

CLINICAL QUIZ

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Gülhane Tıp Dergisi 45 (4) : 397 - 398 (2003)

Quiz :

A 8 month-old girl was referred for evaluation of hepatomegaly with a two months history of abdominal distention. Her past medical and familial histories were irrelevant. On physical examination her weight and length in the 25th percentile and the liver was 4 cm below the right costal margin with soft surface. The rest of physical examination was normal. Results of laboratory studies showed mild elevations of the aminotransferase levels (2x normal). Blood count, bilirubin, albumin, alkaline phosphatase levels including prothombin time were within normal limits. Alpha - fetoprotein (AFP) was 1.5 times higher than normal levels. There was no indicative serologic marker for hepatitis B,C, TORCH and EBV viruses. An abdominal ultrasonography revealed multiple, hypoechoic, irregularly solid lesions (maximum 21 mm in diameter) located in both lobes (fig. 1). An abdominal CT showed that these solid lesions were hypodense in precontrast and usually hyperdense in postcontrast exposures with central hypodensity (fig. 2). The patient underwent laparotomy for biopsy.

What is the most likely diagnosis ?

- A) Hepatoblastome
- B) Infantil hemangioendothelioma (IHE)
- C) Metastatic liver tumor
- D) Hepatic adenoma

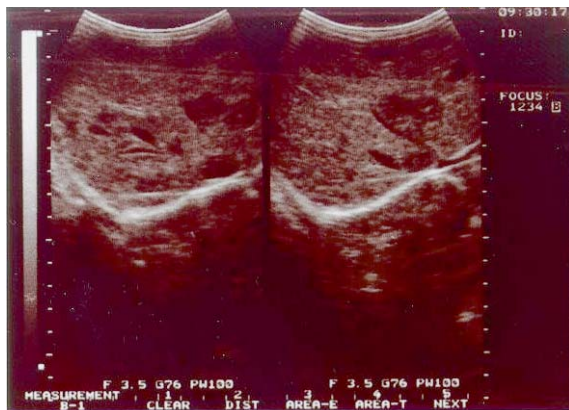


Figure 1: Ultrasonographic image of 8 month-old girl with multiple lesions on both hepatic lobes.



Figure 2: Abdominal CT scanning of the case

Answer :

Histopathological examination of the biopsy specimens revealed well demarcated, non - encapsulated benign tumor consist with vascular channels, lined by endothelial cells demonstrated by staining with factor VIII - related antigen and CD 34 (fig 3, fig 4). The patient was diagnosed with IHE then.

Comments :

A) Hepatoblastoma is the most frequent primary malignancy of the liver. In ultrasonographic study, they are sometimes hypoechoic and multifocal which are involving both lobes. Although AFP is characteristically elevated, some cases with normal level was also reported.

B) IHEs are the most common symptomatic vascular liver tumors presenting before 6 months of age either with a mass or hepatomegaly. Up to 45% of patients have cutaneous hemangiomas. Sonographic images of IHE show variable echogenicity and they may appear as soliter large lesions or multifocal-diffuse lesions. On CT, they show typical vasculer pattern. AFP levels can be within normal range or mildly elevated.

C) Metastases are more common than primary malignancies. With sonography, most metastases appear as hypoechoic, solitary or multiple masses. On CT, they are usually low- density lessions.

D) Hepatic adenomas are uncommon but frequently associated with metabolic disease. In glycogen storage diseases, they are usually multiple rounded lesions of variable size and echogenicity. CT usually reveals discrete hypodense nodules.

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Doğancı

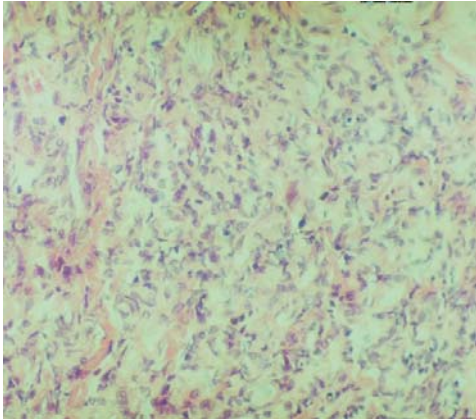


Figure 3: Liver pathology with factor VIII related antigen staining

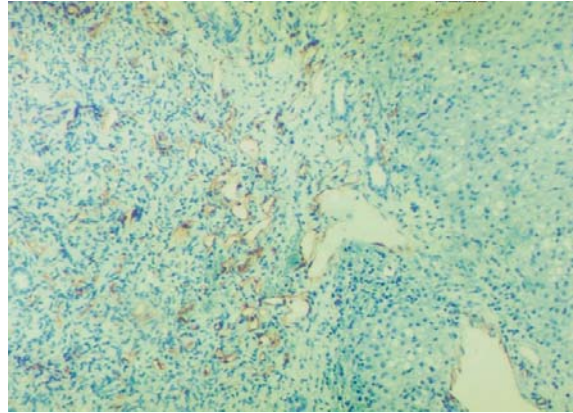


Figure 4: CD 34 staining of the hepatic tissue