Purpura Fulminans

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Purpura fulminans may be seen in three different clinical settings: (1) in the neonatal period from protein C and S deficiencies, (2) during severe bacterial infections such as "sepsis-associated" purpura fulminans, and (3) during the convalescence of an otherwise benign "preparatory" infectious disease most commonly involving the skin. We report a case of a 20-month-old male child with purpura fulminans as a presenting sign of disseminated intravascular coagulopathy (DIC). He had suffered from fever of unknown origin for a month. Although purpura fulminans is not a common disorder to dermatologists, the awareness of this disorder may be the clue to diagnose and treat underlying diseases. (Ann Dermatol 11(2) 75–77, 1999).

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Purpura fulminans is a heterogenous group of disorders characterized by rapidly progressive purpuric lesions, which may develop into extensive areas of skin necrosis and peripheral gangrene. It may be seen in three different clinical settings; (1) in the neonatal period from protein C and S deficiencies, (2) during severe bacterial infections as "sepsis-associated" purpura fulminans, and (3) during the convalescence of an otherwise benign "preparatory" infectious disease most commonly involving the skin. Recently this term has been applied to causes of rapidly progressive hemorrhagic necrosis of the skin with hematologic features of disseminated intravascular coagulopathy (DIC).

We report a case of a 20-month-old male child with purpura fulminans as a presenting sign of DIC was referred from the department of pediatrics. A medical history revealed that he had had suffered from a high fever for 1 month. Even though he had been treated with some antibiotics and analgesics in several clinics, the fever was not controlled. The patient also had a treatment history of acupuncture on both hands and feet for the purpose of lowering the fever one week before admission. A skin examination showed the characteristic findings of purpura fulminans, which showed progressive purpuric lesions on the acral areas accompanied by a necrotic area on the third toe of the right foot (Fig. 1). Also, mottled ecchymotic patches were seen on both lower legs. The skin lesions had developed three days before admission to our hospital. A histopathological examination from the peripheral area of the purpuric patch and necrotic area of the third toe revealed non-inflammatory purpura with thrombi occluding the blood vessels and necrotic epidermis (Fig. 2).

He was treated empirically. Two days after admission, he suddenly fell into a state of shock and laboratory studies showed the characteristic findings of DIC: decreased PT/aPTT (33%/44%); increased fibrinogen degradation product (FDP) (160 µg/dL); increased D-dimer (4 mg/L); decreased fibrinogen (80 mg/dL); decreased antithrombin III (47%). The level of protein C and S were not checked. The bacterial cultures of blood, urine,
cerebral spinal fluid (CSF), sputum, and skin were all negative. His general condition rapidly deteriorated despite treatment with antibiotics and he died 5 days after the onset of shock.

**DISCUSSION**

Purpura may result from abnormalities in any of the three components of hemostasis: platelets, plasma coagulation factors and blood vessels. Platelet disorders manifest themselves as petechiae, coagulation disorders such as echymoses, and vasculitis as palpable purpura.

In classic purpura fulminans the most common antecedent diseases include varicella, streptococcal infections, and febrile exanthems. Also, the etiology is unknown. Purpuric lesions are often symmetric on the lower half of the body. Extensive and confluent purpuric and ecchymotic areas that are frequently associated with bulla formation and full thickness skin necrosis were noted on one study. Sepsis-associated purpura fulminans occurs in the context of severe acute infections with hypotension and septic shock. The organism most commonly involved is Neisseria meningitidis. Bacteria are usually absent from smears of the lesions. Skin lesions are often more acrally located than in the classic purpura fulminans and tend to progress in an ascending fashion as in our case. Mucous membranes are rarely involved. Histopathological examinations revealed occlusion of dermal capillaries and venules by thrombi with hemorrhage and varying degrees of cutaneous necrosis in one study. It has been hypothesized that a preceding event may lead to production of interleukin-1 and tumor necrosis factor-α by keratinocytes and endothelial cells in the pathogenesis of classic purpura fulminans. These cytokines induce procoagulant and antifibrinolytic changes in the dermal vascular endothelium, which initiate clotting with subsequent consumption of protein C and S, as well as antithrombin III. In sepsis-associated purpura fulminans, endotoxin mediates the activation of the intrinsic pathway of coagulation and stimulates the release of cytokines with subsequent shock, consumption of protein C and S, and DIC. In our case, because the boy had been ill with a fever of unknown origin for one month and he was treated with various drugs and acupuncture before documentation of the underlying disorder, it still remains to be a mystery as to whether the skin lesions were the primary focus of infection via accupuncture or some underlying disease was present. Moreover, protein C and S were not checked.

Treatment of idiopathic purpura fulminans includes replenishment of coagulation factors with fresh-frozen plasma or prothrombin concentrate in conjunction with heparin. Antibiotics to cover gram-positive cocci, meningococci, and gram-negative bacilli must be prudently chosen until the results from blood cultures are available. There are also anecdotal reports of the successful treatment with -aminocaproic acid, epoprostenol, dextran, and hyperbaric oxygen. In sepsis-associ-
ated purpura fulminans, treatment is primarily directed at the underlying infection and shock. The role of replacement therapy or heparin administration in sepsis-associated purpura fulminans is difficult to determine. The mortality rate is 20%.

In our case, if the skin lesions had been noticed earlier and pediatricians had had the insight of its association with DIC, prompt treatment of the underlying disease and DIC after the check of the level of protein C and S and search for underlying pathophysiology may have caused a different outcome.

We underrate the fact that purpura fulminans can be a presenting sign of DIC and may be associated with various conditions. A greater awareness may lead to more timely diagnosis as well as reduced morbidity and mortality of this serious disease.

REFERENCES