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Case Report Open Access

Perivascular Epithelioid Cell Tumor: A Rare Liver Primary of Mesenchymal Origin

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Abstract

Perivascular Epithelioid Cell Tumors (PEComas) are a family of rare neoplasms defined as mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells. There are only few reports in the literature of PEComas of liver origin. We presented a female patient with a perivascular epitheloid cell tumor of the liver treated with laparoscopic liver resection that remains disease free after 1 year. Surgical treatment should be considered in all patients with resectable PEComas. Follow-up is indicated based on the potential for malignancy and metastasis of these lesions.

Keywords: Hepatic perivascular epitheloid cell tumor; Hepatectomy; Mesenchymal tumors; HMB-45

Introduction

Perivascular Epithelioid Cell Tumors (PEComas) are a family of rare neoplasms defined as mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells [1]. Pea et al. first noted this unusual cell type in both renal angiomyolipoma (AML) and clear-cell "sugar" tumor (CCST) of the lung [2]. This generated the concept of a family of neoplasms composed of a distinctive cell type, which was immunoreactive with melanocytic markers, and exhibits an epithelioid appearance, a clear-acidophilic cytoplasm, and a perivascular distribution [3]. Cases of primary PEComa of the liver are considered extremely rare [4]. Prior to 2011, approximately 100 cases of PEComa had been described originating from different sites but only few cases were reported of hepatic origin [5]. We present a case of liver PEComa in a young female initially diagnosed as an adenoma with an area of intra-tumoral hemorrhage.

Case Presentation

A 40-year-old white female that presented with right upper quadrant pain radiating to the chest was referred to our center. The patient used oral contraceptives and had a past medical history of early carcinoma of the cervix treated with surgical resection. Laboratory evaluation displayed normal liver function tests and alpha-fetoprotein levels.

A multiphasic liver protocol CT showed multiple focal hepatic masses including several simple hepatic cysts, a 2.5 cm segment VII hemangioma, and a 4 cm heterogeneous arterially enhancing mass in segment VI which became hypodense to the liver parenchyma ("washout") on portal venous phase images (Figure 1). Imaging findings were most suggestive of an adenoma with intralesional hemorrhage.

The case was reviewed at our institutional multidisciplinary hepatobiliary tumor board. Given concerns for future hemorrhage and/or malignant transformation and after discussion with the patient, the decision was made to perform a laparoscopic resection of the segment VI of the liver. The resection was performed without major complications.

Microscopic examination of this lesion demonstrated thick walled vessels, with cells containing clear cytoplasm and prominent perinuclear eosinophilic condensations. Scattered fat cells were seen in less than 5% of the tumor with areas of intratumoral hemorrhage.

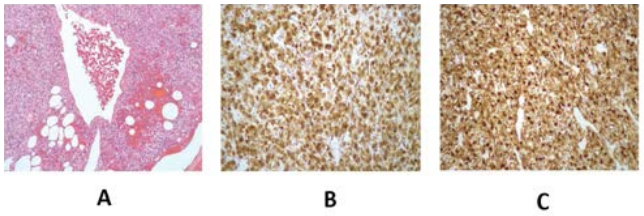

Figure 1: (A) Pathological findings of hepatic PEComa. (B) HE staining: tumor cells containing clear cytoplasm and prominent perinuclear eosinophilic condensations, including scattered fat cells. Immunohistochemical staining showing strong expression of melan-A and (C) HMB45 positive.

Immunohistochemistry demonstrated that this tumor was strongly positive for melanocytic markers HMB-45, Melan-A and vimentin (Figure 2).

Discussion

PEComas are a family of very rare mesenchymal tumors. Renal AML is the most common PEComa, with a reported prevalence of 0.13% in healthy adults which are more commonly seen in women than in men [1]. PEComas show a marked female predominance that may hint at the role of progesterone in the pathogenesis and/or phenotypic expression [1]. Several hypotheses have been suggested regarding the origin of these tumors. The most common is that PEC tumors derive from neural crest cells with the ability to express dual myoid and melanocytic phenotype [2].

Tsui et al. in an extensive review of the literature reported 30 cases of hepatic angiomyolipomas. Twenty-five of 30 patients were female and 3 had Tuberous Sclerosis Complex (TSC) [4].

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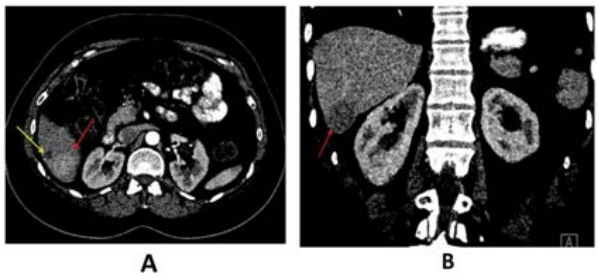

Figure 2: (A) Dynamic post contrast CT images. (B) Hepatic arterial phase image shows a mass in hepatic segment 6 (red arrow) with moderate enhancement relative to background liver. Coronal reconstruction in the portal venous phase shows the partially exophytic mass becomes hypoattenuating relative to the liver ("washout"). Note is made of an adjacent hepatic cyst (yellow arrow, A).

The most extensive review of the literature reported in 2005 found only 61 cases in different locations. The authors described that 100% were HMB-45 positive, 59% were smooth muscle actin positive, 41% were Melan-A positive, 33% were CD117 positive, 31% were desmin positive, 11% were S-100 positive, and 0% were cytokeratin positive [4]. Significantly, the pathologic features of our case were consistent with those of a perivascular epitheloid clear cell tumor, characterized by its predominantly epithelioid appearance with clear to eosinophilic cytoplasm, perivascular distribution, and strong immunoreactivity for HMB-45 antibody, melan-A and vimentin [4,5].

Pre-operative diagnosis of PEComas is very challenging since they have non-specific clinical or radiological features [4]. Some of the lesions considered for differential diagnosis include gastrointestinal stromal tumor, hepatocellular carcinoma, adenoma, leiomyoma, melanoma, and angiosarcoma. Tan et al. performed a retrospective analysis of 7 hepatic PEComas with emphasis on radiologic characteristics of these lesions. They found on contrast-enhanced CT and MRI that these tumors heterogeneously enhanced on arterial phase, and decreased enhancement on portal venous phase and delayed phase. In 2 out of these 7 patients no specific diagnosis was established, one had multiple lesions with delayed enhancement; 3 were diagnosed as HCC and 2 as focal nodular hyperplasia [6].

The majority of the PEComas reported in the literature thus far demonstrated benign behavior, although aggressive PEComas with malignant transformation and metastatic potential have been described [1]. In an attempt to better identify those tumors at higher risk for malignant transformation, Folpe et al. proposed a classification based on criteria such as tumor size greater than 5cm, infiltrative growth pattern, high nuclear grade, mitotic activity greater than 1 per 50 high power fields, and presence of necrosis [4].

Surgical resection in attempt to obtain clear margins remains the mainstay of treatment for PEComas [2,4]. Recently the use of mTOR inhibitors has shown promise for tumors originally deemed unresectable, providing a possible treatment modality for further management of PEComas.

We described a case of a young female with a perivascular epitheloid cell tumor of the liver treated with laparoscopic liver resection that remains disease free after 1 year. In conclusion, PEComas are a group of rare mesenchymal neoplasms which may occur in the liver. These tumors have nonspecific clinical and imaging features with histological and immunohistochemical techniques playing a key role in diagnosis. Surgical treatment should be considered in all patients with resectable PEComas. Follow-up is indicated based on their potential for malignancy and metastasis of these lesions.

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