Actas Urológicas Españolas

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Scientific-clinical letters

Primitive neuroectodermical tumor of the kidney: Case report

Tumor neuroectodérmico primario renal: a propósito de un caso

Dear Editor,

Primitive neuroectodermal tumors (PNET) of the kidney are a rare type of cancer of the group of malignant peripheral neuroectodermal tumors. Their origin is unclear, but they are thought to derive from cells that migrated from the neural tube with variable capability of ectodermal or neuronal differentiation. The first description was written in 1918 by Arthur Purdy Stout, who classified them as "small round cell tumors".[1] In 1975, Seemayer et al were the first to show kidney involvement.[2] Since then, fewer than 50 cases have been published in the medical literature.

Renal PNET is an aggressive tumor that can appear at any age, but it is more common in the second and third decades of life, with no preference for either gender. It is characterized by a high capacity of local recurrence and metastasis. Disease-free survival at five years for all stages is around 45-55%.[3] However, the mean survival of patients with metastases is two years.

We present the case of a 36-year-old male with an unremarkable personal history who visited the emergency service for lumbar pain for two months. He complained of a constitutional syndrome since the onset, with asthenia, loss of appetite and loss of up to 6 kg of weight.

The physical examination revealed a soft abdomen, tender in the left hypochondrium upon deep palpation. A hard, painless mass was palpated in that hemiabdomen. The testicles appeared normal.

An urgent abdominal ultrasound reported a liver of a normal size and homogeneous parenchyma and a normal right kidney. A cystic, necrotic mass with fine echoes within its thick wall was observed in the left hemiabdomen, and displaced the left kidney forward and the spleen upward. Based on the suspicion of a retroperitoneal tumor, a testicle ultrasound and α-fetoprotein and β-HCG were ordered, and were normal. The thorax, abdomen and pelvic CT showed normal lungs and a tumor on the upper pole of the left kidney measuring 16×16×18 cm, with a cystic central area and a thick capsule; it displaced cranially the spleen and the renal vessels, which were not infiltrated. No local regional lymphadenopathies were found (fig. 1). The abdominal MRI confirmed the existence of a single left renal artery cranially displaced by the tumor. No infiltration of the left renal vein was seen, but numerous collateral venous structures in the upper portion of the mass appeared affected, probably by tumor neovascularization. A bone scan ruled out bone metastases.

The patient was offered radical nephrectomy with a left hemi-chevron incision. During surgery, a large cystic mass was observed that originated in the upper pole of the left kidney and locally invaded the spleen; this mandated a left radical nephrectomy and splenectomy. A radical lymphadenectomy was done and a biopsy taken from a suspicious area in the greater omentum. The piece weighed 1,100 g and measured approximately 21×18×16 cm.

The piece was then fixed and carved. A routine hematoxylin-eosin stain was done, plus an immunohistochemical analysis. Macroscopically, the tumor was heterogeneous, surrounded by a pseudocapsule, and markedly different from normal kidney parenchyma. The section surface was grayish, with many necrotic and hemorrhagic areas. Microscopically, the tumor consisted of small round cells organized in small lobules separated by fibrovascular septa. Perivascular pseudorosettes and Homer-Wright rosettes were observed. The material examined did not show lymph node metastases. The spleen was focally invaded by the tumor. A biopsy of the greater omentum was positive for tumor infiltration. The immunohistochemical analysis revealed diffuse cytoplasmic positivity for CD99 in the tumor cells (fig. 2). A neuron-specific enolase and vimentin stain was also positive. Chromogranin and desmin stains were negative.

The clinical presentation and the histological and immunohistochemical phenotype determined the diagnosis of renal PNET. The diagnostic study was completed after surgery with a bone marrow biopsy, which was negative. The

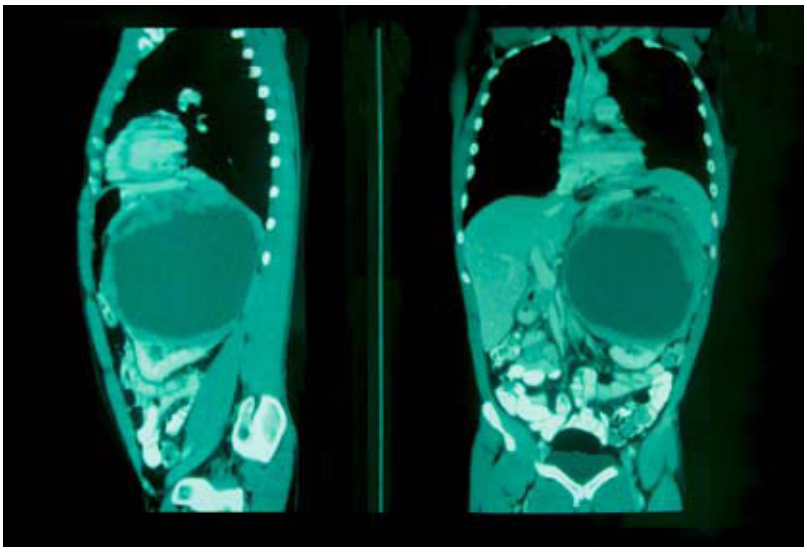

Figure 1 – Sagittal and coronal sections of the thorax, abdomen and pelvic CT showing a large cystic mass pending from the upper pole of the left kidney.

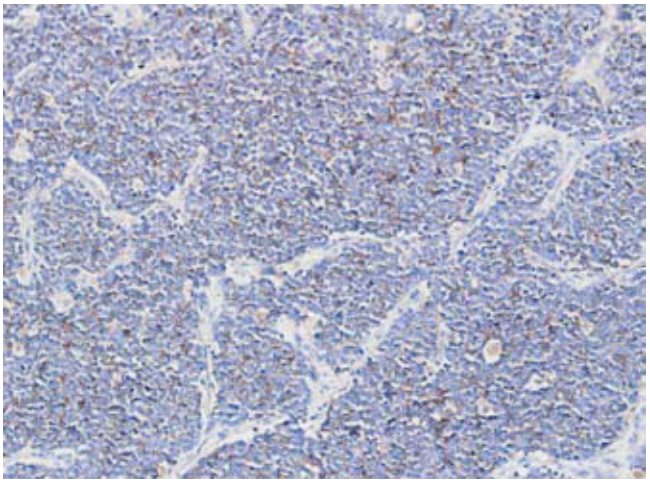

Figure 2 – Optical microscope image (×40), immunohistochemical stain showing diffuse positivity for CD99.

patient subsequently received nine cycles of chemotherapy with vincristin, adriamycin and cyclophosphamide, plus three more cycles with ifosfamide and VP16. Upon completion of the treatment, fractionated radiotherapy was given on the left hemiabdomen at 180 cy/day for 5 weekly sessions, for a total dose of 3,960 cGy, with no complications. After 22 months of follow-up, there is no evidence of distant metastases. The patient's general condition is stable, with a Karnofsky score of 90%.

This is a case of renal PNET in a young patient. PNETs often occur as soft tissue masses in the trunk or axial skeleton in children or young patients.[4-6] The kidney is a very rare location for this type of tumor; a medical literature review shows that fewer than 50 cases have been published so far; however, the exact number is difficult to ascertain because it is not easy to differentiate this tumor from Ewing's sarcoma.[4-6] Renal PNET is a unique clinical entity because it is more aggressive than neuroectodermal tumors in other locations and primary kidney tumors of any other origin. It often appears as a disseminated disease from the onset.[2,3,6]

The clinical manifestations of PNET are completely unspecific. Local progression signs like increasing pain and/or a palpable mass do not distinguish it from other kidney tumors. Imaging tests do not provide specific characteristics. A CT scan may show areas of necrosis and hemorrhage, and MRI helps to rule out the presence of a tumor thrombus.

A combination of conventional optic microscopy and specific immunohistochemical stains is necessary to establish the diagnosis and adequate adjuvant therapy. It is important to use the complete diagnostic repertoire, as using a single tool may be insufficient to exclude the numerous differential diagnoses, such as metastasis, clear cell renal carcinoma, Ewing's sarcoma, Wilms tumor, nephroblastoma, and neuroblastoma, among others.[5] Conventional optic microscopy of renal PNET reveals differentiated, round, small cells with round, uniformly stained nuclei with a non-prominent nucleolus; the cells tend to form rosettes and pseudorosettes. Hommer-Wright rosettes are common in PNET, but scarce in Ewing's sarcoma; they can be also found in neuroblastoma.[5,7] A combination of antibodies should be used for the immunohistochemical analysis. Most authors note that the expression of CD99 is typical of PNET, as was our case. However, differential diagnosis between PNET and extraosseous Ewing's sarcoma dictates the use of other stains, as both tumors are positive for CD99. In order to exclude Ewing's sarcoma, neuronal markers such as neuron-specific enolase, which is positive in PNET and negative in Ewing's sarcoma, must be employed.[5,7]

The progression of renal PNET depends on the stage at diagnosis, the extension of the surgical resection, the

presence of margins, the histological classification, and time to adjuvancy. PNET is an aggressive tumor that tends to recur locally and to metastasize to lymph nodes, lung, liver, bone, and bone marrow, which entails a worse prognosis.[8] The five-year disease-free survival rate for all stages is 45-55%.[3] Patients with a disseminated disease at diagnosis have a mean survival of only two years.[3] Polychemotherapy with high doses of vincristin, adriamycin, and cyclophosphamide has yielded good outcomes.[3,8] However, due to the rarity of the tumor, no randomized studies on the efficacy of different chemotherapy regimens have been published.[3]

We conclude that renal PNETs are an uncommon condition characterized by aggressivity and local and distant recurrence capability, which causes them to have a poor prognosis. This entity must be considered in the differential diagnosis of renal masses in young patients, especially those presenting with a disseminated disease at onset. The diagnosis is reached by exclusion of other small cell renal tumors. An accurate anatomopathological diagnosis is very important clinically because it may determine which of the various chemotherapy regimens will control the disease better.

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Sclerosing sertoli cell tumor. An infrequent type of testicular neoplasm

Tumor de células de sertoli esclerosante. Un subtipo infrecuente de neoplasia testicular

Dear Editor,

Sex cord-gonadal stromal, or non-germ cell neoplasms of the testes account for approximately 5% of testicular tumors, and most of them are Leydig cell tumors.[1] Pure Sertoli cell testicular tumors are uncommon and account for approximately 1% of testicular tumors.[2] Because of their histological and clinical variability, these tumors, usually considered benign, are a heterogeneous entity. They are classified according to their histological characteristics in the following subtypes: classic (unspecified), large cell calcifying variant, and sclerosing variant.[3]

We present the case of a 30-year-old male patient with a history of renal colics and sinusitis who complained of slight enlargement of his left testicle associated with testicular pain. The physical examination revealed a left testis tender upon palpation especially in the tail of the epididymis, with no masses. A testicular ultrasound (fig. 1) showed an 11-mm