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Intrauterine Purpura Fulminans*

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Purpura Fulminans in an infrequently seen syndrome and carries grave prognosis (Crawford and Riddler, 1959). To the best of our knowledge there has only been two reported cases of purpura fulminans in the newborn period[2,3] (Van Der Most, 1962 and Rainier-Pope, 1973). Therefore we would like to report a unique case of purpura fulminans, whose lesions had developed in intrauterine life.

Case Report

Baby B. (HCH: 515080) was a full term male baby who was delivered by Cesarean section since labor had not progressed, 30 hours after the membrane ruptured. At the time of delivery a lesion like ecchymoses was observed on the right inguinal region of the baby and he was given 1 mg vitamin K_1 intramuscularly.

Physical examination at birth, with the exception of the skin lesion on the right inguinal region, was within normal limits (weight, 3580 gm; height, 50 cm) and the one minute Apgar score was 9. Femoral, popliteal and dorsum of the feet pulses on both sites were palpable. Laboratory studies in the first 12 hours revealed hemoglobin to be 17.21 gm/dl, Hct. 48 %, and white blood cells were 20,400/µl. On the peripheral smear, platelets were abundant; 5 % bands, 79 % granulocytes, 2 % basophilic granulocytes, 2 % monocytes, 12 % lymphocytes and one normoblast per 100 WBC were found and the urinalysis did not show any abnormality. IgM was 19 mg/dl; skull and extremity X-rays of the baby did not disclose any abnormality. The mother's and baby's blood groups were A Rh +.

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After obtaining appropriate skin, throat and blood culltures (throat cultures grew out pyocyaneus; others were negative), the patient was given aqueous penicillin (100,000 u/kg/day), and kanamycin (10 mg kg/day) intramuscularly, because of the early membrane rupture.

Skin lesions around the right inguinal area continued to spread quickly and bullae were observed over the scrotum and right buttock in the first 24 hours (Figure 1). The baby looked sick and in pain. The next day new eccyhmotic lesions had started on the left leg and thorax. His

Figure 1
The spread of the lesions. An ecchymotic area on the right malleolar region is also seen.

Hb and Hct dropped to 8.14 gm/dl and 24 %, respectively, and he was slightly jaundiced (total bilirubin was 11 mg % and conjugated bilirubin, 1 mg %); few burr cells were seen and platelets became scanty on the peripheral smear (44,000/μl). Prothrombin time and partial thromboplastine time were found prolonged, both more than 5 minutes. The antibiotics were changed to ampicillin (300 mg/kg/day) and gentamycin (5 mg/kg/day) and prednisone (3 mg/kg/day) was started. Although the spread of the lesions was controlled, the patient became toxic and died on the 3rd day of life. Postmortem L. P. findings were within normal limits for his age. Postmortem examination did not disclose

any evidence of infection, but prominent vasculitis with fibrin deposition seen in small vessels over the involved skin area were observed.

The mother did not disclose any clinical or laboratory evidence of infection; only E. Coli was cultured from the birth canal.

Comments

Diffuse symmetrical hemorrhages with inflammatory vasculitis and necrosis of skin and subcutaneous tissue, usually involving lower extremities and buttocks are characteristics of purpura fulminans. In our case these lesions were observed at birth (Figure 1); therefore it was believed that the lesions started during intrauterine life.

The etiology of this unusual syndrome is not clear, however, it usually occurs in the convalescent phase of bacterial or viral infections. No evidence of infection was found either in the mother or in the baby. We cannot suggest that the pyocyaneus found in the infant's throat or the E. Coli in the mother's birth canal were in any way related to the baby's lesions. Normal IgM level of the baby was also incompatible with congenital infection. Therefore, some other factors, which could be operative even in intrauterine life, should also be looked for in the pathogenesis of this syndrome.

The hematologic findings of this baby on the second day of life, were compatible with disseminated intravascular coagulation, which was described previously[2] (Rainier-Pope, 1973). Heparin treatment has been recommended for the treatment of purpura fulminans[4] (Allen, 1966), but because heparinization cannot easily be controlled in a baby of this size, it was not used in this case. The baby was treated with corticosteroid as we had obtained very good results in two children with this syndrome.

Summary

A unusual case of purpura fulminans, observed in a newly born baby, is presented. Since the lesions were observed at birth, it is believed that purpura fulminans developed in the intrauterine life in this Cesarean section delivered baby.

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