

Gastrointestinal and Liver Pathology at Rush

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Case of the Month Answer – July 2008

Contributed by Drs. Dawn Bradley and Shriram Jakate

Diagnosis: Epithelioid Hemangioendothelioma

Epithelioid hemangioendothelioma is a rare vascular endothelial tumor of unknown etiology and variable clinical course and is intermediate between hemangioma and angiosarcoma. It is a slow-growing tumor that usually occurs in middle-aged patients whom may present clinically with constitutional symptoms including anorexia, vomiting, abdominal discomfort, and weight loss. Most patients are asymptomatic (42%). Grossly, the lesions are multiple and distributed throughout the liver, and often there is a main mass with separate satellite masses. This characteristic mimics metastatic disease. Microscopically, these tumors have central densely sclerotic, hypocellular myxoid areas or occasional necrosis and peripheral cellular regions of epithelioid and/or dendritic cells. The tumor cells may extend through the sinusoids, central veins, and portal vein branches. Factor VIII and CD31 immunostains (Figs. 4 & 5, respectively) establish the endothelial origin of this tumor. The clinical course of epithelioid hemangioendothelioma in liver is variable and unpredictable. Mitosis or necrosis in the tumor is not related to tumor behavior. Surgical excision or liver transplantation is the treatment of choice, resulting in prolonged survival (5-28 years), and the presence of metastasis is not a significant contraindication for surgery and does not appear to affect survival.

References:

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2. G. Klatskin, H. Conn. *Histopathology of the Liver*, Vol. 1, Ch. 25. Neoplasms of the Liver and Intrahepatic Bile Ducts. 1993, p. 385.

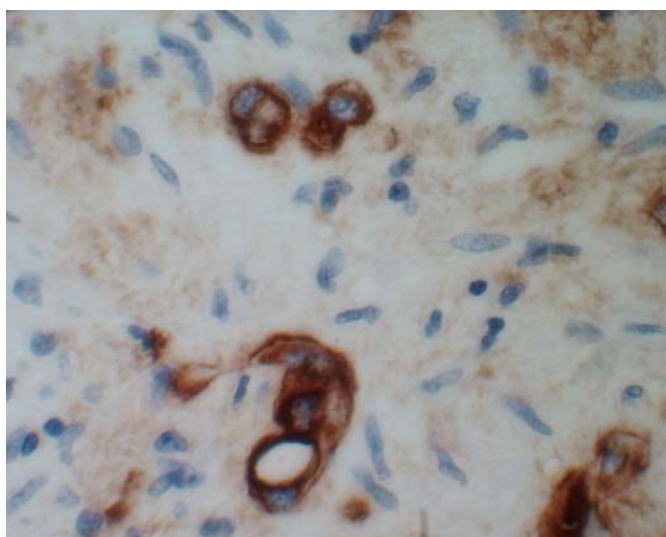


Figure 4

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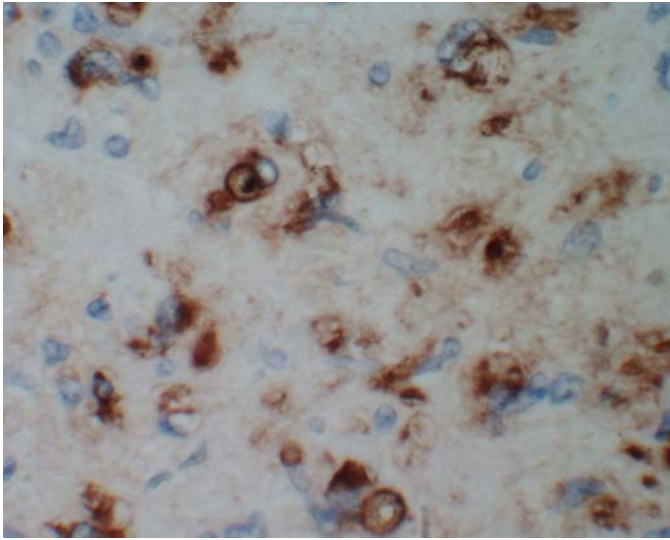


Figure 5