Hepatic epithelioid angiomyolipoma of liver; A diagnostic dilemma

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Abstract
Epithelioid angiomyolipoma (EAML) is a rare mesenchymal neoplasia. EAML is included in group of the perivascular epithelioid cell (PEC) tumors known as PEComas. Most cases of epithelioid angiomyolipomas have been described in kidney we herein report a rare case of hepatic epithelioid angiomyolipoma masquerading as hepatic adenoma in a 30 year old female. The tumor was finally diagnosed after histopathology and immunohistochemical examination.

Introduction
EAML is included in group of the perivascular epithelioid cell (PEC) tumors known as PEComas. These tumors are also associated with tuberous sclerosis. Most of these cases have been reported in kidney, involvement of liver is rare with less than 50 cases described in literature. Most cases of hepatic angiomyolipoma are misdiagnosed as adenomas or other diseases on radiology as cases lack definite adipose tissue component. Final diagnosis could be reached through histology and immunohistochemistry testing.

Case report
A 30 year old female presented with vague abdominal pain and on evaluation was found to have a liver space occupying lesion. The lab investigations showed anemia with haemoglobin of 10.5 g/dl (12-15.0 g/dl). Liver and kidney function test were normal. CECT abdomen performed showed a large homogenously hypodense mass showing no calcification / hemorrhage measuring 80 x 70 x 55 mm seen in left lobe of liver involving caudate lobe. The mass was showing heterogenous arterial phase enhancement (periphery to centre) and remains slightly hyperdense as compared to hepatic parenchyma on hepatic and delayed phases. The possibility of hepatic adenoma was kept based on radiological findings [Figure 1a, b].

Patient underwent segmental liver resection of the mass.

Grossly – A partly nodular partial liver segmental resection specimen was received measuring 8 x 8 x 3.5 cms. Cut surface shows a well defined black brown vascular mass measuring 6.5 x 6 x 4 cms [Figure 2a].

Microscopy from the mass showed mainly epithelioid cells arranged in sheets and trabeculae having abundant eosinophilic cytoplasm and round nuclei [Figure 2b]. There was rich vascularized stroma with thick walled vessels. Aggregates of foam cells with fine lipid droplets noted [Figure 2c]. The neoplastic cells are diffusely HMB45 positive and negative for HepPar 1 [Figure 2d].

There were no features suggestive of aggressive behviour in form of vascular metastasis, pleomorphism or p53 expression.

The case was diagnosed as hepatic epithelioid Angiomyolipoma. Patient was well after resection and on regular follow up. Futher evaluation for tuberous sclerosis was performed but brain scan was negative.

Discussion

In 2013, the World Health Organization (WHO) defined neoplasms with perivascular epithelioid cell differentiation (PEComas) as mesenchymal tumors composed of distinctive cells that show a focal association with blood vessel walls and usually express melanocytic and smooth-muscle markers[1].

PEComas include Angiomyolipoma (AML), lymphangioleiomyomatosis and clear cell ‘sugar’ tumour [2].

AML can occur anywhere in body, but most cases arise in the retroperitoneum, abdominopelvic region, uterus, and gastrointestinal tract [3]. Occurrence of hepatic AML is very rare.

Most cases are asymptomatic and are detected incidentally on routine examination. As the lesion enlarges cases can present with abdominal pain. Tuberous sclerosis is associated with over half of the cases of renal AML and 5-15% of the cases of HAML [4].

Epithelioid PEComas is a variant of AML composed almost exclusively of epithelioid cells with pronounced abnormal blood vessels and few or no lipocytes [5,6]. Due to relative paucity of fat content most cases are misdiagnosed as adenomas on radiology as was also noted in our case. Most cases of EAML are found in kidney. Involvement of liver is very rare. Presence of solitary EAML usually indicates primary hepatic EAML and surgical excision is treatment of choice. In case of multiple lesions within the liver possibility of metastasis from renal EAML remains and have poorer prognosis.

Brimo et al have summarized the pathological characteristics of renal EAML likely associated with malignant progression: [1] ≥ 2 mitotic figures per 10 high-power field; [2] atypical mitotic figures; [3] ≥ 70% of atypical epithelioid cells; and [4] necrosis. The presence of 3 or more features was highly predictive of malignant behavior [7].

The criteria for malignant transformation of HEAML are however not well defined due to scarcity of these lesions. [8, 9] Deng et al have postulated that large tumor size, pleomorphic nuclei with high proliferation activity, and P53 immunoreactivity may predict the existence of malignant transformation of hepatic AML [8].

Because of the rarity of these tumors in the liver, absence of significant fat component most cases pose a diagnostic dilemma. Definite diagnosis can be established only by histopathology and can be confirmed by immunohistochemical marker testing. Due to rare association of these tumors with tuberous sclerosis and renal lesions further evaluation of these patients is suggested as opposed to adenomas where surgery is curative. The aim of this report is to increase awareness among pathologist of this rare entity.

References

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