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Oncology

Renal rupture – Not what it seems

Renate Pichler [a, *, 1], Isabel Heidegger [a], Gert Schachtner [a], Jannik Stuehmeier [a], Abbas Agaimy [b], Hans Maier [c], Bernhard Glodny [d], Peter Rehder [a, 1]

[a] Medical University Innsbruck, Department of Urology, Innsbruck, Austria
[b] Friedrich-Alexander-University, Institute of Pathology, Erlangen, Germany
[c] Clinical Pathology and Cytodiagnostics, Tyrolean State Hospitals Ltd, Innsbruck, Austria
[d] Medical University Innsbruck, Department of Radiology, Innsbruck, Austria

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1. Introduction

Synovial sarcoma represents only 6–10% of all soft tissue sarcomas.[1] Primary synovial sarcoma arising from the kidney is an extremely rare and aggressive disease.

This report is the first describing primary synovial sarcoma of the kidney presenting with renal rupture.

2. Case presentation

A 20-year-old male judo teacher presented with acute left flank pain and gross hematuria. At the time of this presentation, the patient was hemodynamically stable. He had no clear history of blunt abdominal trauma, or preexisting conditions or previous surgery. Multiphase abdominal computed tomography confirmed an active extravasation from a segmental renal artery during the arterial phase (Fig. 1A), with a consecutive 17 × 8 × 4 cm retroperitoneal hematoma (Fig. 1B), and also a hematoma near the renal hilus (Fig. 1B). Nephrographic and delayed pyelographic phase images revealed no urinary extravasation, but with poor function of the affected kidney (Fig. 1C). Selective angioembolisation was performed to manage active arterial bleeding. Retrospectively, the angiography was suggestive of a pathological vascular displacement caused by a centrally located mass (Fig. 2A–B). 24 hours later open surgery due to hemodynamic instability was necessary resulting in hematoma evacuation (2.5 L) and nephrectomy. Surgery revealed complete dehiscence of the renal pelvis from the kidney. Tumorous tissue was noted within the hematoma. Histological examination showed poorly differentiated spindle cell neoplasms with high mitotic activity suggestive of primary monophasic synovial sarcoma of the kidney. Immunohistochemical staining showed positive reactivity for CD99, bcl-2, and TLE1 but not CK7, CK20, S-100, synaptophysin, smooth muscle actin, desmin, myogenin and h-caldesmon (Fig. 3). Molecular fluorescence in situ hybridization (FISH) analysis showed the t(X;18)(p11;q11) chromosomal translocation thus confirming a diagnosis of monophasic spindle cell synovial sarcoma (Fig. 4). Postoperatively, contrast-enhanced computed-tomography combined with ^{18}F-fluorodeoxyglucose (^{18}F-FDG) PET confirmed no evidence of lymphogenic or hematogenic metastasis. The patient was referred to the oncology department for further systemic treatment.

3. Discussion

To date, about 60 cases of primary synovial sarcoma arising within the kidney have been reported in the literature,[2] the first case being reported in 2000 by Argani et al.[3] Diagnosis can be challenging due to various renal tumors with similar histological features such as Wilms tumor, Ewing sarcoma, or renal cell carcinoma with sarcomatoid differentiation, resulting in a diagnostic dilemma with various differential diagnosis.[4] Definitive diagnosis is made by adjunct immunohistochemistry and can be confirmed by molecular-genetic analysis detecting SS18-SSX gene fusion.[5] Immunohistochemical analysis of primary renal synovial sarcoma have consistently shown a positive staining for CD56, Vimentin, CD99 and focal positivity for EMA.[4]

* Corresponding author. Medical University Innsbruck, Department of Urology, 35 Anich Street, A-6020, Innsbruck, Austria.
E-mail address: Renate.Pichler@i-med.ac.at (R. Pichler).
[1] Both authors contributed equally as first and corresponding authors.

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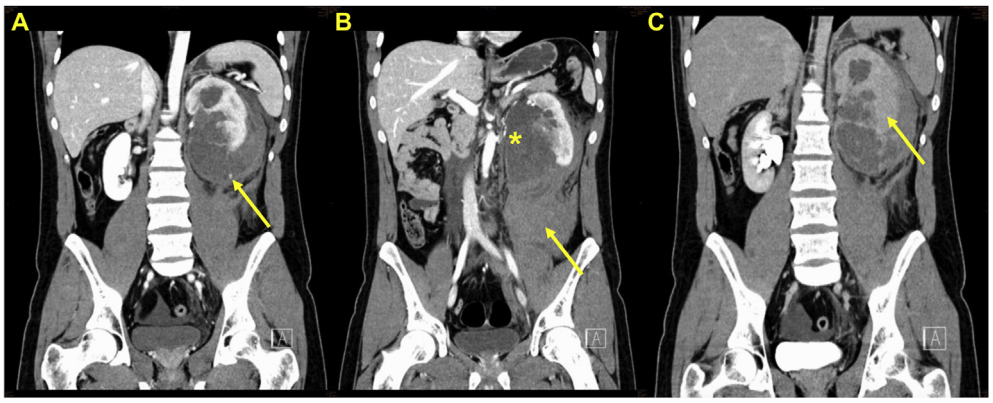

Fig. 1. Abdominal computed tomography (CT) scan showing active extravasation of a segmental renal artery (**A**, arrow), with a 17 × 8 × 4 cm retroperitoneal hematoma (**B**, arrow), and also a hematoma (retrospectively tumor) near the renal hilus (**B**, asterisk). Nephrographic phase revealed no urinary extravasation of the affected kidney, but with poor function (**C**, arrow).

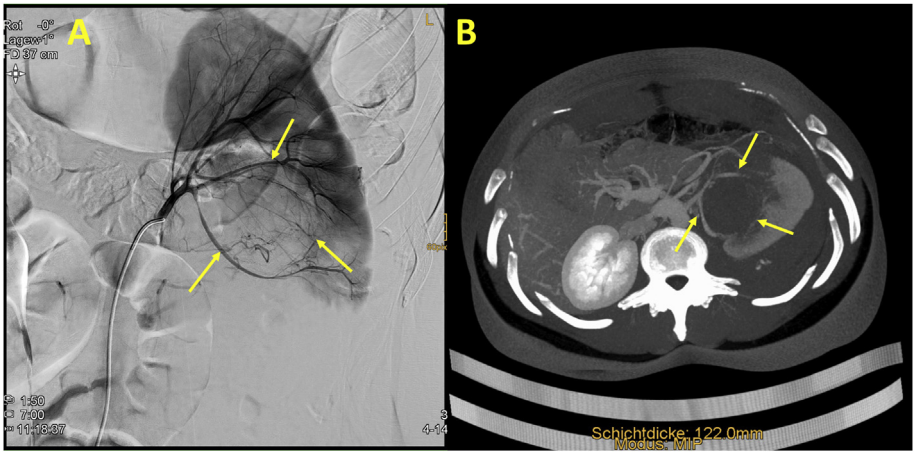

Fig. 2. Angiography demonstrating pathological vascular displacement caused by a centrally located mass and pathological tumor vessels (**A**, arrows). Transversal Maximum Intensity Projection Reconstruction (MIP) of the CT retrospectively showing the displacement as well (**B**, arrows).

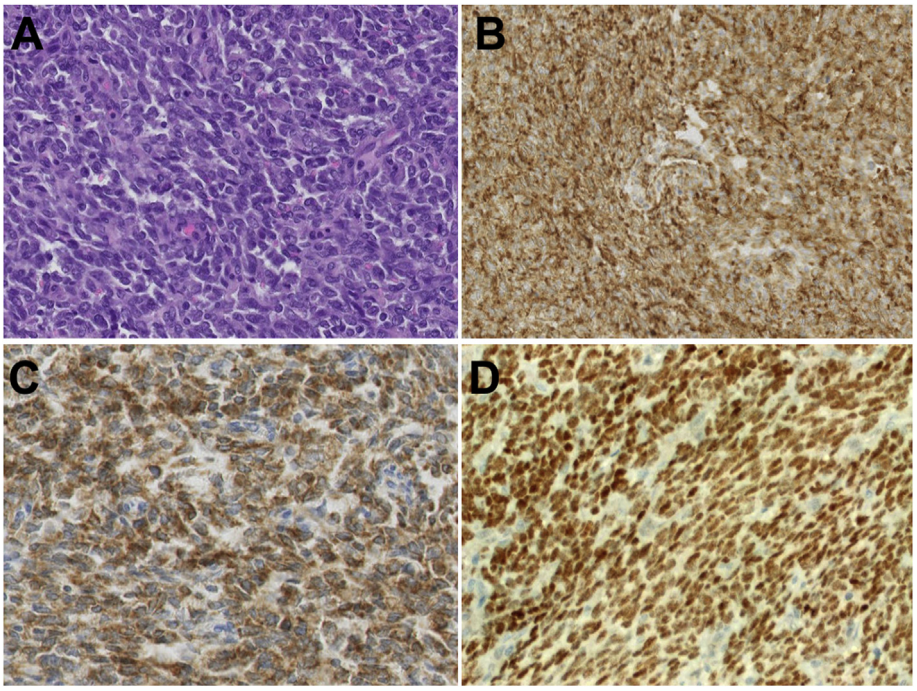

Fig. 3. Hematoxylin and eosin staining of the tumor (**A**) and positive immunohistochemical staining for CD99 (**B**), Bcl-2 (**C**) and TLE1 (**D**).

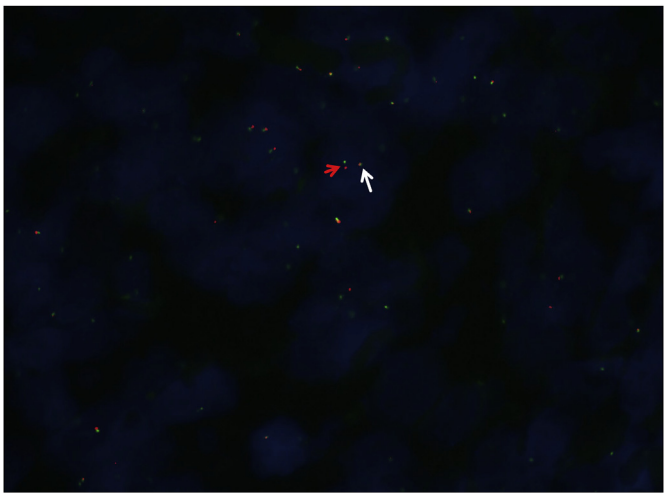

Fig. 4. SYT FISH probe showing fused green and orange signals (**white arrow**) and split-aprat green and orange signals (**red arrow**). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

4. Conclusion

Primary synovial sarcomas of kidney are rare aggressive neoplasms with poor outcome. No specific guidelines for the treatment of primary renal synovial sarcoma are established due to the rarity of the disease. A combined therapeutic strategy including radical surgery and chemotherapy (ifosfamide and doxorubicin) seems to achieve the best oncological results.

Conflicts of interest

None.

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