Perivascular Epithelioid Cell Tumor of the Kidney — Report of an Incidentaloma and its Treatment

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Abstract

Perivascular epithelioid cell tumors (PEComas) are a relatively rare group of mesenchymal tumors that may occur in uncommon sites, including the kidney. We present the case of such a renal tumor that was found incidentally after a radiologic examination in a 53-year-old female. The patient was treated surgically and pathologic examination was diagnostic for the disease.

INTRODUCTION

World Health Organization defines the Epithelioid Cell tumors (PEComas) as mesenchymal tumors composed of histologically, immunohistochemically, ultrastructurally, and genetically epithelioid distinctive cells. These cells coexpress muscle and melanocytic markers. This family of tumors includes angiomylipomas, pulmonary lymphangioleiomyomatosis [1], clear cell tumors of the lung, abdominopelvic sarcomas as well as other tumors with the same characteristics, located in various sites [2]. PEComas are extremely rare tumors that can occur in any part of the human body and their natural history and prognostic features are yet undefined [3]. We report a case of an exophytic renal mass in a middle-aged woman diagnosed incidentally during imaging examinations and treated successfully through mass excision.

CASE PRESENTATION

The patient, a 53-year-old woman, was admitted in our surgical department after being submitted to an abdominal ultrasound examination for the evaluation of epigastric pain. This examination revealed incidentally the presence of an extrarenal mass of the right kidney, of diameter of 10 cm. The patient did not present any signs of renal dysfunction, from her medical history, or the clinical and laboratory examination, and did not complain about any other symptoms other than the aforementioned pain.

Further investigation included an upper-lower abdomen and retroperitoneal CT scan (Figure 1), as well as an abdominal MRI (Figures 2,3). Computerized tomography – guided biopsy was performed and the pathologic examination revealed the presence of a mass containing spindle cells and showing characteristics of a PEComa.

Surgery was considered as an optimal treatment, so an exploratory laparotomy was performed (Figure 4), and the excision of an exophytic mass of a diameter of 12 cm was realized (Figure 5). Right nephrectomy wasn’t considered necessary. The patient’s postoperative condition was stable and she was discharged on the fifth postoperative day. The second pathologic examination also revealed an exophytic benign renal PEComa (Figure 6). No further treatment was considered necessary and

Figure 1 CT imaging of the right pararenal mass.
the patient didn't show evidence of recurrence during the follow-up control.

**DISCUSSION**

In the female genitourinary tract, PEComas have been described in the kidney, bladder, and urethra. Angiomyolipomas can be controlled through ultrasound and CT-scan for any change in their preoperative size and usually are hyper-attenuated on precontrast CT, with or without fat component [4,5].

The expression of cathepsin K, TFE3 gene fusions and the genetic alterations of tuberous sclerosis complex have been associated with the disease [6]. The presence of heavily pigmented PEComas has also been described and must be differentiated from other pigmented tumors, such as malignant melanomas, pigmented clear cell renal cell carcinoma and

Although most PEComas are considered benign, some can be malignant, and classification criteria have been suggested for both renal and extrarenal lesions. Characteristics as carcinoma-like growth factors, extrarenal extension and renal vein involvement are associated with progression of the disease, so a long-term follow-up study is important [7]. In our case, oncologic evaluation didn’t consider necessary any other postoperative treatment and the patient remained healthy during follow-up control.

The differential diagnosis is difficult and a thorough preoperative control should be performed. The pathologic examination sets the final diagnosis of this rather benign, but rare condition, in order to estimate the prognosis or the necessity of a probable additional treatment.

REFERENCES


