Clue on the skin in Acute meningococcemia

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Abstract

A 49-year-old woman presented with diffuse retiform purpura for two days. Skin examination showed an irregular purpuric plaques and central necrosis which is characteristic feature of purpura fulminans. Purpura fulminans was defiined as a cutaneous marker of disseminated intravascular coagulation. She also had a fever with worsening myalgia and then became to septic shock rapidly. In this patient, the purpuric lesions can lead to the diagnosis of acute meningococcemia that is confirm by the bacteriologic isolation of Neisseria meningitidis from blood cultures. Although the patient had no meningitis, here we reported the presenting of purpura fulminans which is the clue to diagnosis in this case. It is vital to detect characteristic skin lesions of acute meningococcemia for early administration of appropriate antibiotics to reduce mortality.

Key Words : Purpura fulminans, Acute meningococcemia

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Introduction

Meningococcal infection, although rare, has severe morbidity and high mortality.[1] It may present as acute meningococcemia with or without meningitis, chronic meningococcemia, or meningitis alone. When acute meningococcemia develops, a classic petechial eruption is present in approximately 60% of patients, in association with fever, chills, myalgia, and headache. In severe cases, retiform purpura and skin necrosis may follow. These skin findings can be helpful to consider a meningococcal infection in the setting of acquired purpura fulminans with sepsis.

Case report

A 49-year-old woman who was known underlying type 2 diabetes mellitus, hypertension and dyslipidemia presented at the outpatient department with diffuse retiform purpura for two days. She also had a fever with severe myalgia. The patient denied headache. On arrival, her temperature was 36°C, her blood pressure was 107/64 mmHg and her pulse rate was 113 beats/min. Physical examination showed normal consciousness and no clinical evidence of meningeal irritation. Skin examination showed generalized irregular purpuric plaques with central gunmetal gray color on the trunk and all extremities (Figure 1).

Investigation revealed white blood cells count of 8,900/uL with neutrophilia (95%), raised blood sugar of 429 mg/dL with normal blood ketone of 0.2 mmol/L, decreased serum bicarbonate of 11 mmol/L, raised serum lactate of 7.4 mmol/L, and highly raised creatine phosphokinase of 2.248 U/L. Coagulation profiles were consistent with disseminated intravascular coagulopathy (DIC).

The patient was subsequently transferred to intensive care unit (ICU) and initially treated with ceftriaxone. The patient was gradually worsened. She had high grade fever, worsening myalgia and progression to fulminant septicemia. Normal saline and vasopressors were administered. Severe metabolic acidosis was treated with 7.5% Sodium bicarbonate. As a result of multi-organ failure, finally the patient was death. Hemocultures revealed gram negative diplococci which are consistent with Neisseria meningitidis, and confirmed the diagnosis of acute meningococcemia.

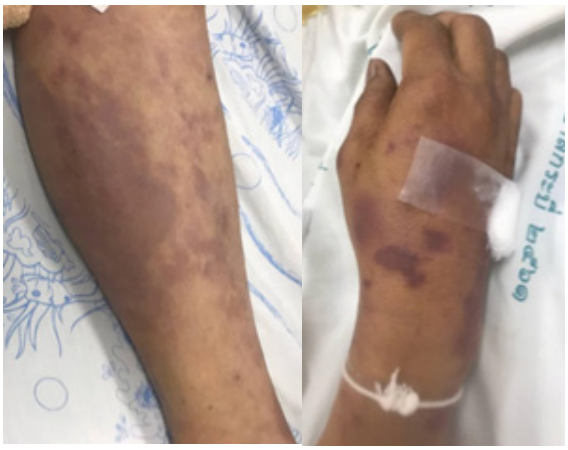

Figure 1 Purpura fulminans. Extensive purpuric plaques with central necrosis on the extremities. Note the irregular outline and a smudged appearance.

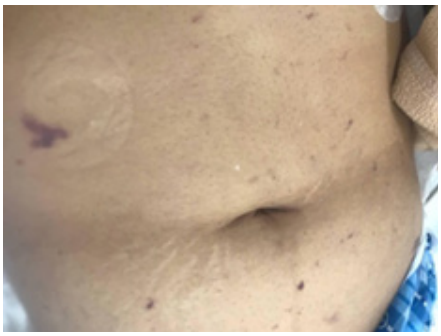

Figure 2 Acute meningococcemia. Retiform purpura in a patient with disseminated intravascular coagulation.

Discussion

Our patient presented with diffuse retiform purpura and central necrosis on the trunk and all extremities. She also had a fever with severe myalgia. The differential diagnosis includes acute meningococcemia, enteroviral infections, sepsis caused by another bacterial organism, acute bacterial endocarditis, Toxic shock syndrome, Rocky Mountain spotted fever and leptospirosis.[2] In this patient, the bacteriologic isolation from blood cultures showed *N. meningitidis* which can established the diagnosis of acute meningococcemia. A diagnosis requires the correlation between comprehensive clinical evaluations and culture of *N. meningitidis* from the skin or body fluid such as blood and cerebrospinal fluid.

Retiform purpura can be the first sign in a septic patient with DIC (Figure 2). The widespread purpura is also called purpura fulminans that is defined as rapidly progressive cutaneous hemorrhage and necrosis due to dermal vascular thrombosis and DIC.[3] The most common type is acute infectious purpura fulminans that commonly caused by *N. meningitidis, S. aureus,* group A Streptococcus, *S. pneumoniae, and V. vulnificus.*[4] Bacterial endotoxin triggers consumption of protein C lead to an acquired protein C deficiency which is thought to play a role in the pathogenesis of purpura fulminans.[5] In addition, purpura fulminans is a crucial predictor of poor prognosis following meningococcal infection.[3]

Meningococcal infection is caused by a Gram-negative diplococcus, *Neisseria meningitidis.* The human nasopharynx is the natural habitat of *N. meningitidis.* The organism is transmitted by respiratory droplets, with an incubation period of 2 to 10 days. The containment of infection is associated by undetermined host, environmental factors and host immunity.[6,7] Acute meningococcemia usually starts with a fever, myalgia, lethargy, and is responsible for septicemia and/or meningitis when the bacteria invade the bloodstream. The skin lesions begin early as a petechial rash, which may develop to retiform purpura with central necrosis or bullae.[8] The petechial eruption most commonly arises in the extremities but can occur in other parts of body.

The hallmark of therapy for acute meningococcemia is early initiation of antibiotics.[9] The recommended treatment is a third-generation cephalosporin such as ceftriaxone or cefotaxime. If the organism is susceptible, penicillin G may be used. Chloramphenicol is an alternative in current treatment option. Standard antibiotic duration is 7 days.[7] In small studies, the improved survival has been reported after administration of protein C concentrate in patient with purpura fulminans.[10]

Purpura fulminans is an important clue to diagnosis of acute meningococcemia. Severe myalgia is also crucial manifestation that must be considered. Early detection and management are necessary to improve outcome of the disease.

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