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CASE REPORT

Perivascular epithelioid cell tumour (PEComa) of the inferior vena cava presenting as an adrenal mass

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Abstract

A 54-year-old woman had a mass located in the right suprarenal area. On imaging, this mass appeared to be infiltrating the inferior vena cava (IVC). Exploratory laparotomy was undertaken and excision of the tumour was done with the sleeve of the involved IVC. The mass turned out to be a perivascular epithelioid cell tumour (PEComa) on histopathological examination. This report describes previously reported cases of PEComa in brief and highlights the problems associated with the management of this tumour.

Keywords: *Perivascular epithelioid cell tumour (PEComa); inferior vena cava; adrenal mass.*

Introduction

Perivascular epithelioid cell tumour (PEComa) is a very rare tumour consisting of sheets of epithelioid cells and is positive for melanocytic and smooth muscle antigen[1]. It is considered a benign tumour but can occasionally behave in malignant fashion. It can have varied presentation depending upon the size and the location of the tumour. Various sites such as skin, colon, bladder and soft tissue have been reported to be affected by this tumour but PEComa of the inferior vena cava (IVC) has not been reported in the English literature.

Case report

A 54-year-old woman presented with pain in the right lumbar region of 6 months duration. Her haematological and biochemical parameters were within normal limits. Ultrasound of the abdomen showed a large upper retroperitoneal mass which was probably arising from the right adrenal gland and invading the IVC.

Contrast-enhanced computed tomography (CT) of the abdomen was performed which confirmed the ultrasound findings, however, the organ of origin of this retroperitoneal mass was not clear. The mass measured approximately 7 × 7 cm and was heterogeneous in attenuation (Fig. 1). The IVC appeared infiltrated and the right renal artery was displaced inferiorly with loss of intervening fat planes. Magnetic resonance (MR) examination confirmed the CT findings and there was a

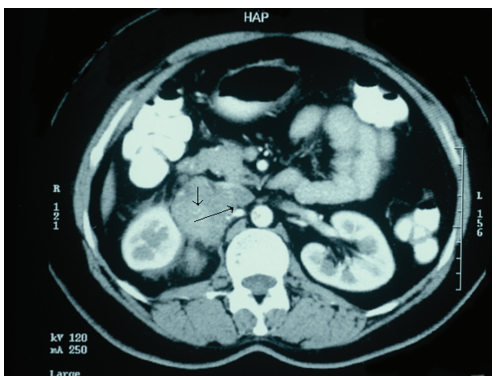

Figure 1 **Axial contrast-enhanced CT image showing a large heterogeneous lobulated mass in the right suprarenal location which is displacing and compressing the IVC (arrow) anteriorly. The IVC appears infiltrated by the mass as is the right renal artery (long arrow).**

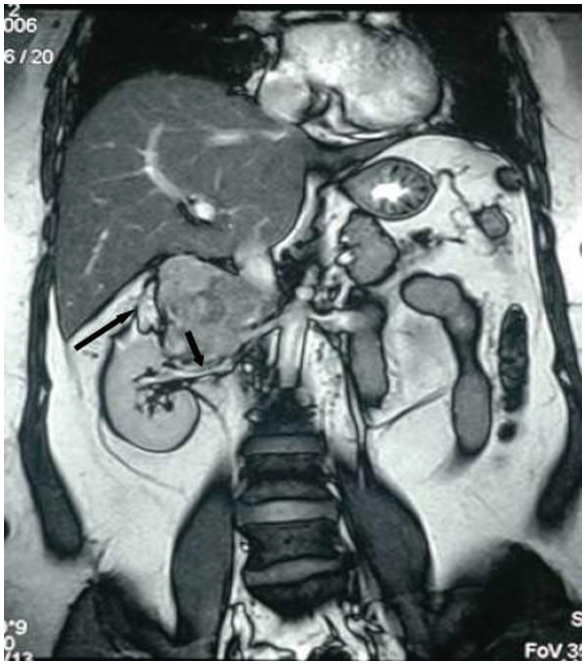

Figure 2 **MR True-Fisp coronal image showing the heterogeneous signal intensity mass displacing the right renal artery inferiorly (arrow). The adrenal (long arrow) appears separate and displaced laterally by the mass.**

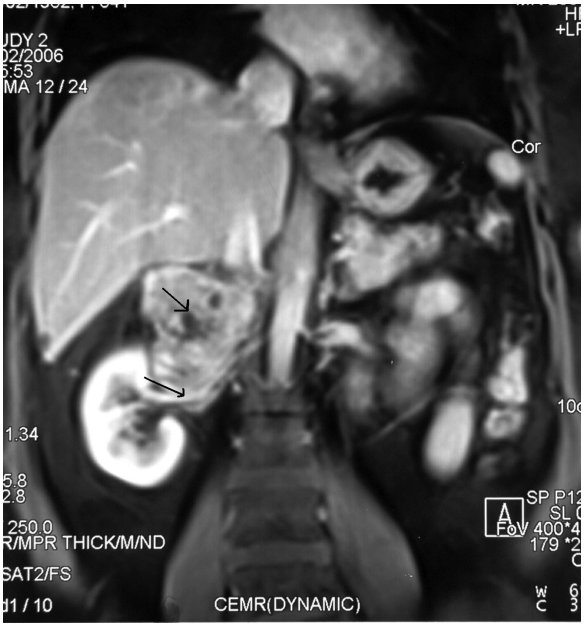

Figure 3 **Coronal contrast-enhanced MR image showing heterogeneous enhancement of the mass (arrow) in the expected adrenal location. The renal artery (long arrow) is displaced inferiorly by the mass.**

suggestion that the right adrenal was being displaced laterally by the mass, however this was not corroborated on the CT scan (Fig. 2). Coronal sections of the contrast-enhanced MR image showed heterogeneous enhancement of the mass in the expected adrenal location (Fig. 3). The renal artery was displaced inferiorly by the mass.

Suspecting pheochromocytoma and adrenocorticoid carcinoma, urinary vanillyl mandelic acid, serum cortisol and catecholamine levels were measured and found to be normal. On laparotomy, an 8×8 cm mass was seen arising from the right posterolateral wall of the IVC. The right adrenal gland was normal and was displaced superolaterally. En-bloc resection of the tumour with the sleeve of the IVC was performed. The histopathological report was consistent with PEComa of the IVC (Fig. 4).

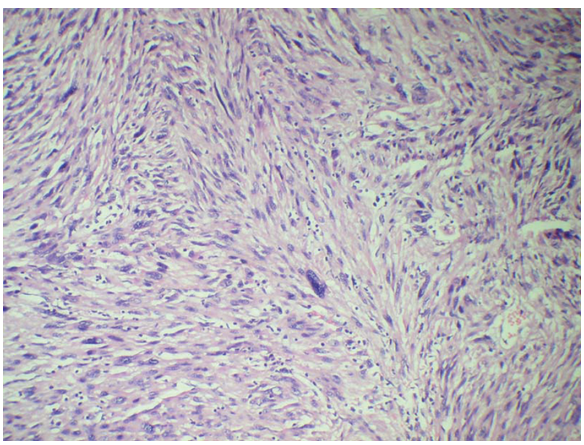

Figure 4 **Nests and sheets of epithelioid and occasionally spindled cells with clear to granular eosinophilic cytoplasm and a focal association with blood vessel walls consistent with PEComa of the IVC.**

Discussion

PEComas are a family of related mesenchymal neoplasms that include angiomyolipoma, lymphangiomyomatosis, clear cell 'sugar' tumour of the lung, and a group of rare, morphologically and immunophenotypically similar lesions arising at a variety of visceral and soft tissue sites[2]. These tumours' all share a distinctive cell type, the perivascular epithelioid cell (PEC). They show a marked female predominance. They are composed of nests and sheets of usually epithelioid but occasionally spindle cells with clear to granular eosinophilic cytoplasm. These tumours' have a focal association with blood vessel walls[2].

They appear to arise most commonly at visceral, retroperitoneal, and abdominopelvic sites, with a subset occurring in somatic soft tissue and skin[2]. Nearly all of them show immunoreactivity for both melanocytic (melan-A and/or HMB-45) and smooth muscle (actin and/or desmin) markers[1]. Although most reported PEComas behave in a benign fashion, malignant tumours have

occasionally been documented[3]. Because PEComas can behave in an aggressive manner, careful follow-up is warranted.

Folpe *et al.*[3] reported PEComa of soft tissue and gynaecological origin in the omentum, uterus and vagina, soft tissues, abdominal wall, etc. The median patient age was 46 years with marked female predominance. The tumours ranged from 1.6 to 29 cm in size. Clinical follow-up of 26 PEComa patients showed 3 local recurrences and 5 distant metastases. They concluded that recurrence and/or metastasis was strongly associated with tumour size greater than the median size of 8 cm, mitotic activity greater than 1/50 high power field, and necrosis.

Kalyanasundaram *et al.*[4] reported 2 such tumours in young female patients involving the urinary bladder and vagina, respectively. Immunohistochemistry was strongly positive for melanocytic markers. The vaginal tumour recurred despite chemotherapy and the bladder tumour was lost to follow-up.

Rigby *et al.*[5] presented a case of 11-year-old girl with 2 large abdominal masses in the left flank and epigastrium and left supraclavicular lymphadenopathy. Subsequent investigations led to the diagnosis of metastatic PEComa arising from the left kidney. The patient was treated with a dacarbazine-based regimen first and subsequently with imagined misplace. The treatment was later discontinued because of non-response.

Guinea *et al.*[6] reported a case of PEComa with a perirenal location.

These case reports highlight the dilemma in treating PEComa as there is not enough literature and evidence regarding this. We are reporting this case to emphasise 3 points: first, tumours arising from the IVC and growing exophytically can mimic an adrenal tumour if the mass is in a suprarenal location. Second, tumours invading IVC are often treated with pessimism about a cure but these tumours may turn out to be potentially benign entities like PEComa as in our case. Third, as the role of adjuvant therapy is questionable based on the failure of chemoradiation in previously reported cases, the best policy in such cases would be to follow-up scrupulously rather than compromising the patient's quality of life by subjecting them to chemotherapy.

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