## **Photoclinic**



Figure 1. Blackish discoloration over nose and upper limb.

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A 38-year-old male presented with a history of fever and chills for the past eight weeks that was complicated by decreased urinary output, discoloration of the extremities, and during the previous two weeks, swelling of the face and extremities. The patient was taking paracetamol for fever; he was not taking any other medications. The severity of fever was not documented. The discoloration of his lower limbs was preceded by a rash, which then led to the development of a blackish discoloration of the extremities and nose. There was no significant travel or past history. His general physical

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E-mail: drsourabh79@gmail.com Accepted for publication: 6 March 2011 examination was unremarkable except for a fever of 102°F and skin discoloration (Figure 1). His laboratory investigations were significant for lower hemoglobin (12.4 mg/dL), raised leucocyte count (15000 cells/cm³), raised serum creatinine (2.5 mg/dL) and urea (154 mg/dL). Blood cultures were positive for *Streptococcus pyogenes*. The diagnosis of streptococcal septicemia-related purpura fulminans (PF) was established.

What is your diagnosis? See the next page for diagnosis.

## **Photoclinic Diagnosis:**

## **Streptococcal Purpura Fulminans**

Purpura fulminans (PF) is a hemorrhagic condition usually associated with a previous infection or sepsis. Features include hypotension, small vessel thrombosis, tissue necrosis, cutaneous hemorrhage, and disseminated intravascular coagulation. PF can be broadly classified into the following three distinct categories<sup>1</sup>: 1) inherited or acquired abnormalities of protein C or other coagulation systems, 2) acute infectious PF, and 3) idiopathic PF.

Inherited and acquired abnormalities of the protein C and protein S anticoagulant pathway are responsible for the majority of cases of PF. Gram-negative organisms are considered to be the most common cause of the acute infectious type, which is often associated with multi-organ failure. The idiopathic variety is uncommon and in contrast to the acute infectious type, where microthrombi mainly occur in the skin's blood vessels rather than vessels of other organs. The most common acute infection with which PF is associated is meningococcal sepsis. The general picture of PF is of widespread microvascular thrombosis. The thrombi consist of fibrin with a mild inflammatory component, which differentiates it from other vasculitic skin disorders such as Henoch-Schönlein purpura and drug-induced purpura, where the inflammatory component is more marked.1 The mortality rate has decreased with the better treatment of secondary infections, supportive care, and new treatments. Supportive treatment and the replacement of deficient blood components, fresh frozen plasma, clotting factors, heparin, and activated protein C may be useful.2

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