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CELL TUMORS VASCULAR EPITHELIOIDS (PECOMES): EXPERIENCES OF ANATOMICAL PATHOLOGY LABORATORY OF THE MILITARY HOSPITAL MOULAY ISMAIL OF MEKNES (ABOUT 4 CASES)

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INTRODUCTION

PEComas are defined by the World Health Organization (WHO) as "mesenchymal tumors composed of histologically and immunohistochemically distinct perivascular epithelioid cells.[1] These tumors belong to a heterogeneous family including angiomyolipoma (AML), lymphangioleiomyomatosis (LAM), clear cell myomelanocytic tumor of the falciform ligament / ligamentum teres (CCMT), clear cell tumor of the lung (CCST) and a subgroup called PEComa-NOS.[2] The aim of this work is to review the epidemiological, clinical, anathomopathological, therapeutic and evolutionary characteristics of PEComas, based on the analysis of a retrospective series of 04 cases of PEComas managed in the pathological anatomy department of the Moulay Ismail military hospital in Meknes over a period of 09 years, from January 2014 to December 2022, with emphasis on our own experience in the management of this type of tumour, in comparison with data from the literature.

MATERIALS AND METHODS

I. **Resources:** Our study focuses on a series of adult PEComa cases (age ≥ 16 years old) comprising 04 patients managed in the pathological anatomy department of the Moulay Ismail military hospital in Meknes and the pathological anatomy department of the Mohammed V military hospital in Rabat, over a 9-year period, extending from January 2014 to December 2022.

II. **Methods:** This is a retrospective, descriptive analysis of the medical records selected for our study, based on the analysis of various anamnestic, clinical, paraclinical, therapeutic and evolutionary parameters, with emphasis on our own experience in comparison with the data in the literature.

- **Inclusion criteria:** We included in this series patients with perivascular epithelioid cell tumors operated on at the Moulay Ismail military hospital in Meknes or the Mohammed V military hospital in Rabat, with histological confirmation.

- **Exclusion criteria:** This series does not include.
- Incomplete files.
- Non-operated patients.
- Without pathological confirmation.

RESULTS

EPIDEMIOLOGICAL DATA

a. **Frequency:** We collected 04 cases of adult PE Comas (age ≥ 16 years old) among 09 cases of PE Comas managed in the pathological anatomy department of the Moulay Ismail military hospital in Meknes, over a period of 9 years, i.e. a frequency of 44.44%.
b. **Age:** The age of the study population ranged from 45 to 55.
c. **Sex:** In the 04 cases discussed, we noted a predominance of women:
d. **Distribution by location:** The most frequent location of PE Comas was the skin, with two observed cases, representing 50% of cases. The other two locations were in the uterus and kidney respectively.

CLINICAL DATA

1. **Diagnostic delay:** The delay ranged from 3 months to 18 months, while 50% of our patients consulted more than 6 months later.

Table 1: Diagnostic delay for the various patients in our series.

Diagnosis time	Number of cases	Percentage
<6 months	2	50%
>6 months	2	50%

2. CLINIC

2.1. Functional signs

2.1.1. Cutaneous PEComa Functional signs reported by both patients included papular swelling of the forearm and earlobe, respectively. One patient also reported itching and tingling around the lesion.

2.1.2. Renal PEComa: The reason for consultation was pain in the right flank associated with the presence of haematuria.

2.1.3. Uterine PEComa: Our patient was admitted with abnormal uterine bleeding and abdominal pain. The functional signs are summarized in the following table.

Table 2: Functional signs of patients monitored.

Functional signs	Number of cases	Percentage
Papular swelling	2	50%
Abdominal/lumbar pain	2	50%
Hematuria	1	25%
Abnormal uterine bleeding	1	25%

I. PARACLINICAL DATA

1. **Abdominopelvic ultrasound:** Abdominal ultrasonography was performed as 1st-line treatment in 2 cases in our series. The results were as follows.

- The presence of a heterogeneous, hyperechoic uterine mass in the case of uterine PEComa.
- The presence of a heterogeneous, hyper-echoic right lumbar mass in the case of renal pecoma.

2. **COMPUTED TOMOGRAPHY:** Abdominal-pelvic CT revealed the location, size and scanographic appearance of two cases in our series. The CT appearance of the tumors was generally characterized by hypodensity, significant heterogeneous enhancement on the arterial and venous phases, and mild hypodensity at late time. The examination also confirmed the absence of locoregional extension in all cases in our series (100% of cases).

3. **MAGNETIC RESONANCE IMAGING:** Abdominal-pelvic MRI was performed in 2 patients (50% of cases).

- Unenhanced MRI revealed hypointense T1-weighted masses and hyperintense T2-weighted masses.
- The uterine PEComa was characterized on T2-weighted images by a heterogeneous tumor with hypointense and hyperintense areas with irregular, blurred margins.
- No lymph nodes or distant metastases.

II. Pathological features

1. **Biopsy specimens:** 1 patient with uterine localization underwent ultrasound-guided biopsy prior to treatment. The 3 other patients were diagnosed after tumor resection. Examination of the surgical specimens yielded the following results.
2. **Macroscopic aspects:** The macroscopic characteristics of the samples were as follows.

- Tumor mass sizes ranged from 1 cm to 4.5 cm at their largest.
- The weight of tumor masses was not specified in the available data.

3. **Microscopic aspects:** Microscopic examination of the samples revealed the following histological features.

- **Tumor architecture:** The tumors were composed of epithelioid cells arranged in clusters or nests. Some tumors showed a vascular arrangement characterized by irregular blood vessels surrounding clusters of tumor cells.
- **Cellular characteristics:** Tumor cells were epithelioid in shape, with abundant eosinophilic cytoplasm. Nuclei varied in size and shape, from small to large, with an often regular nuclear morphology.
- **Nuclear features:** Tumor cell nuclei showed fine, granular chromatin.

4. **Immunohistochemistry:** Tumor cells were positive for PECome-specific markers such as HMB-45, melan-A and SMA. Markers such as PANCK, desmin, S-100, melan-A, EMA, c-kit and CD10 were negative in tumor cells. These histological and immunohistochemical features are consistent with the diagnosis of PEComa.

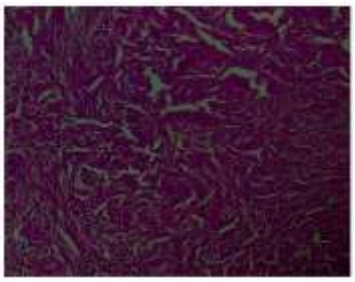

Figure 1. Microscopic Aspect of a Pecome Showing epithelioid cells with slightly clear eosinophilic cytoplasm, ovoid nuclei and small nucleoli.

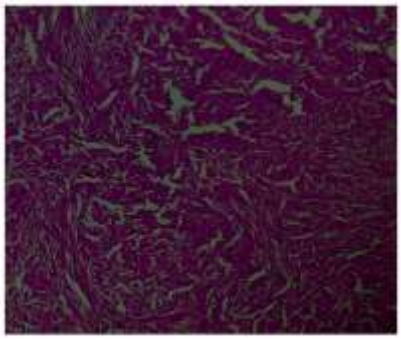

Figure 2. Microscopic aspect of a pecome.

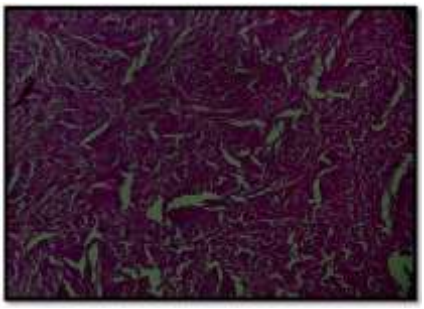

Figure 3: Iconography of the pathological anatomy laboratory at moulay ismail military hospital of meknes.

Table 3. Showing the macroscopic, microscopic and immunohistochemical characteristics of the 04 cases in our series.

Location	Macroscopy	Microscopy	Immunohistochemistry
Cutaneous (forearm)	1 cm skin lesion, dark brown papule	Non-encapsulated intradermal tumor, clear epithelioid cells	HMB45+, Melan-A-, S100+, HMB45, Melan-A-, S100+.
Cutaneous (earlobe)	Skin lesion measuring 1.8 x 0.9 cm, whitish-yellow nodular surface	Well-defined nodule with fibrous pseudocapsule	HMB45+, Melan-A+, SMA+, SMA+.
Renal	4.5 cm x 4 cm, crumbly beige kidney lesion	Triphasic tumor proliferation: predominantly adipose, vascular and smooth muscle, with a muscle component arising from the vascular wall	HMB45+
Uterine	Uterine tumor, 1.5 x 1.0 cm, bumpy surface	Epithelioid component with focal infiltration, absence of pleomorphism and low mitotic rate	HMB45+, SMA+, SMA

TREATMENT

1. **Surgical treatment:** As the location of PEComas is highly variable, the nature of the surgical procedure (approach and surgical technique) naturally depends on the organ affected. 3 cases underwent surgery with histological confirmation, and in one case the diagnosis of PEComa was established after tumor resection. The 04 patients underwent complete excision surgery.
2. **Adjuvant treatment:** Only 1 patient with uterine PEComa received neoadjuvant radiotherapy after excision surgery.

Table 4: Clinical features, location and associated treatment.

Patient	Location	Gender	Age	Surgical margins	Adjuvant radiotherapy
1	Cutaneous	Woman	50	R0	----
2	Uterus	Woman	45	R0	5GY x 5GY
3	Renal	Men	49	R0	----
4	Cutaneous	Woman	55	R0	----

DISCUSSION

PEComas, a heterogeneous group of soft tissue sarcomas (STS), are extremely rare.their incidence is estimated at 0.3 cases per 1,000,000 people.[3] The prevalence of malignant PEComas is even lower, at 0.12 cases per 1,000,000 people, or 0.24 per 1,000,000.[4] PEComas can develop in any part of the body. The most common form of PEComa is angiomyolipoma, which has a prevalence of 0.2% in healthy adults.[5]

A study included 18 patients with PEComes who were admitted to Zhongshan Hospital and Xuhui District Central Hospital in China from January 2006 to October 2018. These patientswere diagnosed on the basis of pathological findings and treated by surgical resection or medication. Over a period of 12.83 years (or approximately 1.4 patients per year), the frequency of these specific cases of radically treated PEComas was observed.[6]

The age of onset of PEComa can vary depending on various factors, such as the specific type of PEComa and tumor location.[7] In a retrospective study of 114 cases of gynecological PEComa between 1997 and 2017, patient age ranged from 9 to 79 years, with peak incidence observed around the forties (median age 43.5 years).[8] In

the study conducted at Zhongshan Hospital and Xuhui District Central Hospital in China, the age range of PEComa patients was 23 to 53 years, with a median age of 38 years.[6] In the 234 cases of PEComa-NOS reported in the US literature, patients had an age range of 3 to 97 years (median age 43 years).[9] The most common sites for the development of PEComas included the uterus, skin, liver and colon.[10] Occasionally, locations were not specified, including the abdomen[11], buttocks[12], pelvis[12] or abdominal wall.[11] Furthermore, large malignant PEComas are mainly diagnosed in the extraperitoneal space.[13] Bonetti et al.[14] first described this rare tumour type in 1992, and Zamboni.[15] et al subsequently coined the term "PEComa" in 1996.The World Health Organization recognized the PEComa as a distinct tumour entityteen 2002.[16]

Perivascular epithelioid cells have no "normal" cellular equivalent in the body. Several hypotheses have been put forward: one suggests a possible origin of PEComes from the neural crest, which also gives rise to the pericytes and smooth muscle cells of facial and forebrain blood vessels.[17] Some researchers have supported this hypothesis based on studies of neural crest precursors, which have shown their ability to differentiate into smooth muscle cells[18] and adipocytes.[19] in vitro. However, it should be noted that some researchers disagree with this hypothesis due to the lack of expression of S-100, a marker specific to neural crest cells,in PEComes. Fukunaga hypothesized that the original mesenchymal cells might have the capacity to differentiate into smooth muscle cells and HMB45-positive epithelioid cells, which would explain the different degrees of differentiation observed in different PEComa cases.[20]

It is also possible to speculate that PEComas are derived from genetically modified mesenchymal stem cells of perivascular origin, which could explain their particular phenotype and behavior.[21] But due to the lack of in-depth knowledge of the specific molecular and cellular mechanisms involved in PEComas, the origin of these tumors remains undetermined in many cases.

Molecularly, PEComas are associated with mutations in the TSC1 or TSC2 genes, which are implicated in the pathophysiology of tuberous sclerosis. These genes encode principles that indirectly inhibit the mTOR pathway.[22]

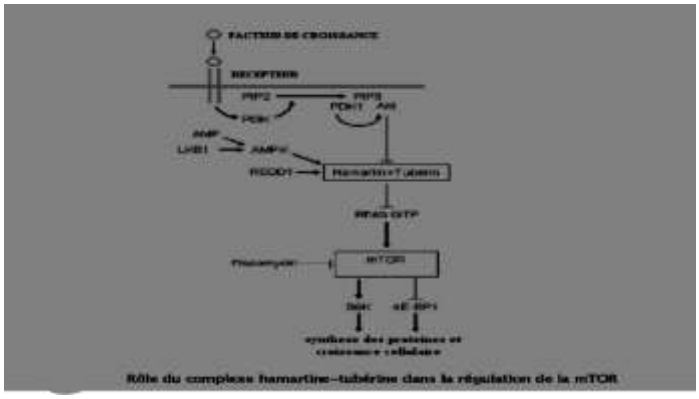

Figure 4: Role of the hamartine-tuberine complex in mTor(175) regulation.

Translocations of TFE3 often play a considerable role in PEComes[23], Transcription factor E3 (TFE3), also known as immunoglobulin light chain activator 3, belongs to the MITF (microphthalmia-associated transcription factor) family, which also includes transcription factor EB (TFEB), transcription factor EC (TFEC) andMITF.[24]

The MITF family regulates cell differentiation and is involved in the regulation of tumorigenesis.[25] The theory suggests that tumors with TFE3 rearrangements show no genetic alterations in either the TSC1 or TSC2 genes.[26]

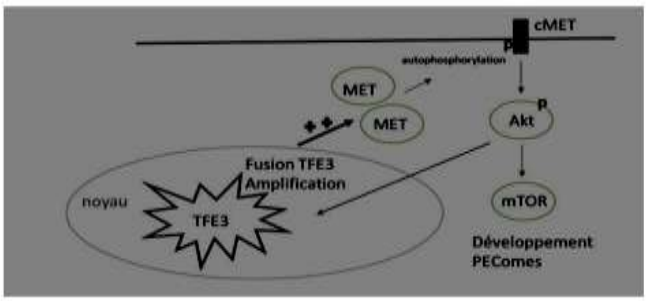

Figure 5: Development of PEComes following translocation of the TFE3 gene according to Utpatel and Tsuda (62,161).

Other translocations have been reported apart from the TFE3 translocation and the TSC1 -TSC2 alteration. Indeed, a case of aggressive PEComa with ATRX mutation has been described.[26]

Among the rare gene rearrangements described in PEComes are two cases ofRAD51B fusion with RRAGB/OPHN1 in a uterine PEComa, as well as two cases of HTR4-ST3GAL1 andRASSF1-PDZRN3 fusion.[20] In addition, there are two cases of RAD51B fusion with RRAGB/OPHN1 in a uterine PEComa, as well as two cases of HTR4-ST3GAL1 andRASSF1-PDZRN3 fusion.

Pathological anatomy establishes the diagnosis of a PEComa, specifies its histological type, and establishes its histopronostic grade. The World Health Organization (WHO) classification describes PEComas as mesenchymal tumors composed of histologically and immunohistochemically distinct perivascular epithelioid cells.[16]

This big family includes.

- angiomyolipomas (AML),
- lymphangioleiomyomatosis (LAM),
- Clear cell tumors of the lung or "sugar tumors" (CCST),
- clear-cell myomelanocytic tumors of the falciform ligament (CCMT)
- and PEComes NOS (no further details)

PEComas are rare tumors with numerous differential diagnoses of varying complexity. These include melanoma, clear cell sarcoma, alveolar soft tissue sarcoma, gastrointestinal stromal tumor (GIST), leiomyosarcoma and renal cell carcinoma with TFE3 translocation. In some cases, paraganglioma, adrenocortical carcinoma and hepatocellular carcinoma may also be discussed as possible differential diagnoses.[3] Neoadjuvant and adjuvant treatments have not yet achieved a well-defined level of efficacy, but they are used selectively according to the specific needs of each patient.

- **Chemotherapy:** The efficacy of chemotherapy in the treatment of PEComas is generally limite.[26] According to the literature, chemotherapy is administered to patients with large PEComas who have not presented with a relapse within 6, 11 or 24 months.[12]
- **Radiotherapy:** Radiotherapy has been used in some cases of perivascular epithelioid cell tumors (PEComa) as an adjunct to surgery, either neoadjuvant or adjuvant.
- **mTOR inhibitors:** In the case of PEComas, mTOR inhibitors have shown some efficacy in treating the disease. Even in the absence of TSC1-TSC2 mutations, mTOR inhibitors can induce a positive response, although this may seem counterintuitive.[27,28]
- **VEGFR pathway inhibitor:** VEGFR inhibitors are drugs that block the VEGFR receptor and are used in the treatment of PEComa to target angiogenesis and reduce the blood supply to the tumor.

CONCLUSION

Perivascular epithelioid cell tumors (PEComes) are rare mesenchymal tumors composed of perivascular epithelioid cells with distinct biphenotypic differentiation, both melanocytic and smooth muscle.[3] These tumors can develop in various organs and tissues, including the kidneys, genital tract, retroperitoneum and lungs.[6] There is no specific clinical or radiological presentation for PEComas, and diagnosis relies mainly on anatomopathological examination with complementary immunohistochemical and molecular analyses.[29]

LIST OF ABBREVIATIONS

PEComa: perivascular epithelial cell tumor
AML: Angiomyolipoma
LAM: Lyphmangioleiomyomatosis
CCMT: clear cell of the calciformligament
Pecome: our unspecified pecome
mTOR: mammalian (mechanistic) target of rapamycin
mTORC1: mammalian (mechanistic) target of rapamycin complex1
mTORC2: mammalian (mechanistic) target of rapamycin complex2
PEComa: Perivascular epithelioid cell tumor
PI3K: Phosphatidylinositol-3 Kinase
PI3P: Phosphatidylinositol-3-phosphate
GTP: Guanosine triphosphate
GTPase : Guanosine triphosphatase
S6K: Ribosomal protein S6 kinase
Irm: magnetic resonance imaging
CT: computed tomography
Mitf: Microphthalmia-associated transcription factor
TFE3: transcription factor 3
D VL2: dishevelled segment polarity protein 2
RBMX: (RNA-binding motif protein X-linked)
PET/CT: positron emission tomography/modensitometry
Fdg: Fluorodeoxyglucose
HMB-45: (Human Melanoma Black-45)
NSE: neuron-specific enolase
Sma: smooth muscle actin
RCP: multidisciplinary consultation meetings
VEGF: (Vascular Endothelial Growth Factor)
GM-CSF: Granulocyte-macrophage colony-stimulating factor
MSC: mesenchymal stem cell
Gap: gtpase activating protein

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