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Angiosarcoma of the Bladder: Review of the Literature and Discussion About a Clinical Case

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ABSTRACT

Our reported case is a 72 year-old man who presented with hematuria. A transurethral resection of the bladder tumor (TURB-T) has been performed. Histopathological diagnosis was an epithelioid angiosarcoma. CT scan revealed a bladder thickening. The treatment consisted in a complete pelvectomy with urinary and digestive diversion. Following the operation, the patient developed liver and pulmonary metastasis. He died 5 months after the initial diagnosis.

Introduction

Angiosarcoma is a vascular malignant tumor developed from blood vessels wall connective tissue. It is a rare neoplasia with bad prognosis and affects any organ. Bladder angiosarcoma is rare and is the subject of very few publications. Information about diagnosis, treatment, evolution and prognosis of this type of tumor are very limited. In this paper we report a 72-year-old patient case, harboring bladder angiosarcoma revealed by macroscopic hematuria.

Case presentation

The patient was 72-year-old man. He was admitted in emergency for a first episode of macroscopic hematuria complicated with acute urinary retention. His medical history was high blood pressure, a weaned smoking addiction, and benign prostatic hyperplasia (BPH). As a formal cinema industry worker, he had undergone chemical products exposure. At the first clinical examination, he presented with no pain, a supple abdomen without palpable mass. The biological checkup was normal. A bladder blood clot was seen on ultrasound. A cystoscopy was performed in emergency revealing a voluminous mass with a wide implantation located in the left side of the bladder. A TURB-T was performed. Pathological analysis reported an epithelioid bladder angiosarcoma (ISUP 2012 classification). The tumor was located in the submucosae bulging the normal urothelium.

On architectural analysis, tumor cells were undifferentiated; they contained abundant cytoplasm and large, irregular nuclei with many mitosis. Necrosis rate was superior to 50%. There was no vascular embolus (Fig. 1). Immunohistochemical (IHC) staining was positive for CD31 (Fig. 2), FLI-1, ERG (Fig. 2) and vimentin, but negative for CD34 and cytokeratin. Proliferation index with Ki67 staining was of 90%.

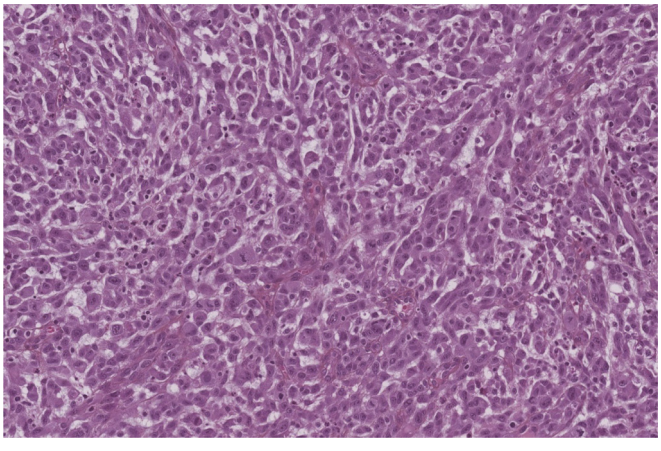

Figure 1. Malignant epithelioid cells, HES stain.

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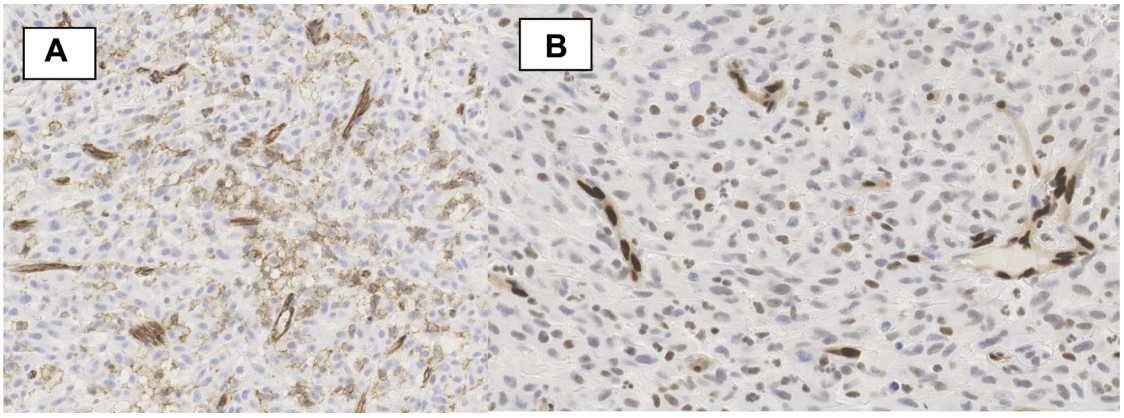

Figure 2. A: CD31 membranous staining of infiltrating tumor cells. We can observe also the normal vessel wall staining. B: Moderate ERG nuclear staining of tumor cells. We can also observe a stronger staining of normal endothelial capillary cells.

Despite a complete resection, a CT scan performed 3 weeks after TURB-T revealed massive local recurrence with a 42 * 37 mm mass (Fig. 3). No secondary lesion was diagnosed at this time.

For the treatment, the institutional sarcoma board has decided an initial external radiation therapy followed by surgery if the mass did not increase. After beginning the radiation therapy, the patient degraded. He presented a sepsis. The CT-scan showed an air bubble inside the bladder near the mass evocating tumor necrosis (Fig. 3).

Despite broad-spectrum intra-venous antibiotics, the patient declined indicating a surgical extirpation. A total pelvectomy with urinary and digestive diversion was performed 3 months after initial presentation.

After surgery, the patient presented an atrial fibrillation. One week after surgery, a CT-scan revealed liver and lung metastasis with peritoneal carcinomatosis.

The patient died 5 months after initial diagnosis.

Discussion

Non-urothelial bladder tumors represent less than 5% of bladder tumors. Bladder angiosarcoma was described for the first time in 1907 by Jungano.[1] Since that date, thirty-two cases had been reported in literature. Some cases are reported after radiation therapy for prostate or gynecologic cancer. Angiosarcoma following radiation therapy was previously described. Hematuria is always the first symptom (macroscopic or microscopic). Associated symptoms are dysuria, pain, obstruction, vaginal bleeding and weight loss. Men are more affected than women with a ratio of 1/5. Liver and lung are usual metastatic sites.

Pathological diagnosis can be difficult.[2] According to Matoso and Epstein, Angiosarcoma is confirmed if at least one endothelial marker including Factor VIII (F VIII), CD31, CD34 or ERG is positive on IHC.[3]

In our case, epithelioid bladder angiosarcoma diagnosis is founded on CD31, FLI-1 and ERG positive staining. CD34 and Cytokeratin are negative. A second expert opinion concluded in angiosarcoma or undifferentiated sarcomatoid carcinoma. An uropathologist board discussion decided that further molecular analysis would not have any value. In the literature, 27 cases are positive for at least one endothelial marker, including ERG (n = 5), F VIII (n = 12), CD31 (n = 20) and CD34 (n = 12). Five cases report no immunohistochemical analysis. No molecular analysis is reported (Table 1).

There is still no consensus on optimal treatment for bladder angiosarcoma. In our case, radiation therapy and radical pelvectomy was performed. Our patient had rapid disease progression after surgery; therefore he did not fit for adjuvant therapy. No significant superiority of any of those strategies has been reported.

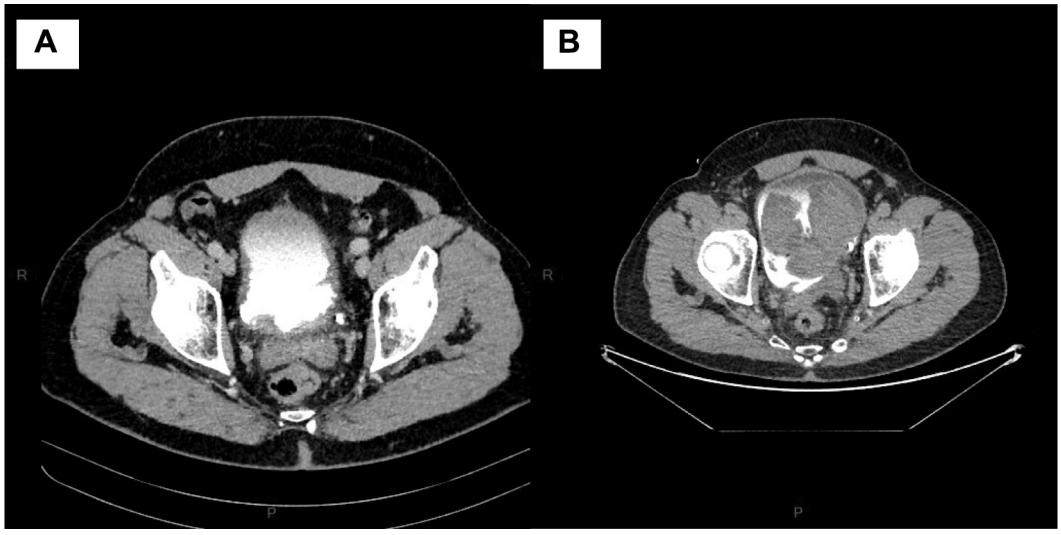

Figure 3. CT images show the rapid local progression of the tumor bladder. A: Day of diagnosis. B: 2 months after diagnosis.