INDIAN J DERMATOL VENEREOL LEPROL 1988 ; 54 : 30-32

CASE REPORTS

PURPURA FULMINANS

D A Parikh, R J Fernandez, S J Bhatia, D R Karand and F D Dastur

Four patients, 2 males and 2 females, 16 to 70 years in age, developed purpura fulminans associated with disseminated intravascular coagulation. Three of these had meningitis, and one had septicaemia. All the four patients had extensive confluent geometric shaped purpura of explosive onset, that was associated with haemorrhagic bullae and focal gangrene. Acral cyanosis/gangrene was seen in all the patients. However, the large arterial pulses of the involved extremities were normal. A histopathological study showed fibrin thrombi in the dermal vessel. This facilitated an early diagnosis of DIC, where initial coagulation tests were unhelpful. In spite of appropriate therapy, 3 of the patients died of DIC.

Key words : Purpura fulminans, Disseminated intravascular coagulation.

Purpura fulminans (PF) is an uncommon, acute, severe and usually fatal disease. Cuielliot first described it in 1884.[1] Since then nearly 200 cases have been recorded.[1] We report 4 cases of PF with disseminated intravascular coagulation (DIC).

Case Reports

Case 1

A 70-year-old female patient was admitted to the hospital for fever and skin rash. At the time of admission, the other vital parameters were normal. There was no bleeding from any site. Skin examination revealed multiple petechiae and large ecchymotic patches predominantly on the extremities. Vesicles and bullae were present. Nikolsky's sign was positive. The fingers of the right hand showed cyanosis. She also had signs of meningitis, and CSF cytochemistry was suggestive of pyogenic meningitis. The patient was treated with crystalline penicillin, but on the 3rd day, the patient started bleeding profusely from the gastro-intestinal tract. Bleeding, clotting and prothrombin time were raised, while platelet count was reduced to 60,000/c mm. The patient died on the same day.

Case 2

A 35-year-old male having squamous cell carcinoma of the rectum was operated for colostomy. On the 2nd day after surgery, the patient developed signs of peritonitis and septicemia. On the 3rd day he developed widespread petechiae and ecchymotic lesions. All the fingers showed dry gangrene. Routine laboratory investigations were normal. Urine, stools and blood culture, done thrice showed growth of *E. coli*. The coagulation profile was within normal limits. After a skin biopsy the patient was immediately heparinised. Heparin was given intravenously in an initial dose of 100 units/Kg, and thereafter 10 units/Kg 4 hourly. Fresh blood transfusions were also given. Antibiotics in the form of gentamicin 80 mg intramuscularly thrice daily and ampicillin 500 mg, 4 times a day were started. Skin lesions on several areas showed necrosis, which subsequently sloughed to heal with scarring. The gangrenous fingers got autoamputated.

From the Departments of Dermatology, Venereology and Leprology and Medicine, Seth GS Medical College and KEM Hospital, Parel, Bombay-400 012, India.

Address correspondence to : Dr. D. A. Parikh, 1, MILAN Dixit Road, Ville-parle (East), Bombay-400 057.

Cases 3 and 4

Two adult male patients were admitted for fever and extensive skin lesions. Both had signs of meningitis. The skin examination revealed widespread, confluent and geometric-shaped purpura with acral cyanosis. Coagulation profiles of these patients were suggestive of DIC. Thrombin time was prolonged in both the cases. Platelet count was 60,000/cmm in case 3, and 40,000/cmm in case 4. CSF cytochemistry was suggestive of pyogenic meningitis. Both these patients died within 24 hours of admission due to extensive bleeding from the GI tract, leading to hypovolumic shock and sepsis.

The skin biopsy from the purpuric lesion done in all the patients, showed keratinocyte degeneration with a sub-epidermal cleft. The papillary vessels showed dilatation with extravasation of red blood cells. Deep dermal blood vessels showed a fibrinous plug. There was complete absence of an inflammatory infiltrate at the dermo-epidermal junction, and around the blood vessel. There was no sign of vasculitis.

Comments

The syndrome of septic shock, purpura, and/or acral cyanosis is highly suggestive of DIC. Skin lesions are frequently the first sign of DIC. All our four patients presented with widespread skin lesions. Purpura fulminans (PF) is common in the paediatric age group,[2] but all our patients were adults. Stanley et al[3] observed PF in patients above the age of 11. Three patients in the present study had meningitis, while one had Gram negative septicemia. Spicer and Rau[4] reviewing 100 cases of PF, observed scarlet fever as the commonest causative agent (27%), followed by post-infective (23%) (These were unverified upper respiratory tract viral infections), varicella (20%) and streptococcal infection in 11% of the cases. Purpura fulminans has also been reported with leukemia,[3] prostatic carcinoma and lymphoma.[5]

Spicer and Rau[4] described three cardinal features of PF viz (1) Large purpuric and ecchymotic areas on the skin, (2) fever, (3) hypotension.

Histopathologically a fibrinous plug in the deep dermal vessel, along with absence of inflammatory infiltrate or signs of vasculitis are suggestive of PF. A skin biopsy helps in early diagnosis.[6] Infarcts of the entire thickness of the skin are extremely rare. This is due to a rich and interconnected cutaneous blood supply.

The treatment of DIC consists of vigorous treatment of the primary cause. The role of heparin in the management of DIC is controversial.[7] Thrombosis involving small or large vessels is an indication for heparin therapy along with blood products. Purpura fulminans is due to thrombosis in the small cutaneous vessels. Thus, heparin should be useful.[7] In the present report, the case that was heparinized early, was the only one to survive. The replacement of other coagulation factors is a must.[8] Waddell et al[9] treated one case of purpura fulminans with hyperbaric oxygen, this induces hyperoxia and leads to a marked improvement in the appearance of the involved extremity. According to Spicer and Rau[4] PF is a type of Schwartzman reaction. They do not advocate corticosteroids as it may potentiate the disease process.

References

1. Esterly NB : Purpura fulminans, in : Clinical Dermatology, Vol 2, 14th ed, Editors, Demis DJ, Dahl MV, Smith EB et al : Harper and Row Publishers, Philadelphia, 1987; p 7-24.
2. Champion RH : Purpura, in : Textbook of Dermatology, Fourth ed, Editors, Rook A, Wilkinson DS, Ebling FJG et al : Blackwell Scientific Publications, London, 1987; p 1119.
3. Stanley JR, Martin CM, Colman RW et al : The skin in disseminated intravascular coagulation.

Prospective analysis of thirty-six cases. Brit J Dermatol, 1973; 88 : 221-229.

4. Spicer TE and Rau JM : Purpura fulminans, Amer J Med, 1976; 61 : 566-571.
5. Hautelecte ML, Berneman ZN, Bieger R et al : Purpura fulminans in pneumococcal sepsis, Arch Int Med, 1986; 146 : 497-499.
6. Pinkus H and Mehregan AH : A Guide to Dermatopathology. Third ed, Appleton-Century-Crofts, New York, 1981; p 181.
7. Paul O : Herparin 1986, indications and effective use Drugs, 1986; 31 : 81-92.
8. Wintrobe MM, Lee GR, Boggs DR et al : Bleeding disorder caused by vascular abnormalities, in : Clinical Haematology. Eighth ed, Editor, Wintrobe MM : Lea and Febiger Company, Philadelphia, 1981; p 1077.
9. Waddell WB, Saltzman HA, Fuson RL et al : Purpura gangrenosa treated with hyperbaric oxygenation, JAMA, 1965; 191 : 971-974.