

Case Reports

Purpura fulminans secondary to rickettsia in a child: A case report

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Purpura fulminans (PF) is a life-threatening hematologic emergency in which there is skin necrosis and disseminated intravascular coagulation (DIC). It was first described by Guelliot in 1884. There is a tissue necrosis, small vessel thrombosis, and DIC. PF may be categorized as: PF due to inherited abnormalities of protein C or other coagulation systems, acute infectious PF, and idiopathic PF¹. There have been many case reports both in adults as well as in pediatric patients with life-threatening fatal outcomes. Here, we report a case of PF in a child secondary to rickettsial infection, who was managed efficiently with timely intervention.

Case report

Eighteen month old female child presented to our outpatient department with 3-days history of low grade intermittent fever, and two episodes of non-bilious, blood stained, projectile vomiting. At the time of admission she developed generalized tonic clonic convulsions which lasted for 2 min followed by post-ictal drowsiness. On examination, the glasgow coma scale was 5/15 and she was in septic shock. Petechial and ecchymotic rashes were seen on the limbs and trunk. There was hepatosplenomegaly. Over the following days, the child developed mottled skin pigmentation and diffused purpuric rashes. She developed symmetrical patchy skin necrosis on both upper and lower limbs and also gangrene of tips of three fingers, tip of the nose and helical borders of both ears. (Figs. 1–4).

On investigation, complete blood count was unremarkable initially, except for hemoglobin 6 g%. But on the following days her platelet count decreased to 22,000/mm³. Widal test, blood and urine culture, X-ray chest and peripheral smear were done to find out the cause of fever and were found negative. Culture of the skin swab showed no growth. So, Weil-Felix (WF) test was done and was found positive for OX19 with a titre of >1 : 160 titre. The routine urine test, the liver function tests, the renal function tests and the serum electrolytes were normal except for hyperkalemia of 7 meq/l. Acid blood gas analysis was



Fig. 1: Necrosis of skin over lower limbs (before) and hypertrophic scars after healing.



Fig. 2: Necrosis of tip of nose and fingers and follow up photo showing autoamputation.



Fig. 3: Necrosis of tip of nose—Before and after treatment.



Fig. 4: Necrosis of helical borders of both ears.

normal. Based on the above clinical findings and investigations, a diagnosis of acute infectious type of PF secondary to rickettsia was made. Due to financial constraints, confirmatory tests for rickettsia and its speciation, and other investigations for the hypercoagulable states, like proteins C and S, and fibrinogen degradation products were not done.

The child was managed with immediate heparinization, and infusion of fresh frozen plasma (15 ml/kg every 12 h) along with the resuscitation measures for septic shock. Multiple units of platelets were transfused. Initially was started on crystalline penicillin, and dexamethasone injections. Later on she was put on azithromycin and meropenem. She was also given low molecular weight dextran intravenously.

Over next two weeks, her general condition improved, blood parameters returned to normal, skin lesions healed with hypertrophic scars and there was autoamputation of the gangrenous parts of the digits, tip of the nose and helical borders of both ears.

DISCUSSION

Neonatal PF is the most common form which is due to inherited or acquired abnormalities of the proteins C and S anticoagulant pathways. They usually manifest within first 72 h of birth. Idiopathic PF follows a bacterial or viral illness² and usually begins 7–10 days after the onset of the infection. The pathogenesis involves acute transient decreases in proteins C and S, or antithrombin III levels.

The most common form, acute infectious PF, occurs superimposed on a bacterial infection. The bacterial en-

dotoxins consume antithrombin III as well as proteins C and S, thus, disturbing the anticoagulant and procoagulant endothelial cell activity³. Meningococci⁴, group A streptococci⁵, Pneumococci⁶ and Varicella are the various bacterial and viral pathogens implicated. The present case which we have reported is the acute infectious form of PF secondary to rickettsia infection.

Irrespective of the cause of PF, its cardinal manifestations are presence of circumscribed ecchymosis of skin and symmetrical gangrene of the extremities with coagulation abnormalities suggestive of disseminated intravascular coagulation⁷. Typical hematological findings include low concentrations of fibrinogen, clotting factors and platelets, and prolonged prothrombin and partial thromboplastin times⁸. Fibrinogen degradation products tend to be raised and concentrations of proteins C, S, and antithrombin III reduced⁹.

Rickettsial diseases are some of the most covert re-emerging infections and are prevalent throughout the world. They are vector borne diseases, vectors being ticks, fleas and louse. They commonly affect the travelers to endemic areas. In India, they are reported from Maharashtra, Tamil Nadu, Karnataka, Kerala, Jammu and Kashmir, Uttarakhand, Himachal Pradesh, Rajasthan, Assam and West Bengal¹⁰. These infections present with vague signs and symptoms mimicking viral infections, thus making diagnosis difficult.

Various serological tests that can be done to diagnose rickettsial infections are microimmunofluorescence, immunoperoxidase assay, latex agglutination, indirect hemagglutination, enzyme-linked immunosorbent assay, dot blot immunoassay (including dipstick test) and Weil-Felix test.

Of these, only WF test is easily available in India. As all these tests detect antibodies, they would be able to make diagnosis only after 5–7 days of onset of disease¹⁰. The sharing of antigens between rickettsia and proteus is the basis of this heterophile antibody test. It demonstrates agglutinins to *Proteus vulgaris* strain OX 19, OX 2 and OX K. The poor sensitivity of the WF test is now well demonstrated but a good correlation between the results of the WF test and detection of IgM antibodies by an indirect immunofluorescence assay (IFA) is often observed¹¹. This can be used as a screening test, when positive, is reasonably specific. Thus, this test can replace the more definitive investigations to diagnose rickettsial infections. In our patient, WF test was positive to OX19 antigen, and since other causes of fever were ruled out, and based on clinical signs and symptoms, we made a diagnosis of acute infectious PF secondary to rickettsia.

Irrespective of the type and etiology, management

includes supportive therapy with aggressive broad spectrum antibiotics, resuscitation with fluids and rectification of acid base and electrolyte imbalance. Fresh frozen plasma must be given 10–20 ml/kg every 8–12 h in order to replace the procoagulant and anticoagulant proteins consumed in the disease process. Platelets and cryoprecipitates must be transfused to manage thrombocytopenia and hypofibrinogenemia respectively. Anti-coagulation with heparin is important to manage large vessel venous thrombosis. Prompt excision of necrotic tissue is recommended and escharotomies and amputations may be indicated¹². Multidisciplinary involvement may be required to facilitate rehabilitation.

Correct diagnosis, timely intervention and appropriate management are important in PF in order to prevent morbidity and mortality associated with this condition.

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