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Case Report

Internal Medicine Section

Purpura Fulminans Secondary to Rickettsial Infection: A Case Report

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ABSTRACT

Purpura fulminans is an acute life threatening disorder characterized by cutaneous haemorrhagic manifestations and necrosis caused by disseminated intravascular coagulation and dermal vascular thrombosis. In this case a 60-year-old male presented with purpuric lesions over both upper and lower limbs and consumption coagulopathy following rickettsial infection. It was diagnosed as purpura fulminans secondary to rickettsial infection with disseminated intravascular coagulation and treated with replacement of platelets and coagulation factors along with antibiotics and doxycycline.

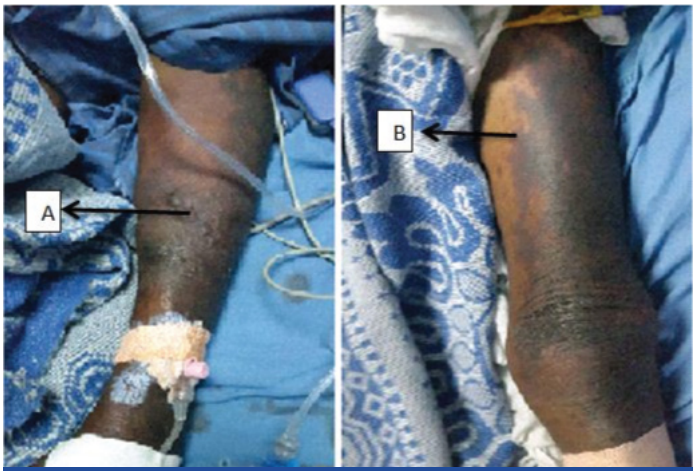
Keywords: Consumption coagulopathy, Disseminated intravascular coagulation, Thrombotic disorder

CASE REPORT

A 60-year-old male presented to the medical emergency room of Shri BM Patil Medical College Hospital in Vijayapur India, with fever of eight days duration associated with swelling of both lower limbs. On examination he had multiple purpuric patches over the upper and lower extremities. Vitals were normal with bilateral inguinal lymphadenopathy.

On the second day the patient developed acute onset breathlessness after he tried to walk to bathroom against the advice of the staff. The patient developed tachypnea, hypotension and his oxygen saturation started to drop to 90%. The patient was shifted to intensive care unit and started on inotropic and supportive treatment. After his arterial blood gases showed metabolic acidosis, correction with soda bicarbonate was done. His oxygen saturation dropped further to 80% after which non invasive ventilation was started. Pulmonary embolism was suspected and d dimer estimation was done. The d–dimer was raised with positive troponin T, echocardiography showed akinesia of the anterior septal and apical regions with distal free wall hypokinesia and an ejection fraction of 32%. The patient was started on fibrinolytic therapy with a bolus dose of 250000 units of streptokinase followed by an infusion of 50000 units every hour for 24 hours.

The patient developed altered sensorium on the fifth day of admission. His haemogram showed leukocytosis with predominance of neutrophils and thrombocytopenia. Blood culture was sent before the administration of antibiotics followed by serial cultures every 8 hours which were sterile. Dengue duo test for NS1 antigen and dengue IgM and IgG antibodies were negative. Weil Felix test showed a titer of 1:160 for OX^2. The patient was started on doxycycline and platelet infusion was given.



[Table/Fig-1a,b]: a) Blebs over the purpuric lesions over forearm
b) Purpuric skin lesions over the thigh

The patient developed blebs over the skin lesions on the seventh day with no improvement in symptoms [Table/Fig-1a,b]. The patient was started on vancomycin with continued doxycycline for the next three days. The patient showed improvement in sensorium after ten days with gradual fading of the skin lesions. His coagulation profile was normal on the 15th day after which he shifted out of intensive care. He was discharged after 20 days of admission and was keeping good health on follow up after a month.

DISCUSSION

Purpura fulminans is an acute often fatal thrombotic disorder which manifests as purpuric lesions over the body with extensive bruising and discoloration of the skin. It was first described by Guelliot in 1884 in a patient with varicella zoster [1]. Since then, it has been shown to be associated mainly with bacterial mainly meningococcal and viral infections [2]. The occurrence of antiphospholipid antibodies, disseminated intravascular coagulation and antibodies to protein C and S is thought to play a role in its pathogenesis. It is also associated with disseminated intravascular coagulation [3,4]. The three forms of this disease are classified according to the triggering mechanisms: (i) Neonatal purpura fulminans, (ii) Idiopathic purpura fulminans, and (iii) Acute infectious purpura fulminans. In inflammatory conditions it is the activation of complement and coagulation pathways by the endotoxins and signaling by inflammatory cytokines or endothelial dysfunction and vasculitis caused by the offending organisms itself [5].

Purpura fulminans presents with circumscribed ecchymotic lesions and symmetrical gangrene of the extremities with consumption coagulopathy regardless of the causative agent [6].

The lesion presents with an indurated erythematous halo with non blanchable central purpura [7]. When the skin undergoes haemorrhagic necrosis there is formation of bullae. The two most common causes are meningococcal and varicella. Gram-negative bacilli and staphylococci have also been reported. Leptospira, rickettsia and other viral conditions are known to rarely cause purpura fulminans [5]. In our case the patient tested positive for rickettsia with high titers for OX^2 antibodies. It is rare for rickettsial fever to present with purpura fulminans and the petechial lesions typically seen may be masked or totally absent [8,9].

CONCLUSION

Purpura fulminans can be caused by any microbial infection. It is hence important to keep a broad range of differential diagnosis and in cases where preceding history of fever is not clear causes other than bacterial should be sought and investigated.

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