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Sarcomatoid Carcinoma with Angiosacromatous Differentiation of the Bladder Mimicking Primary Angiosarcoma: A Diagnostic Challenge

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Introduction

Sarcomatoid urothelial carcinoma (UC) is a high-grade aggressive subtype of UC defined by the WHO classification of tumours of the Urinary System and Male Genital Organs with "the presence of areas that are indistinguishable from sarcoma" [1]. In fact, it is a biphasic neoplasm with both; malignant epithelial and mesenchymal differentiation. Heterologuous components have been described, among which angiosarcoma is exceedingly rare [1,2]. Given its poor prognosis and aggressive clinical behavior, this subtype of UC should be distinguished from other bladder carcinoma, and further immunophenotypic study and /or molecular work-up must be performed. Thus, accurate diagnosis is mandatory for treatment purposes. The main differential diagnosis is primary angiosarcoma and pseudoangiosarcomatous UC; which both, show morphologic and immunophenotypic overlap with sarcomatoid UC with heterologuous angiosaroma (UCAC) component [2]. Histologically, UCAC presents as true angiosarcoma associated to conventional UC component [4]. On immunohistochemistry, the neoplastic cells express all vascular markers: CD31 CD34 and erg, with focal reported pan-cytokeratin positivity, with focal or diffuse expression of urothelial markers such as: GATA3.

Herein we describe the case of 56 you women, without a previous history of radiation therapy, presenting to the emergency department with marked gross hematuria. Transurethral resection of the bladder (TURB) was performed, the bladder showed a diffuse hemorrhagic pattern. Histology, showed atypical epithelioid to spindle cells arranging in anastomosing channels, with decohesion and pseudo lumina. Focal areas of conventional high grade papillary urothelial carcinoma were observed (Figure 1). On immunohistochemistry, angiosarcoma component was diffusely positive for vascular markers: CD31 and erg, focally positive for AE1/AE3 and negative for CK7, CK20. However, GATA3 was also positive in the angiosarcoma cells (Figure 2). No immunohistochemistry was performed on UC component, since the diagnosis was straightforward. The patient has fatal outcome few days later, in the course of massive pulmonary emboli.

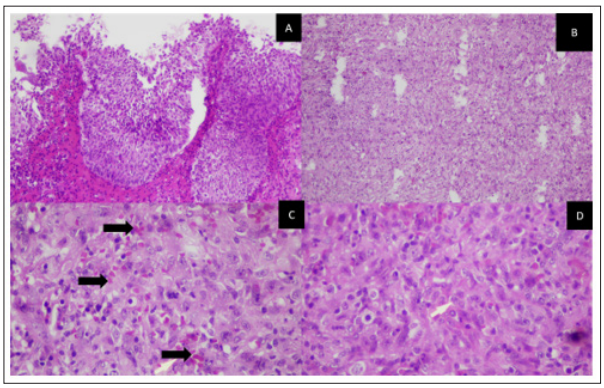

Figure 1: Histology Findings

A: conventional noninvasive urothelial carcinoma component (HE medium power)
B: Angiosarcoma component histological findings (HE medium power)
C: Angiosarcoma component histological findings (HE high power), vascular lumen (black arrow)
D: Angiosarcoma component histological findings (HE power high)

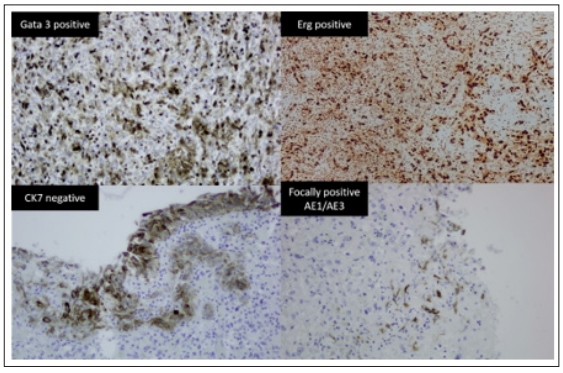

Figure 2: Immunohistochemistry Findings on Angiosarcoma Component

A: GATA3 positive
B: Erg positive
C: CK7 negative (with positivity in the surface urothelial epithelium)
D: AE1/AE3 focally positive
E: CK20 negative

Statement of Significance
This case report shed light on the scarcity of this entity, and highlights the diagnostic difficulty as well, to rule out the main differential diagnosis: primary angiosarcoma. This case will also allow for providing more data about this rare tumor.

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Competing Interests
The authors declare that they have no competing interests.

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Ethics Approval and Consent to Participate
Not applicable.

Consent
Written consent was obtained from the patient.

Availability of Data and Materials
Not applicable.

Authors' Contributions
All authors read and approved the final manuscript.

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