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ORIGINAL PAPERS

Retroperitoneal Precaval Pecoma – an Incidental Finding

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Abstract

PEComas are rare mesenchymal tumours of histologically and immunohistochemically distinctive perivascular epithelioid cells. Most of these tumours are benign, but in malignancies the prognosis is usually very poor. The clinical presentation of this tumours is nonspecific and the imagistic examination is usually not sufficiently sensitive to establish a diagnosis. The best diagnostic method is histopathological examination combined with immunohistochemical examination. Treatment of this tumours is surgery followed by chemotherapy (in case of malignancy). We present the case of a 43-year-old female patient who presented for gynaecological symptoms, and was discovered incidentally during a CT scan with a retroperitoneal precaval mass. The tumour was surgically excised and the histological and immunohistochemical examination led to the diagnosis of PEComa with benign characteristics.

Keywords: PEComa, caval, mesenchymal, excision

Rezumat

PECoamele sunt tumori mezenchimale rare formate din celule epiteloide perivasculare distincte din punct de vedere histologic și imunohistochimic. Majoritatea acestor tumori sunt benigne, dar cazurile maligne au de obicei un prognostic slab. Prezentarea clinică a acestor tumori este nespecifică și imagistica nu este de obicei destul de sensibilă pentru a stabili diagnosticul. Principala metodă de diagnostic este reprezentată de examenul histopatologic combinat cu cel imunohistochimic. Tratamentul acestor tumori este reprezentat de chirurgie, urmată de chimioterapie (în cazurile maligne). Prezentăm cazul unei paciente în vârstă de 43 de ani, ce s-a adresat pentru simptome ginecologice și a fost descoperită incidental in timpul unui CT cu o masă retroperitoneală precavă. Tumora a fost excizată chirurgical și examenul histologic și imunohistochimic au pus diagnosticul de PECom cu caractere de benignitate.

Cuvinte cheie: PECoame, mezenchimatos, excizie, cav

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INTRODUCTION

Perivascular epithelioid cell tumours (PEComas) were initially described as "sugar tumours" of the lung and renal angiomyolipoma's by Bonetti; ten years later, in 2002, World Health Organisation Classification of Tumours defined them as "mesenchymal tumours composed of histologically and immunohistochemically distinctive perivascular epithelioid cells.[1] This group of tumours include angiomyolipoma (AML), clear cell "sugar tumours" of the lung, pancreas, and uterus, and lymphoangioleiomyomatosis (LAM).[2] PEComas-NOS (not otherwise specified), which represent uncommon tumours located in the retroperitoneum, abdominal cavity, digestive tract, skin, soft tissue, and bones sharing similar morphological and immunophenotypically characteristics, are also included in the PEComa group.[3]

CASE PRESENTATION

This is the case of a 43-year-old female patient, who presented with abdominal pain and menometrorrhagia, upon clinical examination and evaluation by a gynaecologist a CT scan of the abdomen and pelvis was recommended. The radiologic assessment revealed multiple uterine leiomyomas and a mixed cystic-solid retroperitoneal mass, located anterior to the vena cava measuring 22/38/50 mm.

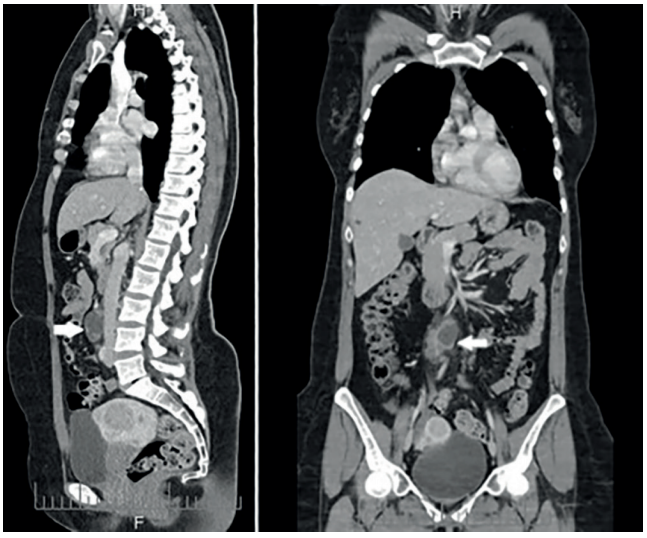

Figure 1. CT images of precaval retroperitoneal PEComa (sagittal and frontal slides)

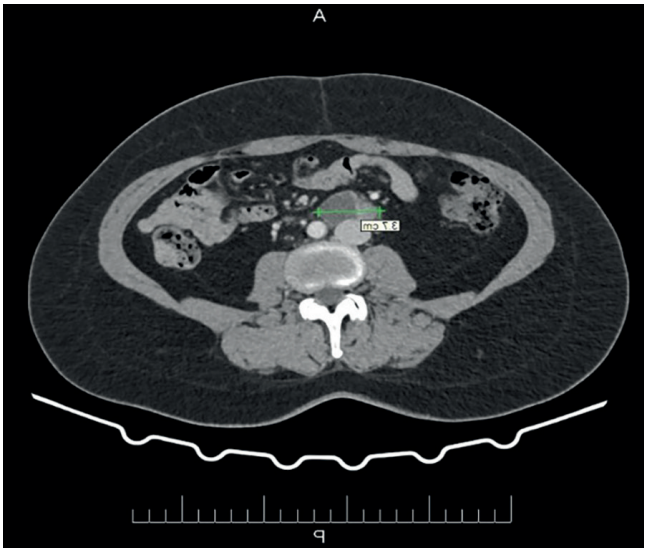

Figure 2. CT images of precaval retroperitoneal PEComa (transversal slide)

The patient underwent surgical intervention, excision of the retroperitoneal tumour and total hysterectomy were performed. The postoperative evolution of the patient was favourable with discharge in the 5th postoperative day.

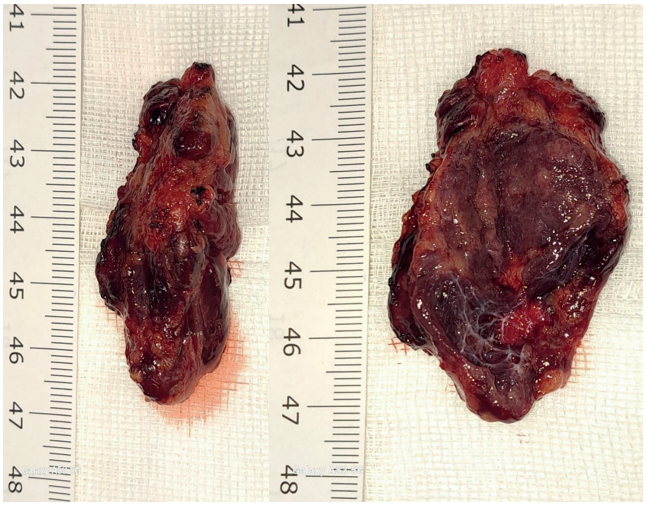

Figure 3. Retroperitoneal PEComa postexcision

The histopathological and immunohistochemical examination of the retroperitoneal tumour revealed a mesenchymal neoplastic proliferation consisting of fusiform and epithelioid cells, with a rich vascular network showing smooth muscle differentiation (SMA

positive immunophenotype), rich vascular supply (CD34 positive immunophenotype), and positivity for melanocytic markers (HMB45, MelanA), in favour of a retroperitoneal PEComa with benign histological and immunohistochemical features. Histopathological examination also confirmed the conventional type of uterine leyomiomas described on the CT scan.

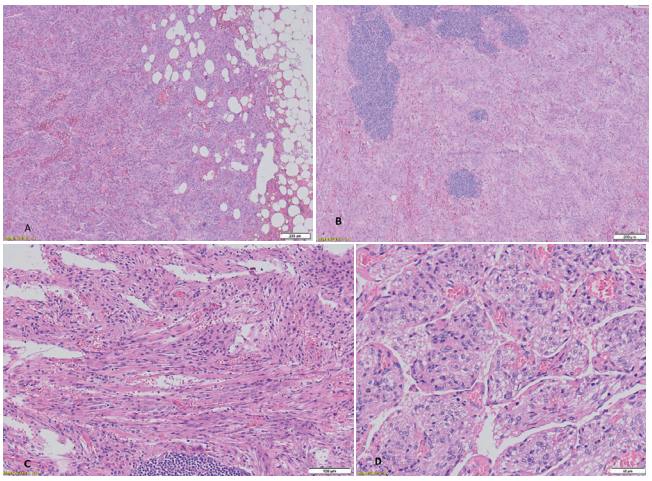

Figure 4: A-H&E slide showing a mesenchymal proliferation with a fascicular, storiform or nested growth pattern, with proeminent arborising vasculature, admixed at periphery with mature adipose tissue. B-There was associated focal inflammation, with frequent lymphoid aggregates and a lymphangioleiomyomatosis-like pattern. On higher magnification, the proliferation is composed of spindle (C) and epithelioid (D) plump cells, with clear to eosinophilic cytoplasm and vesicular slightly irregular nuclei, with inconspicuous nucleoli. There are numerous delicate congested vessels forming arches or having staghorn appearance, with a perivascular condensation of tumour cells.

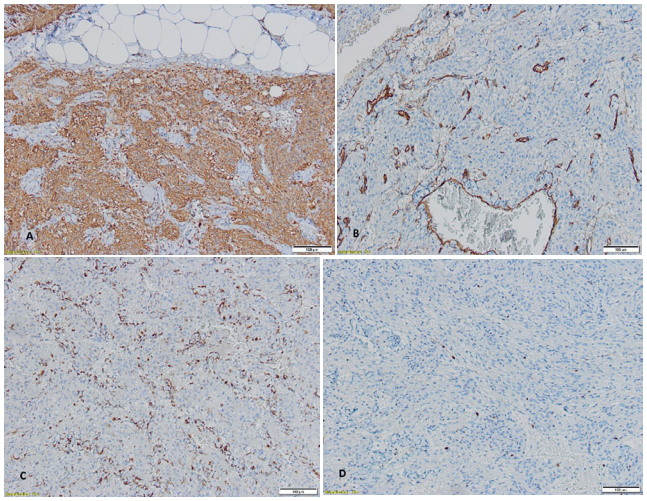

Figure 5: A -IHC for Alpha -smooth muscle actin (SMA) showing diffuse and strong cytoplasmic staining in the mesenchymal cells; B-IHC for CD34 highlighting the prominent vasculature, with negative proliferating cells; C- HMB45 was positive showing intense staining in perivascular cells, diffuse in the mesenchymal proliferation; D- Ki-67 was low, with nuclear positivity in 1% of tumour cells.

DISCUSSION

PEComas are a rare group of mesenchymal tumours. The majority of these tumours are AML or LAM, both of which are typically associated with a prior diagnosis of tuberous sclerosis, this association is not observed in other forms of PEComas.[4] Most of these tumours are benign, with a higher incidence in females, and have a nonspecific clinical presentation, thus making the diagnosis difficult. This issue can lead to locally disseminated or metastasised disease when diagnosing malignant cases.[5] In our case, the patient was female and the tumour was incidentally discovered during a CT scan performed for the evaluation of a gynaecological disease.

Radiological descriptions of PEComas are very variable. Some reports suggested that arteriovenous hypervascularity in contrast-enhanced CT images is a feature of PEComa; most lesions being slightly hypodense in the delayed phase[6]. Similar to other reports, preoperative CT was not sufficiently sensitive to establish the diagnostic in our case.

In the differential diagnosis of PEComas are included neoplasms like melanoma, clear cell sarcoma, gastrointestinal stromal tumours (which can develop in the retroperitoneum) and true smooth muscle cell tumours, therefore immunohistochemical testing is

extremely important for making the correct diagnosis.[7,8] In this case, immunohistochemical examination along with histological examination established the diagnosis of Pecoma with benign characteristics.

Immunohistochemically PEComas are tumours expressing melanocyte and muscle cell markers with various levels of intensity. The most sensitive markers are desmin, HMB-45 and SMA; Melan-A and S-100 being less commonly expressed.[9] In our case, SMA had an intense positive cytoplasmic expression, HMB-45 had a high and medium intensity positive cytoplasmic expression with a focal Melan -A positivity, and S-100 negative tumour cells.

Even though most PEComas are benign, those who are malignant have a poor prognosis. Some reports suggested a set of criteria for their classification, such as: diameter of the tumour, necrosis, vascular invasion, infiltrative growth pattern, nuclear atypia and high mitotic rate.[9] In our case, the diameter of the tumour was less than 5 cm, there was no necrosis or vascular invasion, no infiltrative growth, and mild nuclear atypia, all of which are characteristics of benign tumour.

The standard approach for PEComas is surgery. If the histopathological examination shows worrying features, suggestive of malignancy then chemotherapy (sometimes along with radiotherapy) can be performed after surgery.[10] In this case the tumour had benign characteristics, therefore a complete surgical excision with negative margins was the only recommended treatment according to current guidelines. The patient underwent CT examination 6 months after surgery with no imagistic signs of recurrence.

CONCLUSION

PEComas are a rare group of tumours with unspecific clinical and imagistic characteristics. Diagnosis is based on histological examination correlated with the immunohistochemical expression of smooth muscle and melanocytic markers. Establishing the risk of malignancy is important because of the aggressive evolution of the malignant counterparts. Imaging, histological, and immunohistochemical correlations, as well as treatment strategies, should be further developed for a better characterisation of the difficulty to diagnose tumours with aggressive potential.

The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from the patient included in the study. There are no conflicts of interest regarding this article.

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