Rev Cubana Urol 2022;11(3):e886 ISSN: 2305-7939 RNPS: 2296

Publicación Científica de la Sociedad Cubana de Urología

Presentación de caso

Presentation of three cases of primary renal sarcomas

Sarcomas renales primarios. Presentación de tres casos

Alba María García Cano Fernández[1*] https://orcid.org/0000-0001-9091-4794

Juan Boronat Catalá[1] https://orcid.org/0000-0003-3935-9806

Victoria Diego García[1] https://orcid.org/0000-0001-5416-7963

Luis Llanes González[1,2] https://orcid.org/0000-0002-0162-585X

[1]Urology Department of University Hospital of Getafe. Madrid, España.
[2]Francisco de Vitoria University. Madrid, España.

[*]Corresponding author: albagcf.uro@gmail.com

ABSTRACT

A retrospective review of cases of primary renal sarcomas diagnosed between 1995 and 2020 at the University Hospital of Getafe (Spain) was carried out, with the aim of contrasting the clinical-pathological characteristics, treatment and evolution. Primary renal sarcomas account for 0.58 % of all renal tumors diagnosed at this institution. All patients were treated by excision of the tumor mass. Two patients had localized disease at the time of diagnosis and one of them had metastatic disease. Two of the patients had recurrence and died. The time of progression was 3.7 months. The median survival time was 9.45 months. Primary renal sarcomas are very rare neoplasms. The treatment of choice is surgery, recurrence is very common and the overall prognosis is poor.

Key words: renal malignancies; sarcoma; nephrectomy; disease progression; prognosis.

RESUMEN

Se realizó una revisión retrospectiva de los casos de sarcomas renales primarios diagnosticados entre 1995 y 2020 en el Hospital Universitario de Getafe (España), con el objetivo de contrastar las características clínico-patológicas, el tratamiento y la evolución. Los sarcomas renales primarios representan el 0,58 % de todos los tumores renales diagnosticados en dicha institución. Todos los pacientes fueron tratados mediante la escisión de la masa tumoral. Dos pacientes tenían la enfermedad localizada en el momento del diagnóstico y uno de ellos tenía la enfermedad metastásica. Dos de los pacientes presentaron recidiva y fallecimiento. El tiempo de progresión fue de 3,7 meses. La mediana de tiempo de supervivencia fue de 9,45 meses. Los sarcomas renales primarios son neoplasias muy raras. El tratamiento de elección es la cirugía, la recidiva es muy frecuente y el pronóstico global es malo.

Palabras clave: neoplasias renales; sarcoma; nefrectomía; progresión de la enfermedad; pronóstico.

Recibido: 03/08/2022, **Aprobado:** 16/09/2022

Introduction

Retroperitoneal sarcomas are rare and heterogeneous tumours, accounting for 10-15 % of soft tissue sarcomas. Primary renal sarcomas are very rare tumours accounting for 3 % of all malignant renal tumours. Retroperitoneal sarcomas usually have a peak age incidence in the 50 s, occurring similarly in men and women. Clinically, they are asymptomatic and diagnosed incidentally, or oligosymptomatic with a painful abdominal mass. Anatomopathological analysis is required for definitive diagnosis. The main therapeutic approach is surgical, with the aim of achieving free resection margins. Adjuvant therapies are not well defined at present, so the decision must be individualized. These tumours usually have a poor prognosis, with distant dissemination shortening survival or frequent local recurrences, which makes close follow-up very important.[1,2,3,4]

A retrospective review of cases of primary renal sarcomas diagnosed between 1995 and 2020 at the University Hospital of Getafe (Spain) was carried out. Primary renal sarcomas account for 0.58 % (3/518) of all renal tumours diagnosed in our institution.

A review of the literature was carried out in order to compare clinical pathological characteristics, management and follow-up.

Cases presentation

Clinical case 1

A 35-year-old male smoker was evaluated for constitutional syndrome and a painful abdominal mass in the right hypochondrium. Abdominal computed tomography (CT) (figure 1) and ultrasound showed a 13 cm mass, originating in the right kidney capsule, with interaortocaval adenopathies.

Open radical surgery was performed. The anatomopathological diagnosis was dedifferentiated liposarcoma reaching the resection surface (pT2).

Forty-six days after surgery, he presented early massive tumour recurrence in the form of pulmonary nodules and peritoneal carcinomatosis, finally dying 60 days after surgery.

Clinical case 2

A 59-year-old man, smoker and drinker, was studied for a mass in the hypochondrium and left flank and weight loss. Abdominal ultrasound and CT scan (figure 2) identified a 15cm heterogeneous polylobulated left renal mass with no associated lymphadenopathy.

Open radical surgery was performed. The pathological diagnosis was undifferentiated pleomorphic sarcoma (pT2).

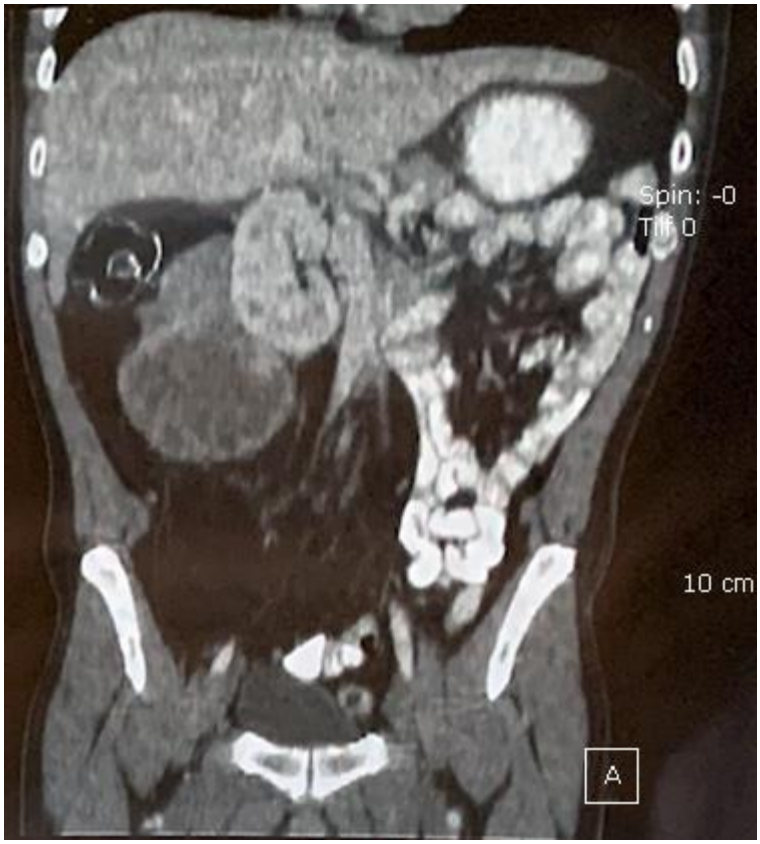

Fig.1 - Abdominal CT scan with contrast. Right retroperitoneal heterogeneous mass originating from the right kidney.

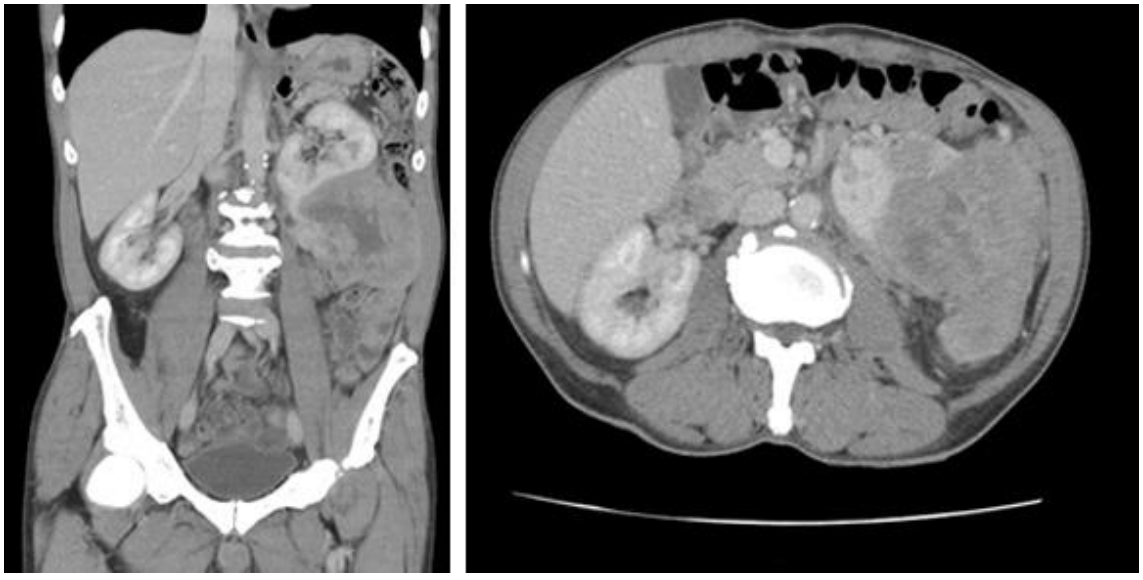

Fig. 2 - Abdominal CT scan with contrast: left renal mass.

At 6 months he presented with disseminated tumour recurrence with two anterior spleen masses, pulmonary nodules and peritoneal implants (figure 3). Chemotherapy treatment was started with adriamycin in the first line and docetaxel in the context of tumour progression. He died of sepsis secondary to pneumonia with severe hypoxaemic respiratory failure at 387 days.

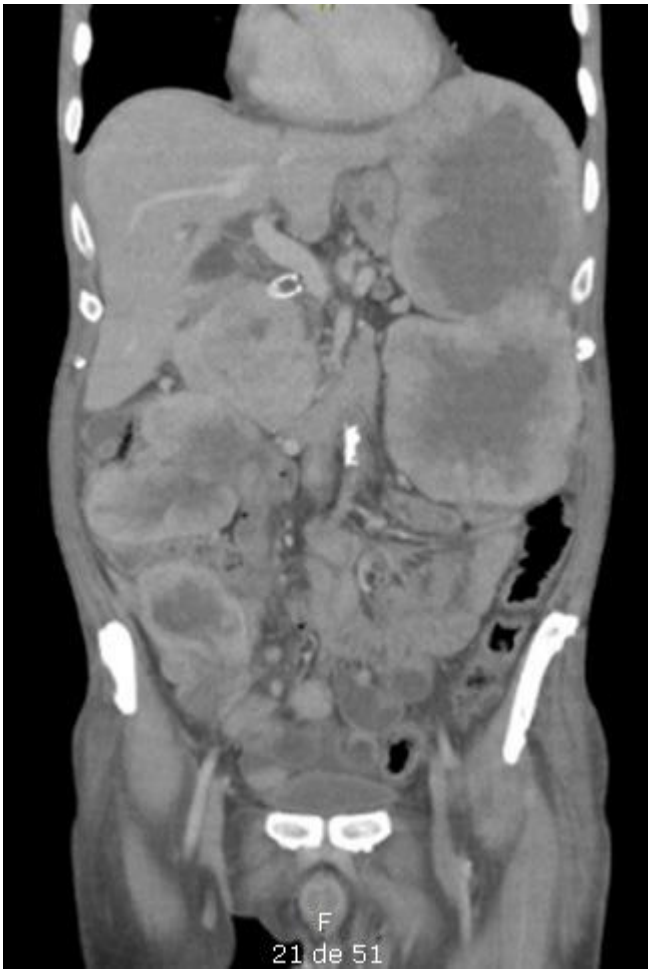

Fig. 3 - Abdominal CT with contrast: tumour progression, two anterior spleen masses with multiple peritoneal implants.

Clinical case 3

A 67-year-old man was studied for anaemia and a painful mass in the left hypochondrium. CT scans (figure 4) showed a 17cm left renal neoformation with no distant lesions.

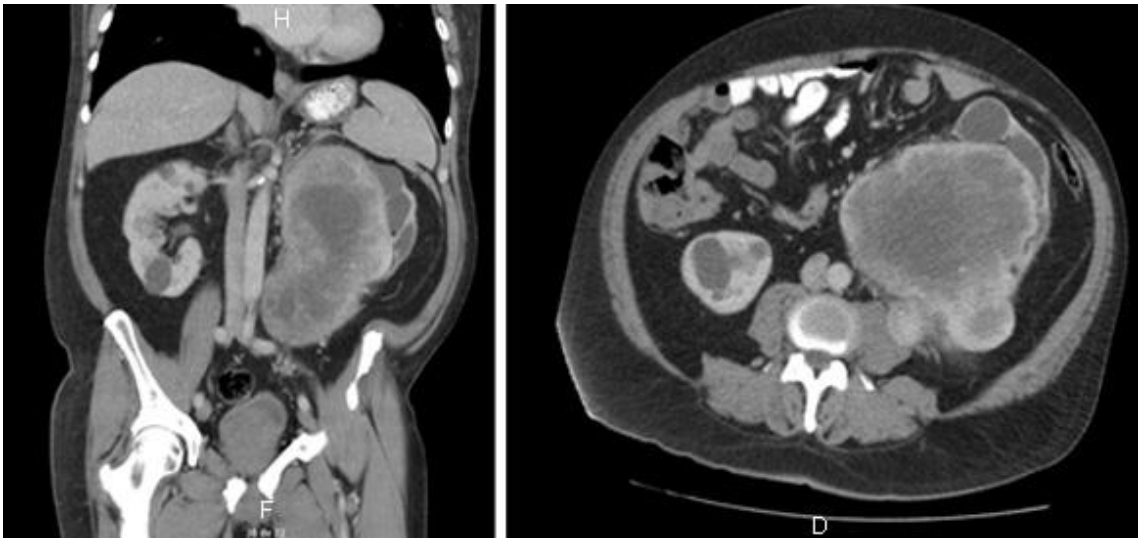

Fig. 4 - Abdominal CT with contrast: left renal mass.

Open radical surgery was performed. The anatomopathological diagnosis was a malignant solitary fibrous tumour infiltrating the renal hilum, respecting the capsule, adrenal and surgical margins (pT2).

At 12 months follow-up, no local or distant recurrence was observed.

None of the cases presented alterations in laboratory tests.

Progression-free survival time (PFS) was 3.7 months (1.53 and 5.8 months in each case). Median survival was 9.45 months (2 and 12.9 months in each case).

Discussion

Renal sarcomas are rare and heterogeneous tumours.[1,2,3] Only three cases have been reported in our centre in 24 years, which represents 0.58 % of all renal tumours diagnosed in our institution.

Dedifferentiated liposarcomas are high-grade tumours that have the potential to metastasise with a high risk of death, as occurred in one of the cases.[1]

Malignant fibrous histiocytoma, or undifferentiated pleomorphous sarcoma, is the most common sarcoma of adulthood. In 12-15 % of cases they are located in the retroperitoneum, being exceptional their origin from the renal capsule. They have a poor prognosis with a survival rate of 60 % at two years and a recurrence rate of between 50 and 82 %. The most frequently metastasised organs are the lung and liver.[4]

Solitary fibrous tumours are generally benign fibroblastic mesenchymal neoplasms, which rarely metastasise. They are predominantly intrathoracic, with renal localization being very rare.[5,6]

Most soft tissue sarcomas arise sporadically; however, in a small proportion of cases, genetic alterations have been identified, such as the presence of neurofibromatosis, hereditary retinoblastoma, Li-Fraumeni syndrome, Werner syndrome or Gardner syndrome, lymphoedema or a history of previous radiotherapy as predisposing factors or factors associated with the development of this type of tumour.[7]

Clinically, this type of tumour produces few and very non-specific symptoms. They may also produce symptoms related to local invasion or distant metastases.[1,4]

Definitive diagnosis requires histological confirmation. The imaging studies of choice are thoraco-abdomino-pelvic CT with contrast to evaluate the primary tumour and as an extension study.[1,2,3,5,8]

The radiological findings of a soft tissue sarcoma on CT images are similar to those of other soft tissue tumours and there are no pathognomonic features for this tumour type. Renal sarcomas typically manifest as a large, well-defined, heterogeneous mass with or without areas of necrosis.[5]

The treatment of choice for sarcomas is surgical resection with the aim of achieving free resection margins. Incomplete resection on a palliative basis is only recommended in selected cases.[1,7,8]

The administration of adjuvant treatments in this type of tumour is not well defined, so it should be done on an individualised basis in all cases.[1]

Although there is no level I evidence to recommend radiotherapy in retroperitoneal sarcomas, there are reviews that suggest it would be beneficial.[9] Neoadjuvant chemotherapy is indicated in locally advanced or high-grade tumours in order to reduce tumour volume and thus facilitate a complete and more conservative surgical resection. New strategies such as immunotherapy are being studied.[2,10]

The main prognostic factor is complete resection to obtain clean surgical margins. Other prognostic factors are tumour size, histology, radiotherapy treatment, age, visceral or retroperitoneal location of the tumour.[3,5]

Despite optimal surgical resection, disease recurrence is common, with 5-year recurrence-free survival approaching 50-60 % for some histological subtypes, and poor overall survival. Distant metastases usually settle in the lung and liver.[1,7,10]

These tumours are characterized by a poor prognosis due to their tendency to recur locally and/or distantly, with a fatal outcome soon after diagnosis in most histological subtypes. Given the high recurrence rate of these tumours, long-term follow-up is mandatory.[1,7,8]

Postoperative surveillance protocols have been proposed according to histology, such that for indolent sarcomas, follow-up with clinical and abdomino-pelvic CT is performed every 6 months for 3 years and annually thereafter. For aggressive tumours, clinical examination and abdomino-pelvic CT scan every 3-4 months for 2 years, every 6 months for the next 3 years and then annually thereafter. Given the rarity of these tumours, it is recommended that patients be included in clinical trials and that their functionality and quality of life be assessed periodically.[8,11]

Conclusions

Renal sarcomas are very rare and heterogeneous tumours characterized by a poor prognosis, with distant dissemination shortening survival or frequent local recurrences, which makes close follow-up very important.

References

1. Mullen JT, Delaney TF. Clinical features, evaluation, and treatment of retroperitoneal soft tissue sarcoma. UpToDate. 2022 [access 22/08/2022]. Available from: https://www-uptodate-com.m-hug.a17.csinet.es/contents/clinical-features-evaluation-and-treatment-of-retroperitoneal-soft-tissue-sarcoma/print

2. Tahri R, Gamra L, El Otmany A. Liposarcome de la loge rénale: à propos de deux cas avec revue de la littérature. PAMJ. 2018;29:167. DOI: 10.11604/pamj.2018.29.167.1850

3. Messiou C, Moskovic E, Vanel D, Morosi C, Benchimol R, Strauss D, *et al*. Primary Retroperitoneal Soft Tissue Sarcoma: Imaging appearances, pitfalls and diagnostic algorithm. Eur J Surg Oncol. 2017;43(7):1191-8. DOI: 10.1016/j.ejso.2016.10.032

4. Sierra Labarta R, Gil Sanz MJ, Muñoz González G, Rioja Sanz LA. Un nuevo caso de histiocitoma fibroso maligno dependiente de cápsula renal. Actas Urol Esp. 2010 [access 22/08/2022];34(1):122-4. Available from: http://scielo.isciii.es/scielo.php?script=sci_arttext&pid=S0210-48062010000100023&lng=es

5. Yilmaz C, Kabatas S, Ozen OI, Gulsen S, Caner H, Altinors N. Solitary fibrous tumor. J Clin Neurosci. 2009;16(12):1578-81. DOI: 10.1016/j.jocn.2009.02.039

6. Demicco EG, Meyer C. Solitary fibrous tumor. UpToDate. 2022 [access 22/08/2022]. Available from: https://www.uptodate.com/contents/solitary-fibrous-tumor

7. Brennan MF, Antonescu CR, Moraco N, Singer S. Lessons learned from the study of 10,000 patients with soft tissue sarcoma. Ann Surg. 2014;260(3):416-21. DOI: 10.1097/sla.0000000000000869

8. Trans-Atlantic RPS Working Group. Management of Primary Retroperitoneal Sarcoma (RPS) in the Adult: A Consensus Approach From the Trans-Atlantic RPS Working Group. Ann Surg Oncol. 2015;22(1):256-63. DOI: 10.1245/s10434-014-3965-2

9. Tiong SS, Dickie C, Haas RL, O'Sullivan B. The role of radiotherapy in the management of localized soft tissue sarcomas. Cancer Biol Med. 2016;13(3):373-83. DOI: 10.20892/j.issn.2095-3941.2016.0028

10. Keung EZ, Lazar AJ, Torres KE, Wang WL, Cormier JN, Ashleigh Guadagnolo B, et al. Phase II study of neoadjuvant checkpoint blockade in patients with surgically resectable undifferentiated pleomorphic sarcoma and dedifferentiated liposarcoma. BMC Cancer. 2018;18(1):913. DOI: 10.1186/s12885-018-4829-0

11. Zaidi MY, Canter R, Cardona K. Post-operative surveillance in retroperitoneal soft tissue sarcoma: The importance of tumor histology in guiding strategy. J Surg Oncol. 2018;117(1):99-104. DOI: 10.1002/jso.24927

Conflict of interests

The authors declare that does not exist an interest conflict.