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Angiosarcoma Tumor – An Unusual Localization of a Rare Tumor: A Case Report

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Abstract

Introduction: Angiosarcoma is a vascular malignant tumor composed of histologically and immunohistochemically distinctive perivascular cells. This type of tumor is rare but bladder localization is even rarer: only 14 cases of primary urinary bladder angiosarcoma have been reported in the English literature.

Case presentation: A case of bladder angiosarcomas in a 81-year-old Caucasian man who presented with hematuria, treated with surgery is described.

Conclusions: The rarity of bladder angiosarcoma tumors hinders the development of a standard therapeutic approach, and thus requires case report descriptions. There is a need for cooperative studies to enlarge the case series and establish the best treatment strategy for this rare disease.

Keywords: Bladder; Metastasis; Angiosarcoma

Introduction

Primary urinary bladder angiosarcoma is an extremely rare mesenchymal tumor [1,2]. It presents mainly in elderly men with hematuria. Information about diagnosis, treatment, evolution and prognosis of this type of tumor are very limited.

We present our case as the 15th reported case of primary urinary bladder angiosarcoma. Case Report

A case of an 81-year-old Caucasian man (body mass index 25kg/m 2) who presented with hematuria, in our Emergency Departed. A history of benign hypertension disease and lower urinary tract symptoms secondary to benign prostatic hyperplasia treated with alpha-blockers.

His physical examination was normal. A digital rectal examination revealed a moderate increase in prostatic gland size without any finding suspicious for neoplastic disease. The results of his blood tests were abnormal with Hb 9,8gr\dl and a mild leukocitosis 14.000 WB. An abdominal ultrasound showed normal kidneys and a bladder with thickened detrusor.

He undergone to cystoscopy, showing a low bladder distension and strange area of 1 centimetre bleeding brown on the left side of the wall. He performed a tc scan (Figure 1) that revealed only a 1 cm solid mass.

A trans urethral bladder resection (TURB) was performed. The first histological diagnosis was not clear and after 1 mounth a re-TURB was performed showing the left side of the bladder with five brown areas too.

Abdominopelvic contrast computed tomography performed before re-TURB showed mild heterogeneous enhancement of the left side of the bladder which infiltrated his bladder wall. No machroscopic metastasis was showed but loco-regional adenopathy was observed (Figure 2).

He then underwent radical cistectomy and extensive bilateral linphectomy (Figure 3).

No intra- and postoperative complications were observed; he was discharged on the seventh postoperative day.

At the medication control 10 days after discharged the patient was evealuated for oncological medical treatment but unlikely we observed a sudden deterioration of the general conditions and definited unfit for chemoterapy.

He died 1 month after surgery in a palliative care institution.

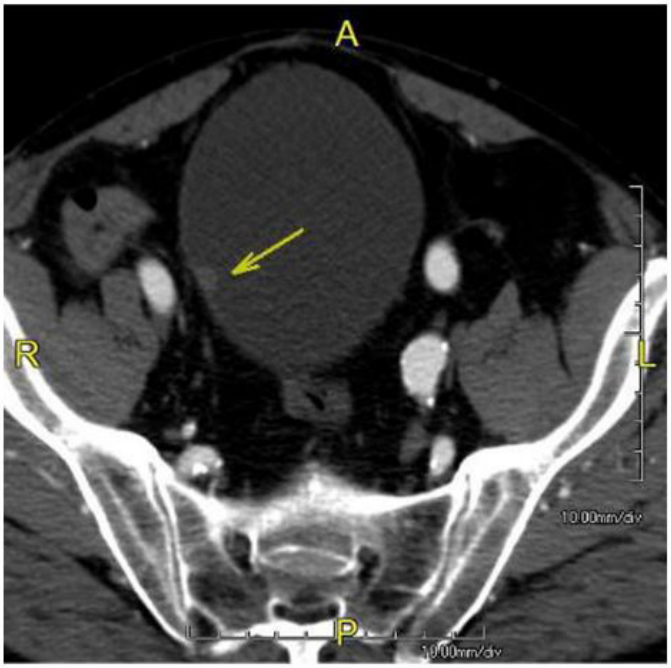


Figure 1: CT pelvic scan

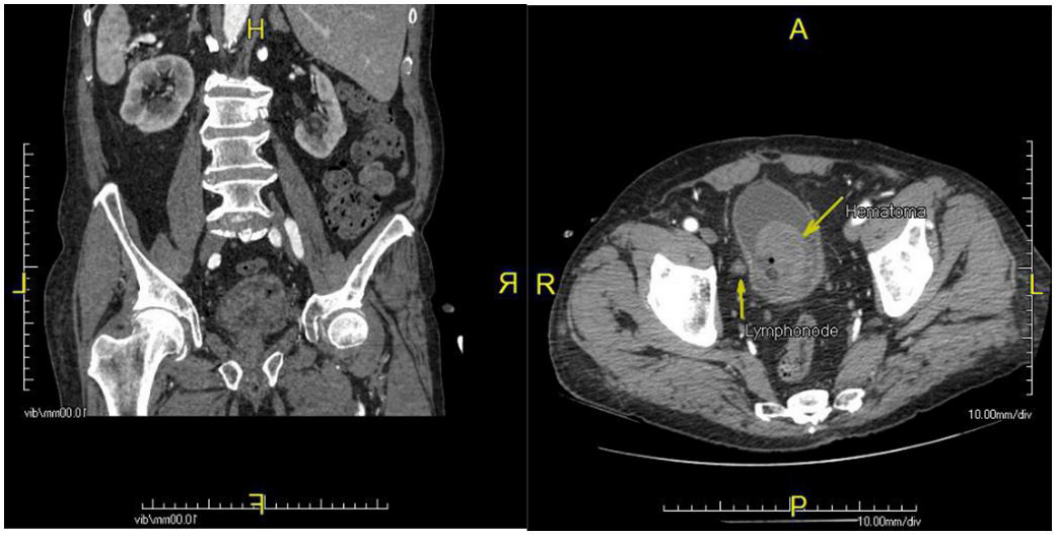


Figure 2: CT abdominal scan

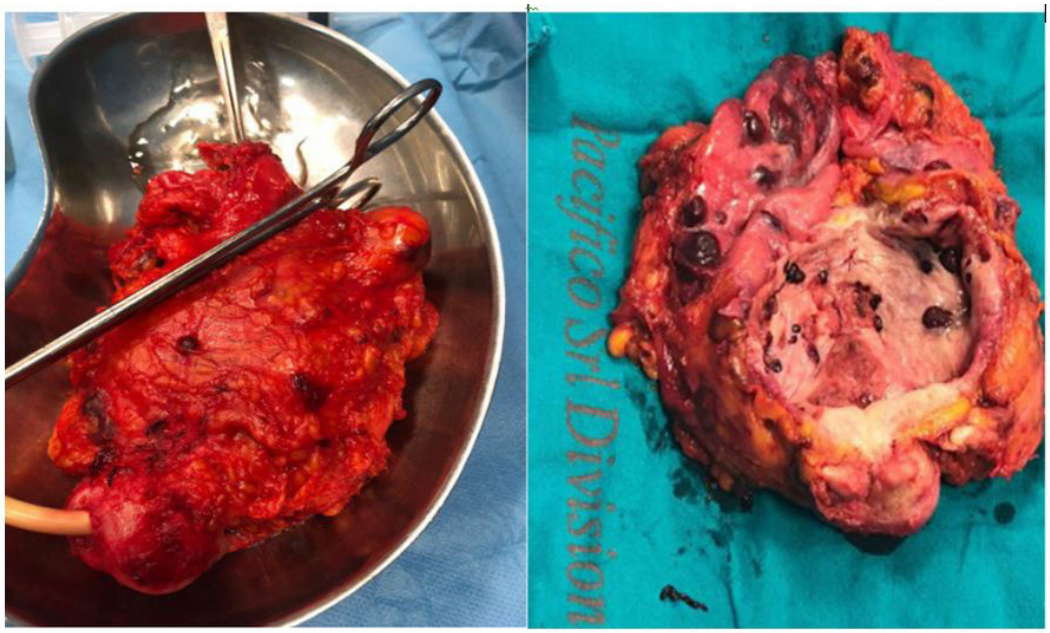
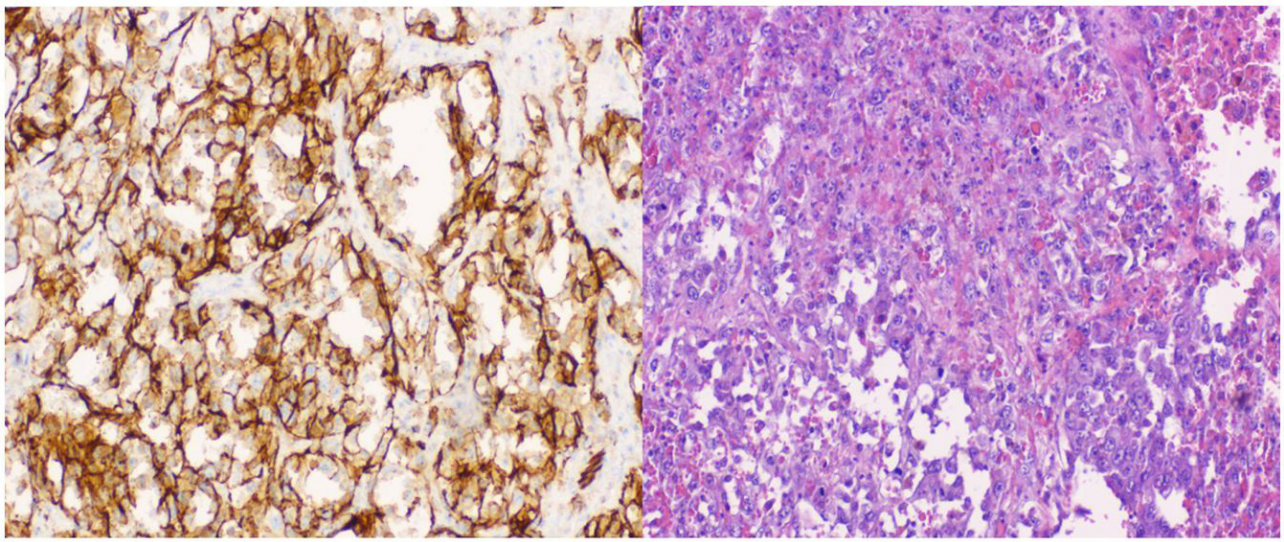
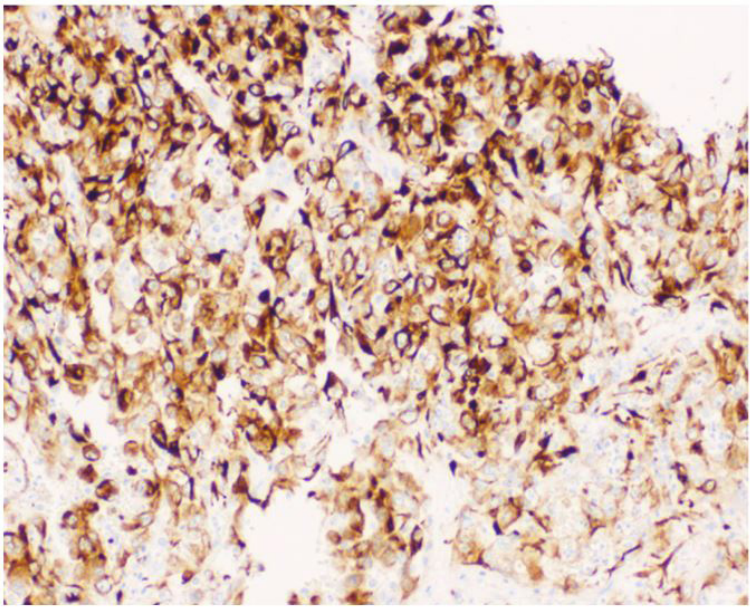


Figure 3: Radical cistectomy and extensive bilateral linphectomy





Discussion

Non-urothelial bladder tumors represent less than 5% of bladder tumors.

Angiosarcoma is a malignant tumor that mimics the morphology and function of endothelium. It can be primary or secondary to direct extension or metastasis. Primary urinary bladder angiosarcomas are very rare. Fourteen cases of primary urinary bladder angiosarcoma (PUBA) have been reported in the English literature [1,2].

PUBA occurs in males more often than females, with a ratio of 3:1. Patients commonly present with hematuria.

The etiology of PUBA is unknown. Radiotherapy and hemangioma are seen in association with PUBA.

Grossly, PUBA is described as large hemorrhagic mass with areas of necrosis.

Due to very few cases of PUBA, there is no consensus treatment algorithm. The primary treatment modalities consist of resection of the tumor with a radical cystectomy with lymph node dissection

followed by chemotherapy and radiation therapy is the recommended therapy [1,2]. In the literature, long-term survival is poor.

Factors that made the diagnosis of PUBA challenging in this case were the unusual clinical presentation and unusual histology.

Histologically, the tumor was poorly differentiated and was rich with osteoclast- like multinucleated giant cells. These cells were immunophenotypically similar to the tumor cells in the expression of vimentin, CD31, and factor XIII. The presence of osteoclast-like multinucleated giant cells in this case with the described immunophenotype is unique. Giant cells have been described in association with urothelial carcinoma in situ, invasive papillary carcinoma, high-grade infiltrating urothelial carcinoma, sarcomatoid carcinoma, and choriocarcinoma of the bladder [3]. The role of these cells in PUBA is not yet clear.

Conclusion

Primary urinary bladder angiosarcoma is a rare and an aggressive malignant neoplasm. It has a male predominance and commonly presents with hematuria. Exposure to radiotherapy is associated with PUBA in 38% of cases. The pathogenesis of PUBA is not yet understood.

For the pathologist, diagnosing this entity is challenging, especially in cases with atypical morphology. Pathologists should have a high index of suspicion in elderly men presenting with hematuria and a urinary bladder mass.

We believe mandatory describe every rare cases of urinary bladder angiosarcoma. More research is necessary to understand the role malignant cells in urinary bladder neoplasm in angiosarcoma in particular.

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