CASE REPORT

Perivascular epithelioid cell tumor (PEComa) of falciform ligament – an extremely rare tumor

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Abstract Primary tumor of falciform ligament is exceedingly uncommon. So far, seven cases of Perivascular Epithelioid Cell tumor (PECom), a recently described histological entity with characteristic perivascular tumor cell arrangement and expression of muscle and melanocytic marker by the cells, have been reported in literature. Here, we report a case of PEComa of falciform ligament, which presented with a huge abdominal lump. Pre-surgical CT scan of abdomen was uninformative regarding the nature and organ of origin of the tumor. On laparotomy, it was surprisingly found to arise from the falciform ligament, and this was easily removed. Histopathological diagnosis confirmed PEComa. There was no recurrence of disease after two years of follow up.

Keywords Pecoma · Falciform ligament

Introduction

Primary tumours of the falciform ligament are extremely rare. Folpe et al [1] had published a brief report of seven patients with primary neoplasms of the falciform ligament culled over a 50 year period. They were characterized by perivascular localization of eosinophilic cells and expressed melanocytic and muscle markers. Such tumours belong to the family of perivascular epithelioid cell tumours (PEComa), a recently described histological entity. We now present one such rare case of falciform ligament tumour.

Case report

A 35-years-old man presented with a history of gradually increasing painless swelling of the upper abdomen since one year. His appetite was normal, but there was some loss of weight. Systemic examination was within normal limits, while abdominal examination revealed a large intra-abdominal lump, but no free fluid. Routine blood investigations and a chest X-ray were normal. CT scan of the abdomen revealed a large heterogeneously enhancing complex mass with solid and cystic component. Due to its size it was difficult to identify the organ of origin. Laparotomy revealed a well encapsulated reddish brown solid-cystic tumour occupying almost the whole of the abdomen (Fig. 1). The flimsy adhesions around the tumour were taken down and the tumour was found attached to the falciform ligament (Fig. 2) on a narrow pedicle. The pedicle was transected and the tumour was removed in toto. Recovery was uneventful.

The tumour measured $22 \times 18 \times 6$ cm in size. Outer surface was lobulated. The cut section revealed solid and

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cystic components (Fig. 3). Cystic spaces were filled with hemorrhagic fluid. Microscopically, the tumour shows perivascular arrangement of cells (Fig. 4). The tumour cells are round to oval and having eosinophilic cytoplasm. Cytoplasmic vacuolation was also noticed. The cells were negative for both smooth muscle actin and HMB 45. Electron microscopical examination showed presence of premelanosomes in the cytoplasm of the tumour cells. The laboratory diagnosis was PEComa. Two years follow up period was available. There was no evidence of recurrence of disease.

Fig. 1 Operative picture showing a well encapsulated reddish brown solid-cystic tumour

Fig. 2 Operative picture showing the attachment of tumour to the falciform ligament

Fig. 3 Gross photograph of tumor showing solid and cystic areas

Fig. 4 Photomicrograph of tumor showing perivascular arrangement of tumor cells

Discussion

Our patient presented with a large mass on the abdominal CT scan, which was removed successfully. Due to the large size, the organ of origin was not easily evident, and only following laparotomy, it became clear that the mass was arising from the falciform ligament. Isolated neoplasm of the falciform ligament is extremely rare. Folpe et al [1] published a series of seven patients with tumours of falciform ligament, which upon histology showed clear to faintly eosinophilic spindled cells with elaborate vasculature. Immunohistochemically, all the tumours were HMB-45 positive and in three cases the tumours were also positive to smooth muscle actin antibody. The authors proposed the term 'clear cell myomelanocytic tumour' for such tumours, which is believed to be a member of the newly described family of tumour called PEComa. Our case was

negative for HMB 45 and smooth muscle actin. However, electron microscopy revealed presence of premelanosomes in cytoplasm of tumor cell consistent with the diagnosis of PEComa.

The term PEComa owes its origin to the finding of ‘perivascular epithelioid cell’ in a variety of seemingly unrelated tumours like clear cell ‘sugar’ tumour of lungs, angiomyolipoma of kidney and liver and lymphangiomatosis [2]. The tumours are usually HMB-45 positive upon immunocytochemistry and electron microscopy often reveals premelanosomes. These perivascular cells have no known normal cellular counterpart and are thought to originate from the walls of blood vessels. Subsequently, such clear cell tumours have been reported elsewhere: rectum, vulva, heart, breast, common bile duct, ligamentum teres hepatis, urinary bladder, skull base, skin and soft tissues of thigh and abdominal wall [3]. Due to the universal presence of such perivascular epithelioid cells in all these tumours, the term PEComa was later coined to embrace these apparently disparate neoplasms [4].

Epithelioid cell is the commonest cell type, while; uncommonly some tumours exhibit spindle cells. Interestingly, the vast majority of extra-renal extra-pulmonary PEComa show epithelioid cells [3], but in

contrast, falciform ligament tumours are composed predominantly of spindle cells [1].

The characteristic feature of such tumour is finding of neoplastic cells arranged around blood vessels. This perivascular finding of neoplastic cells should prompt confirmatory immunohistochemistry. Prognosis of PEComa is variable. Most of the reported cases behaved as benign fashion. However, lung metastasis occurred in one case [1]. In two years follow up there was no recurrence in our case.

References

1. Folpe AL, Goodman ZD, Ishak KG, Paulino AFG, Taboada EM, Meehan SA, Weiss SW (2000) Clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres. *The Am J Surg Pathol* 24:1239–1246
2. Bonetti F, Pea M, Martignoni G et al (1992) PEC and sugar. *Am J Surg Pathol* 16:307–308
3. Hornick JL, Fletcher CDM. PEComa: what do we know so far? *Histopathology* 48:75–82
4. Zamboni G, Pea M, Martignoni G et al (1996) Clear cell ‘sugar’ tumour of the pancreas. A novel member of the family of lesions characterized by the presence of perivascular epithelioid cells. *Am J Surg Pathol* 20:722–730