

Acute abdomen as an unusual presentation of hepatic PEComa. A case report

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ABSTRACT

Perivascular epithelioid cell (PEC) tumors (or PEComas) are myomelanocytic lesions defined by coexpression of melanocytic and muscle markers, suggesting dual differentiation. They are rare mesenchymal tumors and include subtypes with distinct clinical features: angiomyolipoma, lymphangiomyomatosis, and clear cell "sugar" tumors of the lung, pancreas and uterus. Consequent upon the World Health Organization's recognition of PEC-derived tumors as a distinct entity, an increasing number of reports has documented PEComas arising at various anatomical locations. Clear cell myomelanocytic tumors of the falciform ligament/ligamentum teres (CCMTs) represent a rare variant of the PEComas. These hepatic PEComas, different from angiomyolipoma of the liver, pose a clinical, radiological and morphological diagnostic challenge. Because of their rarity, the clinical features and biological behavior of these tumors have yet to be established.

We experienced our first case of CCMT in a 36-year-old woman who presented to our emergency department with a 3-day history of abdominal discomfort and progressive growth of an epigastric bulk. Intralesional hemorrhage was causing abdominal distension, which progressed to acute abdomen soon after. The hemoglobin concentration was 9.9 g/dL. Liver laboratory tests showed slight elevation of AST, ALT and gamma-GT. The alpha-fetoprotein level was not elevated. The radiological images showed a hemorrhagic mass with some bizarre features in left hepatic lobe, immediately adjacent to the ligamentum teres and falciform ligament. The patient underwent a left hepatic lobectomy. The diagnosis of CCMT was based on histological and immunohistochemical staining. The postoperative course was uneventful. The patient received no adjuvant treatment and is currently, 34 months after surgery, alive and disease free.

In this report we describe a peculiar and hitherto undescribed clinical presentation of this tumor and its further course. Moreover, we discuss previously undescribed diagnostic imaging. We recommend that all unusual carcinomas and mesenchymal tumors of the liver should be tested for HMB-45: when positive, there is a high likelihood of PEComa.

Key words: hepatic tumor, PEComa, acute abdomen.

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