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PLEOMORPHIC HYALINIZING ANGIOECTATIC TUMOR (PHAT) OF RENAL PARENCHYMA. FIRST CASE REPORTED IN LITERATURE

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Introduction: The pleomorphic hyalinizing angiectatic tumor (PHAT) is a rare non-metastatic tumor of soft tissue identified in 1996 (1). In adults, it generally occurs in the subcutaneous tissue of the lower limbs, although reported also in chest wall, buttock and arms. In the literature, only one case of PHAT has been described in the kidney at the level of the hilum but not involving renal pelvis or parenchyma (2). The clinical behavior of PHAT is characterized by a slow growth and a rate of local recurrences higher than 50%. Metastasis has not been reported. **Case Report:** A 61-year-old Caucasian obese female with a medical history of hypertension and hypercholesterolemia was admitted at the Emergency Unit for recurring gross hematuria since one year, becoming more frequent and severe in the last 3 months. Due to the severe anemia (hemoglobin of 7.7g/dl), the patient was transfused. Computed tomography (CT) scan revealed a parenchymal lesion of 4 cm in diameter of the lower pole of the right kidney. The lesion was only partially capsulated, mixed with a well evident cystic component, in strict contact with the lower calyx suspicious for infiltration. At cystoscopy, a clot emerging from the right ureteral meatus was evident. Urine cytology was negative and no imaging was indicative of transitional upper urinary tract tumor. After written informed consent, a right nephrectomy was performed and the patient was discharged on the 4th day. The histological exam revealed a partially capsulated lesion (3.7 cm in diameter) with a pseudo-cystic structure, including hemosiderin depositions, compressing but not invading the dilated lower calyx. The lesion was characterized by hyalinized clusters of thin-walled ectatic blood vessels within a stroma composed of sheets and fascicles of spindled and pleomorphic atypical

cells with intranuclear inclusions. At immunohistochemical analysis, AE1/AE3, EMA, CD31, S100, desmin, actin of smooth muscle, HMB45, ALK resulted negative. The lesion was classified as a PHAT. The patient is maintained in follow-up. **Discussion:** PHAT is a low-grade mesenchymal neoplasm of uncertain lineage described in soft tissue and characterized by diffusely infiltrative borders, although some do have well-circumscribed margins. The immunohistostaining for S-100 protein, actin, desmin, cytokeratin, CD-31, factor VIII antigens or epithelial membrane antigen are negative. To date approximately 22 cases of PHAT and 40 cases of its precursor, "early PHAT", have been described in the world literature. At least 3 cases of PHAT were reported to progress to high-grade myxofibrosarcoma (1). A case of PHAT arising in the hilum of the kidney, clinically mimicking an infiltrating tumor of the renal pelvis, has been described in 2012 (2). Our case is the second described in retro-peritoneum and the first of the renal parenchyma. In our patient, a partial nephrectomy was not carried out due to the absence of well-defined margins and apparent involvement of the lower calyx, although not confirmed by the pathological exam. If a partial nephrectomy is performed, a strict follow-up should be considered due to the high percentage of local recurrence characterizing the clinical behavior of PHAT (3).

1 Shi Wei *et al*: Complex analysis of a recurrent pleomorphic hyalinizing angiectatic tumor of soft parts. *Human Pathology* 43: 121-126, 2012.

2 Muhammad T *et al*: Pleomorphic hyalinizing angiectatic tumor of renal hilum. *Annals of Diagnostic Pathology* 16: 489-493, 2012.

3 Folpe AL and Weiss SW: Pleomorphic hyalinizing angiectatic tumor: analysis of 41 cases supporting evolution from a distinctive precursor lesion. *Am J Surg Pathol* 11: 1417-1425, 2004.

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