

CASE 1999 [7]

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Clinical History:

A girl was born from a 27-year old primigravida at 40 weeks of gestational age. The body weight was 3,088g and APGAR score was 8 at one minute. She presented with generalized dermal exfoliation, severe peripheral edema, respiratory insufficiency and hypotonia. She had several craniofacial anomalies including hypertelorism, high arched palate, flat nostrils, small earlobes and failure to close eyes.

She was intubated under control respiration. She presented with progressive liver failure and DIC, associated with hepato-splenomegaly. She fell into the phase of fulminant hepatitis on the 40th hospital day, subsequently followed by generalized infection of MRSA and fungi. Repeated transfusion of platelets did not improve the clinical condition.

She died of respiratory failure on the 52nd hospital day

Necropsy findings:

The body weight was 3.3 kg and the body length was 53 cm. The weight of brain, liver and spleen was 150g, 260g and 120g respectively.

Materials submitted:

1. One hematoxylin and eosin stained section of the frontal lobe
2. One Kodachrome slide of a ultramicrograph of a swollen cell in the spleen (black bar = 3.6 μ m)

Points for discussion:

1. Diagnosis
2. Mechanism of neuronal cell death