

Image and Diagnosis

Contributed by V Ertekin¹, MA Selimođlu¹, N Gürsan²,
CF Öztürk¹

Clinical Data

A two month-old boy was admitted to the paediatric department with the complaint of bleeding from the mouth. It was stated that the patient fell from the crib and uncontrolled bleeding of his oral mucosa started just before admission. This was the sixth child of the family, and the other five children were healthy. There was no consanguinity between the parents. The length of gestation was 40 weeks. His natal and postnatal history was normal. His birth-weight and height were not known because he was born at home. The infant had received no vitamin K prophylaxis and was exclusively breastfed.

On physical examination his weight, height and head circumference were between 50–75 percentiles. His skin and sclera were jaundiced and pale. Laceration in the oral mucosa was detected due to the mentioned trauma. His liver was 6 cm below the costal margin and spleen was not palpable.

Jaundice appeared at 15 days of age. He had central facial paralysis and his eyes deviated to the left. Laboratory studies on admission revealed the following results: White blood cell count 10×10^9 , haematocrit 19%, haemoglobin 6.5 g/dl, platelet count 505 00/L, aspartate aminotransferase 359 IU/l, alanine aminotransferase 189 IU/l, total bilirubin 12 mg/dl, direct bilirubin 5.9 mg/dl, alkaline phosphatase 2260 IU/l, and the PT, APT, INR were 23, 105, 2.08 respectively. After intravenous administration of 5 mg vitamin K, PT and INR normalized and the bleeding stopped. However, liver dysfunction persisted. Brain CT and liver biopsy are shown in figures 1 and 2, respectively.

What is the diagnosis?

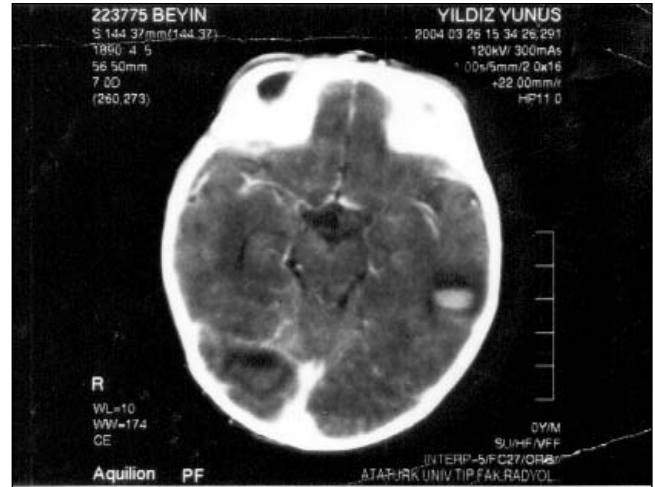


Fig 1: CT brain scan.

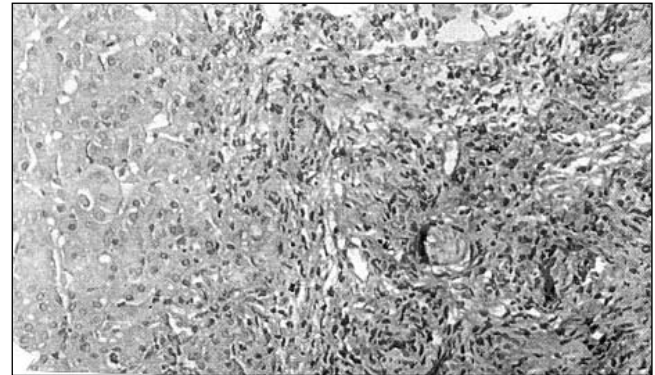


Fig 2: An expanded, inflamed portal tract at left contains many proliferated bile ducts, some of which are filled with inspissated bile. There is also panacinar cholestasis.

Answer on page 397

From: Faculty of Medicine, Departments of Pediatric Gastroenterology and Nutrition¹, Pathology², Ataturk University, Erzurum, Turkey.

Correspondence: Dr V Ertekin, Atatürk Üniversitesi, Tıp Fakültesi, Çocuk Sağlığı ve Hastalıkları AD, 25200, Erzurum, Turkey. Fax:+90 442 2361301, e-mail: vildanertekin@hotmail.com.