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PURPURA FULMINANS*

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Purpura fulminans is a rare disease characterized by skin lesions which start with acute symmetrical necrosis and hemorrhagic blebs. Most of the cases are lost as a result of disseminated intravascular coagulation (DIC) which leads to shock and development of infection. The extensive tissue loss may progress to auto-amputation or surgical removal of gangrenous parts; debridement and grafting may be necessary in many survivors[1-3].

This article reviews the related literature, presents the results of treatment of 7 cases with purpura fulminans at the Hacettepe Children's Hospital and points out the need for psychotherapy as well as medical and surgical treatment.

Cases

All cases were brought to the Hacettepe Children's Hospital due to purple patches on their bodies appearing after an infectious disease (except Case 3). Clinical and laboratory findings on the patients, treatment and outcome are shown in Tables I-III.

Discussion

The etiology and pathogenesis of purpura fulminans are still unknown. The disease is usually defined by characteristic skin lesions, presence of DIC and histopathological occurrence of microthrombosis in the arteries and veins. These findings mimic Schwartzman's phenomenon, that can be experimentally induced in animals[4,5].

Despite the fact that it is usually observed during childhood, neonatal (Case 7), intrauterine and adult cases have also been reported[6-8]. A benign infection usually precedes the onset of the disease. However, it can also appear suddenly (Case 7).

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The onset has been described following such diseases as varicella (Cases 1, 2, 4), streptococcal infections (scarlet fever, tonsillitis, pharyngitis), measles, rubella, meningococcemia, other rashes (Case 5), upper respiratory tract infections and viral gastroenteritis (Case 6)[1,3,5,9,10].

Lesions are frequently localized on the hips and lower extremities. It is very uncommon for the lesions to spread to the face and the mucosal membrane (Case 1). There can also be visceral involvement[9].

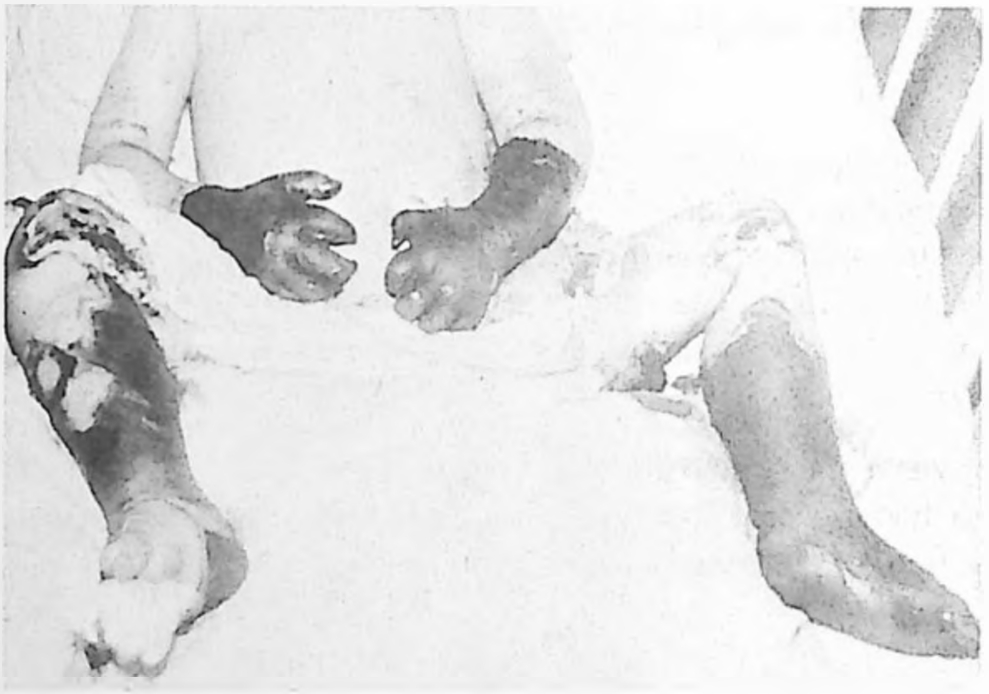

Figure 1.

In this 5-year-old boy with varicella, gangrene developed on the four extremities (Case 1).

In most reported cases DIC was often associated with purpura fulminans. There is a decrease in the platelet count, hypoprothrombinemia, hypofibrinogenemia and an increase in the fibrin degradation products. Anemia can be observed at various stages of tissue extravasation, external bleeding and hemolysis during the microangiopathic disturbances. Leukocytosis with a left shift can be observed due to tissue necrosis[5,10,11].

The hematological findings of our cases were supported by DIC as shown in Table II. Heparin is the only medicine preferred for anti-coagulation therapy. Heparin sodium (100-150 U/kg) is given intravenously every four or six hours and is adjusted to maintain a clotting time of 25-30 minutes. Response is usually obtained within 48 hours. Heparin therapy should be continued until the lesions are stabilized and the coagulation problems are eliminated (minumum 8 days, maximum 5-6 weeks). Heparin should not be discontinued too early, and it should be readministered if the lesions are reactivated[4,5,9,10,12,14].

TABLE I

Summary of the Seven Cases with Purpura Fulminans

Case	Age (years)	Sex	Preceding infection	Localization of the lesions
1	5	M	Varicella	Face, four extremities below the knee and elbow, abdomen
2	10	F	Varicella	Bilateral lower extremities to gluteal regions
3	5	M	Absent	Feet, glans penis, gluteal regions
4	5	F	Varicella	Bilateral on legs
5	4	M	Exanthematous disease (viral?)	Bilateral lower extremities, left arm
6	11/12	M	Enteritis	Both legs and abdomen
7	10/360	M	Jaundice	Bilateral lower extremities, scrotum, abdomen

Although it is reported that corticosteroids may lead to Schwartzman's phenomenon, in contrast to this it has also been observed that if they are given after heparin therapy, better results are obtained in certain cases (Case 2). Hydrocortisone, prednisolone or dexamethasone may be given[4,5,12,15]. Most of the patients go into shock due to bleeding. To prevent this, fresh plasma and blood transfusions should be administered after heparin sodium is given to the patient. In addition, to prevent the infection's spread from the necrotic areas, antibiotics are used as in our cases and others[1,4,5,9,10].

In a few cases favorable results have been obtained by using vitamin K, fibrinogen, exchange transfusion, epsilon aminocaproic acid and hyperboric oxygen[12-15].

For the differential diagnosis, toxic epidermal necrolysis, meningococcal septicemia, massive hepatitis, Reye's syndrome, uremia and various viral and bacterial diseases should be considered[2,9,10].

Survivors require amputation, debridement of the necrotic lesions, and grafting. Most important is the psychological preparation of the patients who require amputation[16], as in Cases 1 and 2.

TABLE II

Hematological Laboratory Data of the Patients with Purpura Fulminans

Patients	Hb (gm/dl)	WBC (/mm^3)	Platelets (PBS)	PT	PTT	Fibrinogen (mg/100 ml)	FDP
1	12.47	11,400	Rare	5′	5′	—	—
2	9.59	8,300	Rare	64″	194″	—	+
3	14.60	15,900	Rare	1′	65″	334	—
4	10.50	7,600	Rare	5′	5′	—	—
5	7.51	12,800	Rare	18″	118″	—	—
6	10.35	10,600	Rare	41″	189″	—	—
7	22.50	12,000	Rare	—	—	—	—

Hb : Hemoglobin
WBC : White blood cell count
PBS : Peripheral blood smear
PT : Prothrombin time (control 12″ - 14″)
PTT : Partial thromboplastin time (control 45″ - 60″)
FDP : Fibrin degradation products

TABLE III

Treatment and Outcome of the Cases

Case	Treatment	Outcome
1	Heparin sodium, dextran, fresh blood and plasma, antibiotics, psychotherapy (pre- and postoperative)	Amputation, grafts
2	Heparin sodium, cortisone, fresh blood and plasma, antibiotics, psychotherapy	Amputations, debridement, grafts
3	Heparin sodium, fresh blood, antibiotics	Debridement, grafts
4	Heparin sodium, fresh blood, vitamin K, antibiotics	Debridement
5	Heparin sodium, cortisone, fresh blood, antibiotics	Death on the 7th day in hospital
6	Heparin sodium, antibiotics	Death 18 hours after admission to hospital
7	Heparin sodium, cortisone, antibiotics	Death on the 3rd day in hospital

Summary

Clinical and laboratory findings and treatment of seven cases with purpura fulminans are reviewed. The importance of psychotherapy for the well-being of the patient, in addition to medical and surgical intervention, is mentioned.

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