

Case 8

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Clinical Abstract:

The patient was a 36-day old girl born at 38 weeks gestation to a P₂₋₀₋₀₋₂ 20-year old woman. Emergency cesarean section was indicated because of fetal distress. There was no history of maternal infections or any other medical problems during pregnancy. At birth, the Apgar scores were 2 and 3 at 1 and 5 minutes, respectively and the baby weighed 2500 grams. She was apneic and cyanotic with a heart rate of 112 per minute requiring intubation and ventilatory support. The head was small (circumference: 30 cm); there were no spontaneous movements and she had generalized edema and petechiae. All cranial sutures were closed, the palpebral fissures were small and there were no obvious cataracts. Examination of the chest revealed diffuse respiratory rales. There was abdominal distention, the liver edge was palpable 3.5 cm below the right costal margin, the spleen was palpable and the bowel sounds were absent. Examination of the extremities revealed deformed distal phalanges in both hands.

Initial laboratory data revealed blood group O positive (mother was O positive); Coombs' test was negative; red blood cell count was $4.21 \times 10^6/\text{uI}$ with micro, macro and poikilocytosis; some nucleated RBCs were present; total leukocyte count was 2500/uI (lymphocytes 78%, monocytes 4%, neutrophils 15%, bands 2%, basophil 1%); the platelet count was 137,000/uI, PT 22.7/12.7 and PTT 39/27; total bilirubin was 6.1 mg/dI with a direct fraction of 3.7 mg/dI; total protein 3.7 g/dI and SGOT 4620 IU/L. Serum titers for Toxoplasma, rubella, and herpes simplex were negative.

On the first day after birth the patient developed grand mal seizures which were controlled with phenobarbital. Although she developed a few spontaneous movements, efforts at extubation remained unsuccessful. On her twelfth day in the hospital peripheral blood cell count and a CSF sample obtained by lumbar puncture were suggestive of sepsis and meningitis. Blood cultures grew Staphylococcus epidermidis. Her hospital course was further complicated by the development of congestive heart failure and she was noted to have a patent ductus arteriosus which did not respond to pharmacologic therapy with Indomethacin. Skull x-rays revealed punctate calcifications within the brain parenchyma. The patient remained ventilator dependent and in view of the dismal neurologic prognosis, she was extubated at the parents request 36 days after birth.

The brain at autopsy weighed 80 grams.

Material submitted: one H+E section of the cerebellum and brain stem or cerebral hemisphere and brain stem.

Point for Discussion: Diagnosis