafulminans and two case reports. BJA. Apr 2001; 86(4): 581-6.
2. Adcock DM, Bronza J, Marlar RA. Proposed Classification and Pathologic Mechanisms of PurpuraFulminans and Skin Necrosis. Seminars in Thrombosis and Thrombosis. 1990; 16(4): 333-40.
3. Chu DZ, Blaisedell FW. PurpuraFulminans.AJS. Mar1982; 143(3): 356-62.
4. Talwar A, Kumar S, Gopal MG, Nandini AS. Spectrum of purpurafulminans : Report of three classification prototypes and review of management strategies. IJDVL. 2012; 78(2): 228-228.
5. Darmstadt GL. Acute Infectious PurpuraFulminans: Pathogenesis and Medical Management. Pediatric Dermatology. Jun 1998; 15(3): 169-83.
6. Branson HE, J. A Structured approach to the Management of PurpuraFulminans.Journal of the National Medical Association. 1983; 75(8)821-25.