CASE REPORT

A rare hepatic tumour mimicking hepatocellular carcinoma

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Abstract

Rare hepatic tumours are often only confirmed after image guided biopsy or excisional biopsy with histology and immunohistochemistry studies. In this case we present a rare benign hepatic tumour in a 53 year old female which mimicked hepatocellular carcinoma. She was managed successfully with hepatic segmentectomy and had an uneventful recovery with no signs of recurrence 6 months after surgery. Post operative immunohistochemistry of the resected sample confirmed the rare diagnosis.

Keywords: hepatic PEComa, hepatocellular carcinoma, hepatic segmentectomy

Introduction

Primary hepatic tumours can be benign or malignant with the commonest benign tumours being haemangiomas while the commonest primary malignant tumours being hepatocellular carcinoma. The tumours are diagnosed by a combination of radiological imaging and biopsy. Rarer benign hepatic tumours are often only confirmed after image guided biopsy or excisional biopsy and histology and immunohistochemistry. In this case we present a rare tumour which mimicked hepatocellular carcinoma. Perivascular Epithelioid Cell tumour (PEComa) is a rare mesenchymal tumour which shows female predominance and consists of spindle cells with granular eosinophilic cytoplasm and perivascular accentuation. Only about 100 cases have been reported in English medical literature with most of them arising from the uterus and more rarely as distant metastases to liver, lungs, intestines, bone and lymph nodes(1). {Armah, 2009 #29;Armah, 2009 #29} Furthermore only about 20 to 25 cases of primary hepatic PEComas have been reported in the current literature with most of them being single case reports(2, 3).

Case presentation

A 53 year old female presented with generalized weakness and malaise for 2 months and vague abdominal discomfort for about 1 month not associated with vomiting, fever or jaundice or any other constitutional symptoms.

Diagnostic assessment

An abdominal ultrasound scan had found an incidental mass in segment VI of the liver.

The patient had a contrasted abdominal CT scan done which showed a mass (51x26x40) millimetres in segment VI of the liver which was isoattenuating in the prephase, hyperattenuating in the arterial phase and isoattenuating in the venous phase (CT characteristic wash out) with grossly normal

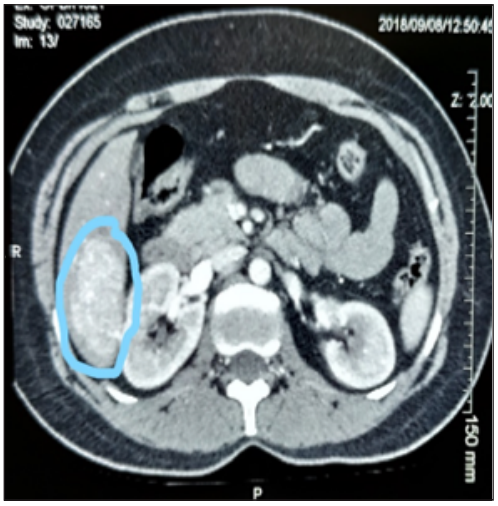

Figure 1. CT image of the liver lesion

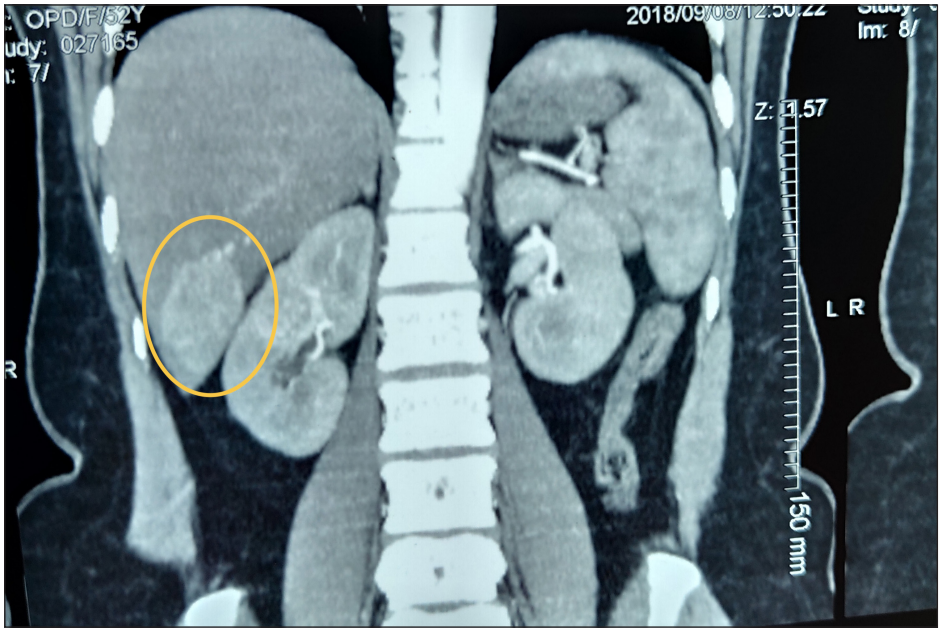

Figure 2. CT image of the liver lesion

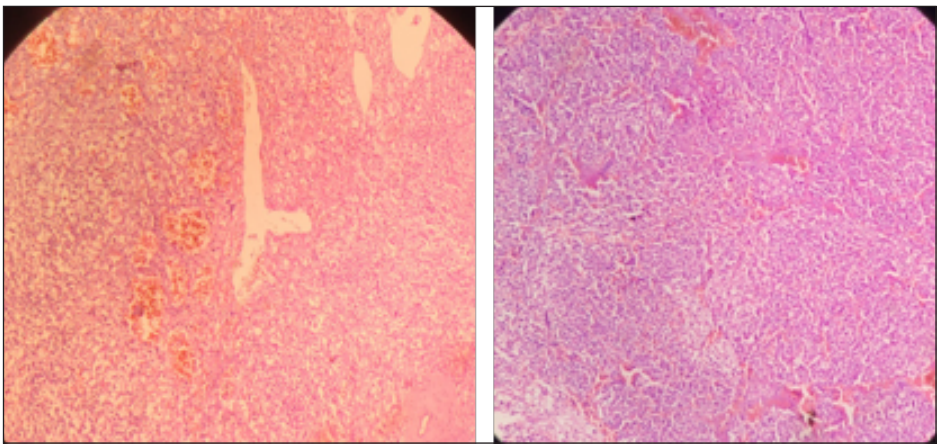

Figure 3. Immunohistochemistry staining for PEComa

surrounding liver parenchyma (Figures 1 and 2). There were no enlarged lymph nodes and the no other lesions seen in the rest of the abdominal organs. She had a normal complete blood count, liver function tests and renal function tests. She also had a normal chest x-ray. An echocardiogram done showed mild pericardial effusion along the right atrium and right ventricle. She was diagnosed with possible hepatocellular carcinoma localized in segment VI (six) of the liver and hepatic segmentectomy was planned. The patient did not do any test for alpha-fetoprotein levels.

Management

The patient underwent resection of segment VI of the liver where tumour was located. She made an uncomplicated recovery.

Histopathology showed a vascular tumour 5cm by 3.5cm by 2.5cm with normal surrounding liver parenchyma. Further immunohistochemistry results showed a tumour which was staining positively for HMB45 and Melan A which confirmed a diagnosis of Perivascular Epithelioid Cell tumour (PEComa), a rare mesenchymal tumour (Figures 3 and 4).

Follow up and outcomes

The patient has been followed up for 6 months and has no symptoms and no radiological evidence of recurrence. We plan to follow up the patient every 3 months for the one year following surgery then every 6 months for the following two years.

Table 1. Correlation coefficients, limits of detection, and limits of quantification of heavy metals in a certified reference sample

Date	Clinical presentation	Intervention
10 September 2018	Abdominal CT Scan showing vascular liver mass segment VI following 1 month history of vague abdominal pain	Worked up for surgery with 2D Echo, Chest x-ray, complete blood count, liver function and renal function tests, all normal
22 September 2018	Admitted for hepatic segmentectomy	Final work up for surgery complete blood count, liver function and renal function tests, PT/INR
23 September 2018	Underwent laparotomy and hepatic segmentectomy of segment VI	Surgery uneventful. Specimen sent for histopathological diagnosis
1 October 2018	Discharged in good general condition	Preliminary histopathological results show vascular tumour with normal surrounding liver parenchyma
10 October 2018	Reviewed and had no complaints	Immunohistochemistry results confirmed PEComa
30 January 2019	Reviewed no complaints	Repeat abdominal CT scan no evidence of recurrence or any other lesions in abdominal organs.

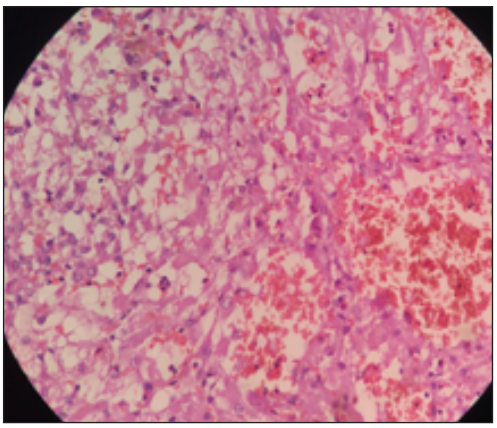

Figure 4. Immunohistochemistry staining for PEComa at a higher magnification

Discussion

A PEComa is a rare mesenchymal tumour which shows female predominance and consists of spindle cells with granular eosinophilic cytoplasm and perivascular accentuation. Only about 100 cases have been reported in English medical literature with most of them arising from the uterus and more rarely as distant metastases to liver, lungs, intestines, bone and lymph nodes. Almost all cases of non-uterine PEComas have been reported in females with a wide age range at diagnosis and an asymptomatic painless mass which can occur in any part of the body including skin, heart, gastrointestinal tract, ovaries and genitourinary tract(1). Upto 2014 only 33 cases of primary hepatic PEComas have been reported in the English literature with most of them being single case reports(2, 4). Other literature have reported not more than 25 reported cases of hepatic PEComa(3). The primary lesion can occur in any part of the liver with case reports of lesions in segment four (IV) of the liver(2),the right lobe(5), segment eight (VIII) of the liver(3) and in the left lobe of the liver (6). In most of the cases the lesion was initially described as possibly a haemangioma due to its high vascularity on Doppler abdominal ultrasound (2, 5, 6). Most of the hepatic PEComas reported were benign tumours with no evidence of local recurrence while in one case report it was a malignant lesion which recurred after 8 months(3). The liver is one of the sites for metastases from primary malignant PEComas in the kidney or uterus and especially if the primary tumour is large and well circumscribed(7). Diagnosis of a PEComa is mainly by immunohistochemistry whereby it will stain positively for Human Melanin Black 45 (HMB45), Melan A and smooth muscle actin (SMA) (1).Patients usually present with an asymptomatic mass in the liver which may be misdiagnosed as a haemangioma, hepatocellular carcinoma or adenomas(6, 8-10). The lesion is usually identified as an incidental finding following abdominal CT scan or PET scans for other unrelated symptoms. In patients presenting with liver lesions which on enhancement of radiological imaging look like hepatocellular carcinoma, PEComa as a differential diagnosis should be considered if there are no other abnormalities in the liver parenchyma and hepatitis virus markers are negative(4). In a review of case reports only one of the patients was diagnosed by preoperative image guided biopsy with the rest being diagnosed after liver resection surgery(11). Most hepatic PEComas are benign but some may show aggressive behavior with recurrence occurring from 3 months upto 9 years after surgery (3). According to Folpe et al PEComas may be classified as benign, uncertain malignant potential or malignant based on the following criteria tumor size >5cm, infiltrative growth pattern, high

nuclear grade, necrosis and mitotic activity >1/50 HPF and subsequent aggressive clinical behaviour of PEComas (12). The management is mainly surgical resection of the tumour through right or left hepatectomy or by segmentectomy. There is no follow up protocol for these patients but most of the cases reported were following up patients every 3 months with some following them up for upto 9 years with no evidence of recurrence(13). Our patient has been followed up for 6 months with no evidence of recurrence.

Conclusions

Hepatic PEComa though a rare condition should be considered as a differential diagnosis in female patients with vascular liver masses mimicking hepatocellular carcinoma and can be managed successfully by surgery with good outcomes.

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