Malignant Hemangiopericytoma of the Liver Masquerading as Hepatocellular Carcinoma

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Abstract

Isolated, metastatic hemangiopericytoma of liver is an extremely rare entity. We present a case of hemangiopericytoma of the liver, metastatic from a meningeal hemangiopericytoma, who presented 10 years after the surgical excision of the primary tumour and morphologically mimicked a hepatocellular carcinoma. We review the literature regarding this entity and discuss the difficulties in preoperative diagnosis and the need for a thorough preoperative evaluation.

Keywords Metastatic hemangiopericytoma · Hepatocellular carcinoma · Isolated hepatic metastases · HCC

Introduction

Hemangiopericytoma is an extremely rare mesenchymal tumour which arises from the Zimmerman's pericytes [1]. An intracranial origin is rarer still (< 1%) and primarily involves the meninges [2]. Though hemangiopericytomas are known to metastasize to liver, an isolated, single, metastatic hemangiopericytoma of the liver is distinctly unusual. We present a case of metastatic hemangiopericytoma of liver (primary from meninges), who presented 10 years after the surgical excision of the primary

and had the morphological appearance of a hepatocellular carcinoma (HCC).

The Case

A 55-year-old lady presented with intermittent, dull aching pain over the right upper abdomen for 2 months. She also noticed a lump over the right upper abdomen which was gradually increasing in size. This was associated with significant loss of appetite and weight (~5 kg). There was no history of jaundice, fever, gastrointestinal bleeding or abdominal distension. She had a past history of excision of a parasagittal meningioma 10 years back and was on maintenance with oral phenytoin. She also underwent laparoscopic hysterectomy and bilateral salphingo-oophorectomy for uterine leiomyoma 9 years back. She was also diagnosed with symptomatic lumbosacral perineural cyst in 2011 and was planned for surgery which she deferred due to symptomatic improvement. She did not have any co-morbidities other than well-controlled hypertension. The general physical examination was unremarkable. Abdominal examination revealed a large, non-tender, firm mass in the right hypochondrium. There was no lymphadenopathy or ascites. An ultrasound abdomen revealed a solid, well-circumscribed, encapsulated mass involving almost the whole of left lobe of the liver. Rest of the liver showed normal echotexture, and there was no ascites or collaterals. Computed tomography of the abdomen (Fig. 1) showed a well-capsulated mass arising from segments II/III of the liver with intense enhancement on arterial phase with washout on

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Fig. 1 **a** Approximately 5 × 6 cm mass in segment II and III of the liver, with intense arterial enhancement. **b** Lesion shows wash out in venous phase. Cut-off of left portal vein can be noticed

venous and delayed phase, suggestive of a HCC. The main and right portal veins and hepatic arteries were free, though the left portal vein and hepatic artery were involved by the tumour. There was no calcification or

satellite lesion. Computed tomography of the chest and upper gastrointestinal endoscopy were normal. Patient had undergone a fine needle aspiration cytology (FNAC) at another hospital which was reported as being suggestive

Fig. 2 Microscopic examination showed a well-circumscribed (a) malignant mesenchymal tumour of spindle cell type (b) having the characteristic staghorn type of vessels (d). Frequent mitotic activity was seen on histopathology (15–16/1-HPF) which was suggestive of a malignant potential (c)

of HCC. Liver function tests, viral serology (HBsAg and anti-HCV IgG antibody) and serum alpha-fetoprotein (AFP) were within normal limit. She was diagnosed with HCC of left lobe of the liver arising in non-cirrhotic background and planned for left hepatectomy. Intraoperatively, there was no evidence of any dissemination and a large mass was found involving segments II/III and a left hepatectomy was performed. Postoperative period was uneventful and she was discharged on postoperative day 5. The final histopathology held a surprise. Gross examination showed a $13 \times 13 \times 8.5$ cm well-circumscribed tumour replacing almost the whole of left lobe of the liver, lying <0.1 cm from the capsule and 1.2 cm from the cut surface. Multiple sections were suggestive of a malignant mesenchymal tumour, spindle cell type with dilated hemangiopericytoma-like vessels. Mitotic activity was 15–16/10HPF and tumour cells were immunopositive for CD34 (focal) and Bcl2 while being negative for CD117, DOG-I, SMA, desmin and EMA. Overall features were suggestive of malignant hemangiopericytoma (Fig. 2).

In view of the rarity of this tumour, we reviewed the previous slides of meningioma which was also suggestive of a hemangiopericytoma. So, a final diagnosis of metastatic hemangiopericytoma of the liver was made. She is doing fine without any evidence of recurrence at 21 months of follow-up. (Informed consent was obtained from the patient for publication of the case.)

Discussion

Hemangiopericytomas originate from the contractile spindle cells surrounding the capillaries and postcapillary venules known as Zimmerman's pericytes [1]. Though they can arise anywhere in the body, they most commonly occur in the extremities and retroperitoneum [3]. Hemangiopericytoma has been reported intracranially in less than 1% of cases, mostly from meninges [2]. A preoperative diagnosis of meningeal hemangiopericytoma is usually unavailable due to its rarity and the clinical and radiological similarity to meningioma [4]. In our case too, the patient underwent the meningeal surgery with a presumed diagnosis of meningioma. Role of adjuvant therapy after complete excision of this tumour is controversial [5] and our patient did not receive any adjuvant treatment. Delayed recurrence after primary excision has been reported, but most of these recurrences occur either at the primary site or at multiple metastatic sites such as the brain, lung, bones and liver [5]. Most of the recurrences are managed with palliative intent owing to the widespread nature of disease or the poor general condition of the patient [6]. Delayed presentation of an isolated, single site of recurrence in the liver is distinctly unusual, especially with a meningeal hemangiopericytoma [5, 7, 8]. The literature on isolated liver metastasis of a meningeal hemangiopericytoma is thin and consists only of a few case

reports. Only 14 patients have undergone a successful excision of the isolated liver recurrence in the entire reported English literature [7]. Preoperative diagnosis of hepatic metastatic disease becomes difficult when there is a long duration between the excision of the primary meningeal tumour and the recurrence (as in our case), and the hypervascularity of these tumours lends the morphologic appearance of a HCC. In the current case, a preoperative diagnosis of HCC was made on the basis of the typical radiological characteristics and was further supported by the cytological report. Since these tumours arise from capillary pericytes, they show arterial enhancement and washout on delayed phase. On reviewing the literature, we found that the majority of patients with isolated hepatic metastases are misdiagnosed as HCC on preoperative imaging, especially in the absence of the histological report of the primary [8–10]. Hence, these patients are treated like HCCs by resection, liver transplantation, trans-arterial chemoembolization and radiofrequency ablation [8–10]. Though resection would be the treatment of choice in patients with a true single site of recurrence, an evaluation for other sites of recurrence is mandatory before proceeding with any treatment. Lack of the correct preoperative histological diagnosis leading to incomplete evaluation has resulted in a variable recurrence-free and overall survival ranging from months to years [8, 9].

In conclusion, isolated, metastatic hemangiopericytoma of the liver is an extremely rare entity but one that should be kept in the differential diagnosis of patients who have previously undergone surgery for lesions elsewhere in the body. It is important to entertain the possibility of metastases in any patient who presents with a liver lesion and has undergone treatment of lesions elsewhere in the body irrespective of the radiological appearance of the liver mass. All attempts should be made to obtain and confirm the histology of primary lesions as well as to rule out other sites of dissemination.

Author Contribution ANS, RK, SP: Substantial contributions to the conception or design of the work;

ANS, RK, KSM, PD: Acquisition, analysis, or interpretation of data for the work;

ANS, RK, KSM, PD, SP: Drafting the work or revising it critically for important intellectual content;

ANS, RK, KSM, PD, SP: Final approval of the version to be published;

ANS, RK, KSM, PD, SP: Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Compliance with Ethical Standards

Conflict of Interest Singh AN, Kilambi R, Das P, Madhusudhan KS, and Pal S declare that they have no conflict of interest.

Consent Informed consent was taken from the patient before submission.

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