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Sclerosing PEComa of the Ureter

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ABSTRACT

A 55-years-old men was referred to the urology department because gross hematuria. Cystoscopy and urine cytology did not show abnormalities. The computed tomography (CT) showed 46 mm × 45 mm tumor located in the right ureteropelvic junction, suggestive of a urothelial tumor, which causes a slight ectasia of the pelvis and calyces. The patient underwent right radical laparoscopic nephroureterectomy without any complications. The postoperative pathological diagnosis of sclerosing PEComa. At the most recent follow-up, 18 months following the surgery, there is no evidence of recurrence.

KEYWORDS

PEComa; Sclerosing PEComa; Ureter

INTRODUCTION

Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms with unpredictable behavior. The main pathological feature is that are composed by epithelioid cells, which stain with melanocytic markers, associated with spindle cells reactive for smooth muscle marker [1].

According to the World Health Organization, PEComas (perivascular epithelioid cell neoplasms) are a "mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells" [2]. This kind of tumor was first described by Zamboni et al. [3] as a family of mesenchymal neoplasms containing the distinctive ''PEC'' (perivascular epithelioid cell), which typically shows a focal association with blood vessel walls, and shows immunophenotypic features of melanocytic differentiation and smooth muscle and The PEComa family tumors include: angiomyolipoma (AML), clear cell ''sugar'' tumor (CCST) of the lung, lymphangiomyomatosis (LAM), lymphangiomyoma, and a variety of unusual visceral, intra-abdominal, and soft tissue/bone tumors, described under the terms clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres , abdominopelvic sarcoma of perivascular epithelioid cells, and primary extrapulmonary sugar tumor , among others. 1) A search of the English literature indicated that no one previous cases of ureteral PEComa have been reported. We present this case as an unusual site of origin.

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CASE REPORT

A 55-year-old man was referred to our urology department with history of two months of gross hematuria. Non-smoker. No other malignancies. The cystoscopy and urine cytology did not show any abnormalities. A computed tomography scan (CT) was performed demonstrated the presence of a 46 mm × 45 mm tumor located in the right ureteropelvic junction, suggestive of a urothelial tumor, which causes a slight ectasia of the pelvis and calyces (Figure 1 - Figure 3).

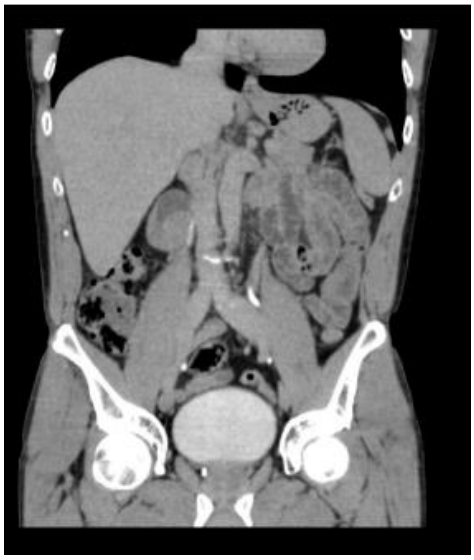

Figure 1: Coronal CT: 46 x 45 mm tumor located in the right ureteropelvic junction, suggestive of a urothelial tumor.

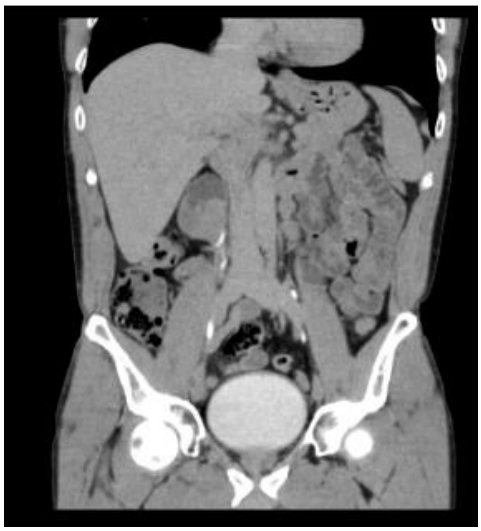

Figure 2: Coronal TC: slight ectasia with contrast into the urinary tract.

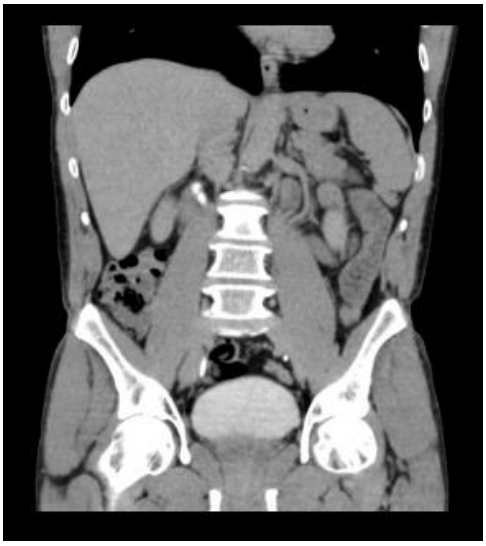

Figure 3: Dilatation of the renal pelvis.

Subsequently, the patient underwent a laparoscopic radical right nephroureterectomy without complications. He was discharged two days after the surgery. The postoperative pathological diagnosis showed in the next report.

Histologically

The tumor size was 5 cm allocated in upper ureter, composed of spindled cells mixed with epithelioid cells arranged in nests, focal areas of prominent stromal hyalinization with extensive stromal sclerosis. Neither necrosis, nuclear atypia, nor vascular invasion were observed. Infiltration of the ureter wall was present. Surgical resection margins: Tumor free Immunohistochemical revealed positive for HMB45, muscle-specific actin (MSA), calponin, caldesmon and desmin. Negative for Melan-A, S-100, AE1/AE3 EMA.

These findings suggested that the tumor was a sclerosing PEComa.

Three months later, a follow-up CT scan was normal. At the most recent follow-up, 18 months following the surgery, there is no evidence of recurrence.

DISCUSSION

The relationship between AML, LAM, and clear cell ‘‘sugar’’ tumor of the lung was first proposed in 1991[4] after recognition that the epithelioid cells that comprise these lesions express HMB-45 and show morphologic similarities. After that, Bonetti et al (5) 1992 coined the term ‘‘perivascular epithelioid cell’’ for this cell type, that are commonly associated with blood vessel walls. They proposed the designation ‘‘PEComa’’ for the lesions within this unusual family of tumors.

Occasional, PEComas show extensive stromal hyalinization, with some different clinical and pathologic tissue. They are uncommon and that cases have been adopted the name of the ‘‘sclerosing’’ variant of PEComa [6].

PEComas are usually indolent neoplasms that show a marked women predominance, ratio of women to men remains 4:1, with a peak incidence in middle-aged adults [7].

PEComas usually appear as solitary tumors, although some cases have been reported showing multiple satellite nodules of PEComa cells with perivascular distribution, a condition called PEComatosis [8].

The most common primary sites of PEComa are the mesentery, omentum, gastrointestinal tract and uterus. In the genitourinary tract, PEComas have been described mainly in the kidney; uncommonly, cases also occur in bladder, urachus, and prostate [9]. Most sclerosing PEComas arise in the retroperitoneum (renal and pararenal area) [6]. Till now, no case has been described in the ureter. So, to our knowledge this is the first case on the literature.

Histopathology this family is characterized by admixture of epithelioid and spindled cells arranged radially around blood vessels. Conventional PEComas show a nested or alveolar appearance. They appear as a proliferation of epithelioid cells arranged in solid nests and lobes. Proliferating cells show eosinophilic granular cytoplasm. Nuclear atypia and mitosis are not seen. When PEComa shows markedly hyalinized stroma, Hornik et al. have proposed the designation of ‘‘sclerosing PEComa.” [6]. Sclerosing PEComas show a slightly different microscopic appearance, with sheets of cells lying in a sclerosing stroma, they lack the characteristic delicate vasculature, and showing a predominant sheetlike growth pattern [1,6,10].

Immunohistochemical profile of PEComas is characterized by co-expression melanocytic markers (HMB-45), Melan-A, micropthalmia transcription factor (MiTF); and muscle smooth (markers muscle actin (SMA), calponin and desmin) [1,7]. and negativity for epithelial markers (EMA, CKs) cytokeratin AE1/AE3, myoglobin, synaptophysin, chromogranin, or vimentin.

Sclerosing PEComa seems to show more extensive expression of smooth muscle markers (including desmin and caldesmon) than other PEComas [6,7].

A recent study by Pan et al involved comparative genomic hybridization studies of PEComas. All exhibited chromosomal aberrations [7,8] like losses on chromosome 19, 18p,16p, 17p, 1p, and 18p and gains on chromosome X, 12q, 3q, 5 and 2q [11].

PEComas demonstrate uncertain tumor biology and unpredictable clinical evolution. While the majority of reported "PEComas" have a benign behavior, a minority have demonstrated locally recurrence, distant metastases and patient death [12].

According to Folpe [1] PEComas have been stratified into 3 prognostic categories - benign, uncertain malignancy potential, and malignant - according to histologic features such as size of the lesion, growth pattern, nuclear grade,

mitotic activity, necrosis, and vascular invasion [1]. The most useful features to predict poor outcome are large tumor size (>5 cm), mitotic activity >1/50 HPF, necrosis, high nuclear grade and cellularity, and infiltrative growth. Folpe et al. [7] suggested that tumors showing only one of these features should alert to a case of probably malignancy [7].

Optimal treatment for PEComas is not known at this time. PEC of the urinary tract can be preoperatively considered as urothelial [13]. Primary excision is usually curative, as most tumors are benign. However, locally advanced or metastatic disease portends a poor prognosis and strategies incorporating chemotherapy, radiation and immunotherapy have been reported [14].

PEComas show alterations in the mTOR pathway being sensitive to anti-mTOR drugs [15].

Several studies have stressed that mTOR-targeted therapy may be useful in the treatment of PEComas. The proven presence of mTOR pathway activation in these tumors presents the possibility of potentially promising therapeutic option.

DECLARATIONS

All the co-authors approve to submit the manuscript.

I confirm that the content of the manuscript has not been published or submitted for publication elsewhere.

This manuscript has not been published or is under consideration for publication with any other journal.

CONFLICT OF INTEREST

The author declares no having any potential conflict of interest.

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