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Primary hepatic perivascular epithelioid tumor (PEComa)

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A 49 year-old woman was found to have a 2.9 cm hepatic lesion in segment 8, initially identified on an abdominal ultrasound performed for chronic abdominal pain and new onset diabetes mellitus, type 2. On computed tomography study the lesion was arterial enhancing with washout on portal venous and delayed phase and was classified as a LI-RADS category 4 (LR-4), suggestive but not diagnostic of hepatocellular carcinoma (HCC); the remainder of the exam was normal with no evidence of metastatic deposits. Of note, the background liver did not show any features of advanced fibrosis, but the patient had a past medical history of non-alcoholic fatty liver disease (NAFLD). The patient subsequently underwent a partial hepatectomy and on gross examination a well circumscribed grey tan lesion was identified. Histologically, the lesion showed a nested architecture, composed of epithelioid cells with round to oval nuclei, variable nucleoli and prominent granular eosinophilic cytoplasm (Figures 1A and 1B; * denotes background normal hepatic parenchyma). In areas the tumoral cells condensed around blood vessels (Figure 1B). The lesion also showed prominent fibrous trabeculae and pericellular fibrosis, raising the morphological differential diagnosis of a scirrhous or fibrolamellar subtype of hepatocellular carcinoma.

By immunohistochemistry the tumour cells were positive with antibodies to gp100 protein (HMB-45; Figure 1C), melan- A but negative for hepatocyte specific antigen (HSA) and S-100 protein.

The perivascular epithelioid group of tumors (PEComas) is characterized by their co-expression of melanocytic and muscle markers and includes angiomyolipoma, lymphangioleiomyoma, and clear cell ('sugar') tumors of the lung, pancreas, and uterus.[1-4] Primary hepatic PEComas are rare, but usually present in non-cirrhotic younger patients and on imaging studies often are suggestive of hepatocellular carcinoma. Although renal angiomyolipoma and pulmonary lymphangioleiomyomatosis are often seen in the setting of tuberous sclerosis complex, there has been no defined hereditary predisposition for hepatic PEComas to date. Fortunately, the majority of hepatic PEComas

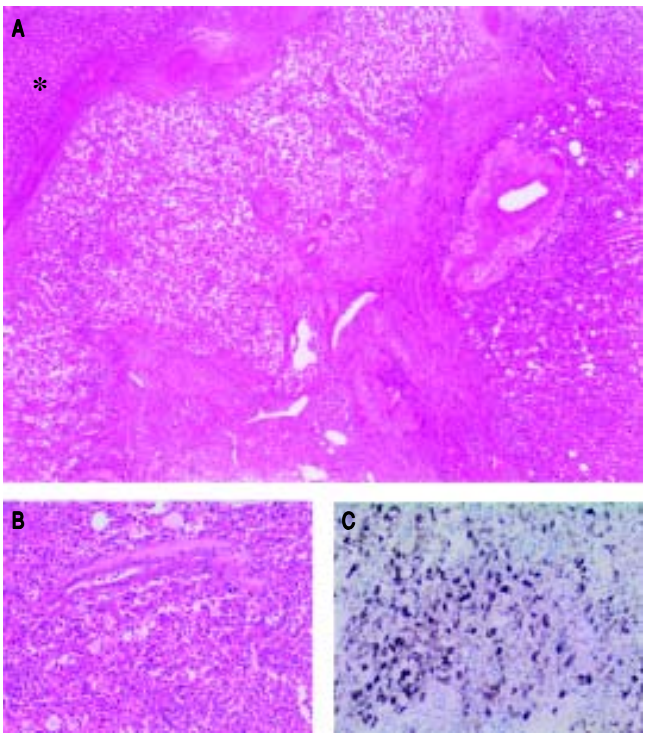

Figure 1. *Representative microscopic photomicrographs of a primary hepatic perivascular epithelioid tumor (PEComa).* ***A.*** *The lesion is well circumscribed and separated by a fibrous capsule from the normal background hepatic parenchyma (*). Also note the broad fibrous bands and prominent vasculature within the lesion itself.* ***B.*** *High-power image shows epithelioid tumor cells characterized by variable nucleoli and granular eosinophilic cytoplasms that condense around a vessel.* ***C.*** *The tumor cells strongly express the melanocytic marker HMB-45 (Magnifications: A 2x, B and C 200x).*

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reported in the literature behaved benign after complete resection with only one pulmonary metastasis reported.[4]

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