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CASE REPORT

Retroperitoneal Sclerosing Perivascular Epithelioid Cell Tumour

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ABSTRACT

Perivascular epithelioid cell tumour is a relatively new entity with rising incidence. This is a rare mesenchymal neoplasm that can occur in various organs and is characterised by proliferation of perivascular cells and expression of myomelanocytic markers. Here, we present an asymptomatic 52-year-old female patient with an incidental radiological finding of a large retroperitoneal mass, histologically and immunohistochemically proven to be a sclerosing type of perivascular epithelioid cell tumour. The mass showed typical morphological and microscopic features consistent with those described in the current literature. However, it had computed tomography findings of neovascularisation and hyper-vascularity, not often documented in previous case reports of the sclerosing type of tumours. Literature review, using PubMed, of intraperitoneal / retroperitoneal type of tumours, and specifically the sclerosing type, was performed. To the best of our knowledge, less than 20 sclerosing perivascular epithelioid cell tumours have been reported and few describe the associated radiological features.

Key Words: *Diagnostic imaging; Kidney neoplasms; Perivascular epithelioid cell neoplasms*

中文摘要

腹膜後硬化型血管周上皮樣細胞瘤

杜婉筠、曾佩琪、楊子慧、袁銘強

血管周上皮樣細胞瘤是一種較新的疾病，發病率呈上升趨勢。它是一種罕見的間葉細胞瘤，可以發生在各種器官內，組織學特徵是血管周圍細胞增生和細胞肌黑色素標記物的表達。本文報告一名52歲女性於影像檢查偶然發現一個腹膜後大腫塊，組織學上和免疫組織化學上均顯示為血管周圍上皮樣細胞瘤的一種硬化型。腫塊顯示典型的形態學和鏡下特徵，與以往文獻描述的一致。然而，電腦斷層掃描結果顯示有新生血管和富血管性，這種情況在該腫瘤硬化型的病例報告並不常見。我們利用PubMed進行腹膜內／腹膜後該腫瘤（尤其是硬化型）的文獻回顧。據我們所知，文獻中有關硬化性血管周上皮樣細胞瘤的病例不足二十個，且其中很少相關影像學特徵的描述。

INTRODUCTION

Perivascular epithelioid cell tumours (PEComas) are a new category of tumours defined in the World Health Organization Classification of Tumours since 2002.[1] These are rare mesenchymal neoplasms characterised by proliferation of perivascular cells and expression of

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myo-melanocytic markers.[2]

The PEComa family includes angiomyolipoma (AML), clear-cell sugar tumour (CCST), lymphangiomyomatosis (LAM), clear-cell myomelanocytic tumour (CCMMT) of the falciform ligament and non-specific types occurring in the pelvic cavity, abdominal cavity, digestive tract, genitourinary tract, and the surrounding soft tissues or skin.[3] They show a female predilection and affect middle-aged adults most commonly. Conventional PEComas usually arise from the abdominopelvic and uterine regions. In contrast, the sclerosing subtype, which is characterised by extensive hyalinised stroma, is predominantly found in the retroperitoneum.[4] Some authors suggest that macroscopic fat and hypervascularity might be the radiological features of the conventional PEComa family,[5,6] especially when considering AML as the most common PEComa. To the best of our knowledge, to date, there are less than 20 cases of sclerosing PEComas reported in the English literature and few of them include radiological findings.[4,7,8] Herein, we present the detailed radiological features of a histopathologically proven sclerosing PEComa and a literature review focusing on the radiological features of sclerosing PEComa and intraperitoneal / retroperitoneal PEComa.

CASE REPORT

We present the case of a 52-year-old woman who was followed up by Department of Medicine at Tuen Mun Hospital, Hong Kong, for hypertension. She was asymptomatic all along. Physical examination was unremarkable. Blood results showed renal impairment. Thus, ultrasound of the kidneys was performed.

Ultrasound revealed a large, well-defined echogenic mass in the right lower abdomen closely abutting the kidney and liver. Internal vascularity was noted. No internal fat or calcification was identified (Figure 1).

Further investigation with computed tomography (CT) was subsequently performed. There was a large (8.5 cm) retroperitoneal mass closely abutting the posterior aspect and displacing the right kidney (Figure 2). There was no internal hypodense focus suggestive of macroscopic fat component on precontrast scan. It demonstrated heterogeneous enhancement upon contrast administration. Multiple tortuous branches from the aorta supplied the lesion. Neither kidney showed any focal solid mass. There were no enlarged intra-abdominal lymph nodes. Lung bases were clear. No osseous destruction was seen.

The patient was referred to the urology team and elective excision of the retroperitoneal mass was performed uneventfully. The right kidney was successfully preserved.

Morphologically, the tumour was solid with greyish-yellow surface and tiny cystic changes. Histologically, low-power examination showed that the tumour cell clusters were arranged in a markedly hyalinised

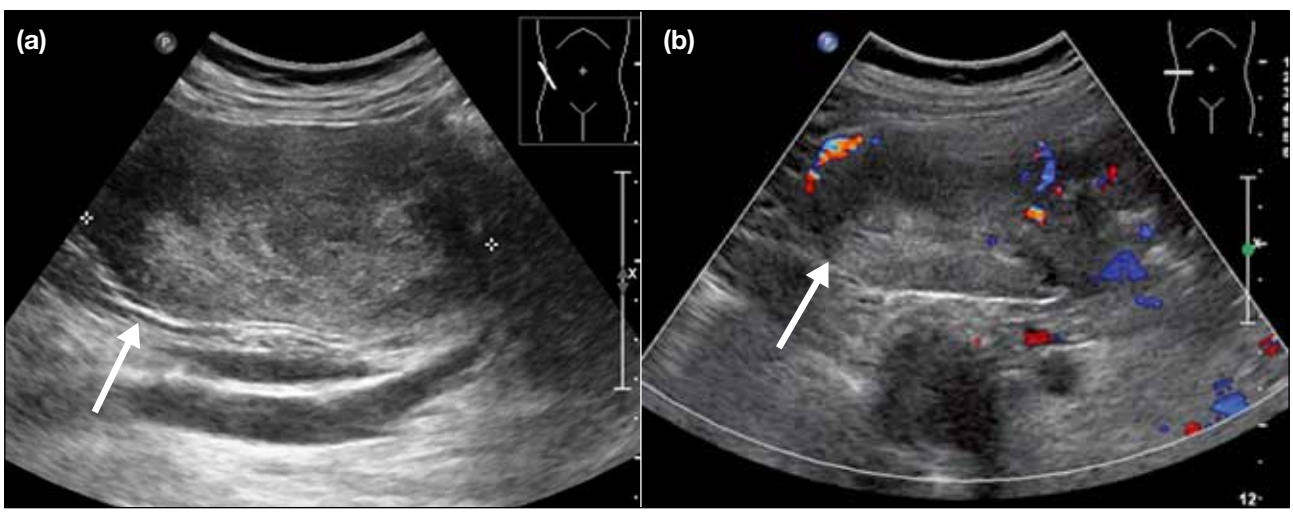

Figure 1. Ultrasound images show (a) a large well-defined echogenic mass in right lower abdomen closely abutting the kidney (arrow). No internal fat or calcification was identified; (b) internal vascularity was noted (arrow).

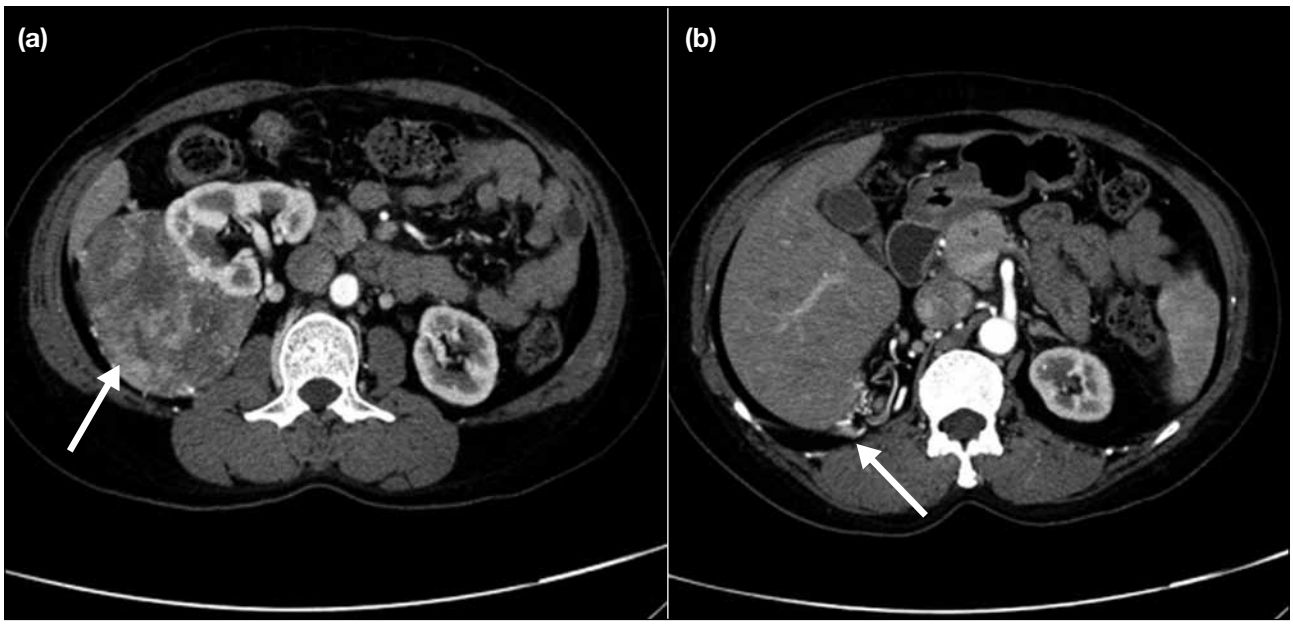

Figure 2. Computed tomography of the abdomen with contrast. (a) A large heterogeneous, enhancing retroperitoneal mass (arrow) is found closely abutting the posterior aspect and displacing the right kidney. (b) The lesion (arrow) was supplied by multiple tortuous branches from the aorta. Neither kidney shows any focal solid mass. There are no enlarged intra-abdominal lymph nodes.

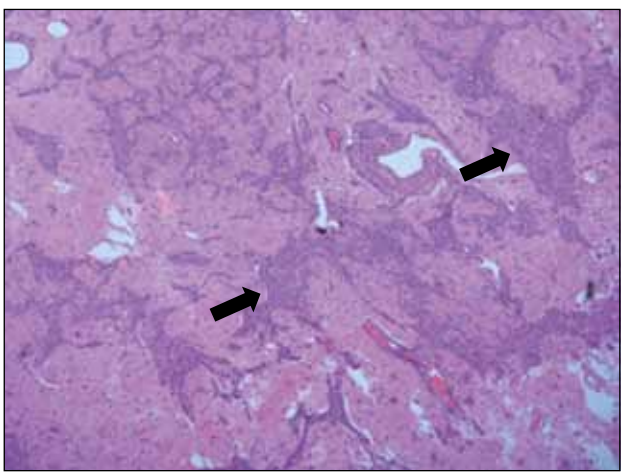

Figure 3. Low-power examination of a photomicrograph of the tumour. Tumour cell clusters (arrows) are arranged in an extensive sclerotic stroma (H&E; original magnification, x 4).

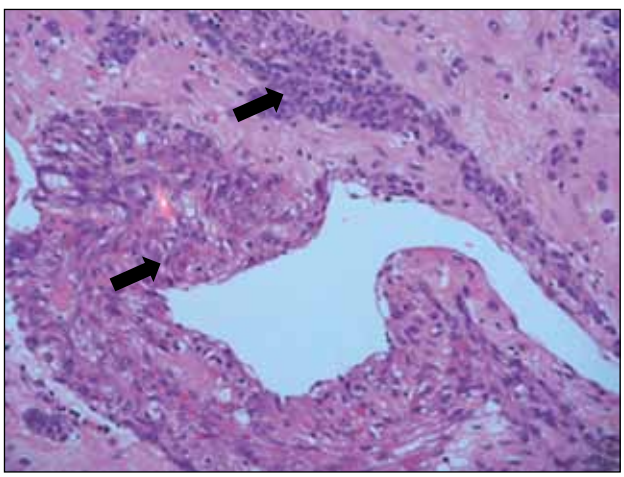

Figure 4. High-power examination of a photomicrograph of the tumour. Tumour cells are spindle-shaped (arrows) with moderate amount of lightly eosinophilic or vacuolated cytoplasm (H&E; original magnification, x 200).

sclerotic stroma (Figure 3). High-power examination showed that the tumour cells were spindle-shaped with moderate amount of lightly eosinophilic or vacuolated cytoplasm (Figure 4). Immunohistochemically, the tumour cells were positive for HMB-45, melan-A, actin, desmin, and calponin.

Follow-up CT scans done 9 months and 23 months after the operation showed no signs of residual tumour or recurrence. No lung changes or bone metastases were detected. Clinically, the patient remained symptom free.

DISCUSSION

PEComa is a relatively rare and newly discovered group of mesenchymal neoplasms sharing distinct morphological, histological, and immunohistochemical

features.[2] The PEComas are characterised by epithelioid cells and a close relationship with blood vessels. They are consistently immunoreactive for the melanocytic marker HMB45, and are variably immunoreactive to smooth muscle actin. They are negative for epithelial markers.[9]

The PEComa family includes AML, CCST, LAM, CCMMT, and non-specific types occurring in the pelvic cavity, abdominal cavity, digestive tract, genitourinary tract, and the surrounding soft tissues or skin.[3]

The sclerosing PEComa subtype was recently described as a distinctive variant, which shows markedly hyalinised stroma. It has a female predominance and peaks in middle-aged adults, similar to the conventional PEComa. However, most sclerosing PEComas arise in the retroperitoneum, while the conventional type most

Table 1. Literature review on radiological features of intraperitoneal / retroperitoneal PEComa.[6,7,10-12]

Study	Site (No. of cases)	Histological subtype if specified	Macroscopic fat element	Enhancement	Hyper-vascularity / dilated vessels	Other radiological description	Remarks
Tirumani et al,[10] 2014	Retroperitoneum (14), female genital tract (10), intraperitoneal (6), lower extremity (3), mediastinum (1), unknown primary (2)	Malignant PEComa	1/36 (in PEComa associated with AML in one patient)	Yes (CT: all enhanced except one with predominate necrosis and all case in MRI)	NA	CT: all except one were well-circumscribed CT: 7/13 necrosis MRI: 3/7 haemorrhage USG: 3/5 well-circumscribed	CT (n=13), MRI (n=7), USG (n=5)
Rekhi et al,[7] 2012	Retroperitoneal soft tissue (1)	Sclerosing PEComa	NA	NA	NA	Well-defined, round, hypodense mass	
Rasalkar et al,[11] 2011	Kidney (1)	Malignant pigmented clear cell PEComa	NA	Yes	NA	Ill-defined, exophytic, partly calcified mass	Liver metastasis
Fang et al,[12] 2007	Liver (2)	(Case 1) PEComa	NA	Yes	NA	Well-demarcated mass with striking enhancement on portal phase than arterial phase	
		(Case 2) PEComa	NA	Yes	NA	Well-demarcated mass with arterial enhancement and hypodense on portal phase	
Prasad et al,[6] 2007	Renal PEComa	Classic AML	Yes	Yes	Dysmorphic vessels, aneurysm formation	Intense enhancement	
		Monotypic epithelioid AML	NA	NA	NA		
		LAM of renal sinus	NA	NA	NA		
	Urinary bladder and prostate	PEComa	NA	NA	NA	Well-circumscribed, expansile or infiltrative soft tissue mass	
	Liver	AML	50%	Yes	NA	Early arterial enhancement	
	Falciform ligament/ ligamentum teres	CCMMT	NA	NA	NA	NA	
	Pancreas	CCST	NA	NA	NA	Well-circumscribed soft tissue mass	
	GI tract (most commonly colon)	Classic AML or epithelioid AML	NA	NA	NA	Well-circumscribed mass, may cause intestinal obstruction	
	Spleen	AML	NA	Yes	Yes, haemorrhage	Progressive centripetal contrast enhancement	
	Retroperitoneal (perinephric)	PEComa	Yes	NA	NA	Well-circumscribed soft tissue mass	
	Peritoneal (omentum, mesentery)	PEComa	NA	NA	NA	Well-circumscribed soft tissue mass	
	Adrenal	PEComa	Yes	NA	NA	NA	
	Uterus	PEComa	NA	NA	NA	Lobulated heterogeneous soft tissue mass with necrosis and haemorrhage	

Abbreviations: AML = angiomyolipoma; CCMMT = clear-cell myomelanocytic tumour; CCST = clear-cell sugar tumour; CT = computed tomography; GI = gastrointestinal; LAM = lymphangiomyomatosis; MRI = magnetic resonance imaging; NA = not available; PEComa = perivascular epithelioid cell tumour; USG = ultrasonography.

commonly arises from the abdominopelvic and uterine sites.[4]

There is, as yet, no consensus on the diagnostic criteria for the benign or malignant type, as its clinical and radiological behaviour is still not well documented. One of the currently accepted classifications for malignant PEComas was proposed by Folpe in 2002.[1] They proposed that a tumour size greater than 5 cm, an infiltrative growth pattern, high nuclear grade, necrosis, mitotic activity of >1/50 high-power fields, and aggressive clinical behaviour suggest a more malignant pathology. In our case, the size was greater than 5 cm and had high mitotic activity but did not have features of invasion or significant clinical symptoms. Thus, our case had mixed characteristics, typically demonstrating the difficulty in predicting the biological course and prognosis of this tumour.

The imaging features of intraperitoneal / retroperitoneal PEComa family other than AML among English radiology literature are relatively scarce. Most intraperitoneal / retroperitoneal PEComas reported in the previous literature are well-circumscribed enhancing mass lesions. A few of these lesions were shown to be hypervascular, pathologically or radiologically. Some authors suggested that macroscopic fat and hypervascularity might be the radiological features of the conventional PEComa family,[5,6] especially when considering AML as the most common PEComa. A few of the recent case reports, including ours, do not show macroscopic fat on CT (Table 1[6,7,10-12]).

To the best of our knowledge, to date, there are less than 20 reported cases of sclerosing PEComa in the English literature and few of them include radiological findings.[4,7,8] None of the reports mentioned macroscopic fat or hypervascularity as a significant feature in the sclerosing type of the tumour (Table 2[4,7,8,13]).

Consistent with previous case reports on retroperitoneal PEComa, our patient was typically asymptomatic, despite the large tumour size. This could be related to the well-circumscribed nature of the mass with lack of invasion of the adjacent organs.

Radiologically, our case showed neovascularisation by multiple tortuous branches from the aorta. Perhaps in future practice, this new rare entity of sclerosing PEComa may also be considered in the differential diagnosis of hypervascular retroperitoneal masses in addition to hemangiopericytomas, sarcomas, or lymphoma. However, differentiation between these differentials can be difficult as they all typically present as large, well-encapsulated, retroperitoneal masses. Since only around 20 cases of sclerosing PEComa have been reported, it remains challenging to identify the typical imaging features of this disease.

Morphologically, our specimen was a greyish-yellow, retroperitoneal, solid mass with tiny cystic changes. Histologically, the tumour showed intimate association

Table 2. Literature review on radiological features of sclerosing PEComa.[4,7,8,13]

Study	Site (No. of cases)	Histological subtype if specified	Macroscopic fat element	Enhancement	Hypervascularity / dilated vessels	Other radiological description	Remarks
Leão et al,[8] 2013	Pararenal (1)	Sclerosing PEComa	NA	Slight enhancement	NA	Round-to-oval mass	
Rekhi et al,[7] 2012	Retroperitoneal soft tissue (1)	Sclerosing PEComa	NA	NA	NA	Well-defined round hypodense mass	
Yamada et al,[13] 2011	Female genital organ (2)	(Case 1) Sclerosing PEComa	NA	NA	NA	Well-circumscribed mass	
		(Case 2) Sclerosing PEComa	NA	NA	NA	NA	
Hornick and Fletcher,[4] 2008	Retroperitoneal soft tissue (10)	Sclerosing PEComa	NA	NA	NA	NA	
	Abdominal wall (1)	Sclerosing PEComa	NA	NA	NA	NA	
	Pelvis (1)	Malignant sclerosing PEComa	NA	NA	NA	NA	Metastasis
	Uterus (1)	Sclerosing PEComa	NA	NA	NA	NA	

Abbreviations: NA = not available; PEComa = perivascular epithelioid cell tumour.

with the vessel wall and extensive hyalinised stroma, which is characteristic of the sclerosing type of PEComa.[4] Our case, agreeing with cases in the previous literature, also showed immunoreactivity to HMB45, which is one of the unique features of this disease.[2-4,9,14]

Surgery is currently the mainstay of treatment, as chemotherapy and radiotherapy have not shown significant results. The majority of PEComas reported in literature are benign with good prognosis.[3,9,15] Our patient remained disease-free for at least 2 years, also suggesting a benign course.

In conclusion, we present a rare case of a histopathologically proven, retroperitoneal, sclerosing PEComa, with radiological findings of neovascularisation and hypervascularity, which are less emphasised in previous case reports. To the best of our knowledge, less than 20 cases of sclerosing PEComa have been reported in the English literature to date, and few of them include radiological findings. The radiological findings of neovascularisation and hypervascularity propose the possibility of adding sclerosing PEComa to the list of hypervascular retroperitoneal masses. Moreover, there was no macroscopic fat identified on CT in our case, which some authors believe to be a characteristic imaging feature in the conventional PEComa family. Due to the rarity of the disease, the distinct imaging findings are yet to be established.

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