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Clinical Image

Hepatic Epithelioid Hemangioendothelioma in a 2-Month-Old Infant

Zouari M [1,2*], Mansour MH[1,2], Aloulou H[2,3], Kammoun T[2,3] and Mhiri R[1,2]

[1]Department of Pediatric Surgery, Hedi-Chaker Hospital 3029 Sfax, Tunisia

[2]Sfax Medical School, University of Sfax, Sfax, Tunisia

[3]Department of Pediatrics, Hedi Chaker Hospital, Sfax, Tunisia

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1. Clinical Image

A 2-month-old male child presented with multifocal liver lesions, noted by abdominal ultrasonography. The Ultrasonography was done as part of a work-up for abdominal distension. He was asymptomatic and had been previously healthy. On examination, the liver was enlarged (liver span–15 cm). Blood investigations showed abnormal liver function tests with mild elevation in aspartate transaminase and alanine transaminase and deranged coagulation profile. The patient was started on vitamin K supplements. Ultrasonography and computed tomography of the abdomen showed hepatomegaly with diffusely scattered multiple target lesions in both the lobes, largest lesion measuring 5 x 4 cm. Based on radiology, provisional diagnosis of Epithelioid Hemangioendothelioma and metastatic liver disease were considered. Fine needle aspiration biopsy revealed epithelioid tumor cells with intracytoplasmic lumina containing red blood cells and a myxoid to sclerotic stroma, consistent with a diagnosis of HEHE. The patient developed acute hepatic failure and died before chemotherapy.

Hepatic epithelioid haemangioendothelioma (HEHE) is a rare vascular sarcoma of endothelial origin [1]. The etiology of this tumor is unknown and has a variable clinical outcome. It usually affects adults and is extremely rare in children [2]. Owing to the paucity of available information regarding its clinical course, pathogenesis, prognostic indicators and treatment outcome, the diagnosis of this tumor is challenging for the pathologist and the management disappointing for the clinician.

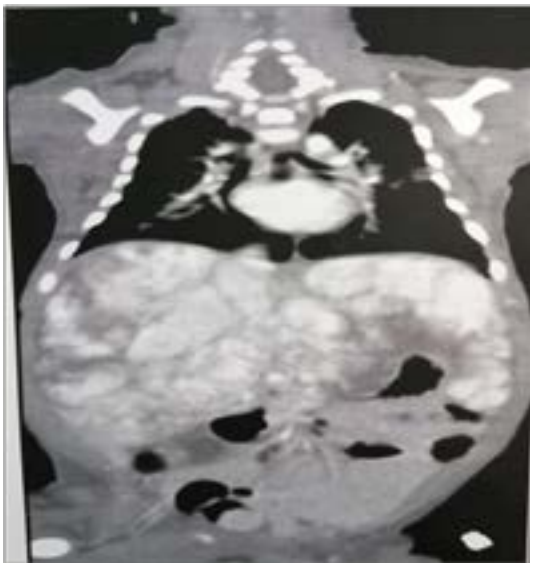

Figure 1: Frontal computed tomography scan cuts showing diffusely scattered multiple target lesions in both the lobes of the liver

***Corresponding Author (s):** Mohamed Zouari, Department of Pediatric Surgery, Hedi Chaker Hospital, 3029 Sfax, Tunisia, Tel: +21697459586, E-mail: zouarimohamed.1982@yahoo.fr, mohamed.zouari@rns.tn*

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