



Oncology

A round cell sarcoma of the bladder following radiotherapy: A case Report

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ARTICLE INFO

Keywords:

Sarcoma

Bladder

Anterior pelvectomy

ABSTRACT

Sarcomas are uncommon and extremely aggressive malignancies. Sarcomas of the bladder are exceedingly rare. We report the case of 67-year-old female patient having had 20 years ago a total hysterectomy with bilateral adnexectomy followed by adjuvant external radiotherapy to an undifferentiated uterine sarcoma, was presented with hematuria. An abdomino-pelvic Computed Tomography (CT) Scan was done showing a 5 cm budding lesion of the bladder floor. An anterior pelvectomy with external urinary diversion was undergone. The histological examination showed a high grade urothelial carcinoma infiltrating the muscle with a sarcomatoid component.

1. Introduction

Sarcomas are uncommon and extremely aggressive malignant tumors. Although they can occur anywhere in the body and at any age, they are more frequently found in the skin of the head. Sarcomas of the bladder are extremely rare.¹ The association of uterine sarcoma and bladder sarcoma in a meta-chronous way is almost non-existent. In our knowledge, this is the first case that describes a uterine sarcoma followed by a bladder sarcoma in the same patient in the literature. They usually arise in adult patients with a history of radiation therapy, cigarette smoking, or exposure to chemical agents.

2. Case presentation

67-year-old female patient, asthmatic since childhood, exposed to wood smoke for 30 years, having had 20 years ago a total hysterectomy with bilateral adnexectomy followed by adjuvant external radiotherapy: 50gy divided over 25 sessions to an undifferentiated uterine sarcoma, was admitted in our department with hematuria, suprapubic pain, frequency and sensitivity of the left lumbar evolving for 2 months and vitals were stable. An abdomino-pelvic Computed Tomography (CT) Scan (Fig. 1) showed a 5 cm budding lesion of the bladder floor, which invaded the left ureteral meatus and left uretero-hydronephrosis without secondary lesions.

The patient had a transurethral resection of her tumor: it was a solid tumor with an infiltrating appearance, very bleeding on contact. The

pathological examination showed a high grade urothelial carcinoma infiltrating the muscle with a sarcomatoid component. Moreover, the patient developed a left pyonephrosis with multiresistant germ. It was controlled by percutaneous drainage by nephrostomy and imipenem for 14 days. An anterior pelvectomy with external urinary diversion: bilateral ureterostomy with bilateral ilio-obturator lymph node dissection was considered: the dissection was very difficult: there was a very significant pre-vaginal collateral venous circulation with very bleeding cardboard tissue in contact.

The postoperative course was simple and the histological examination showed an undifferentiated tumoral proliferation organized in clusters with large cells and on immunohistochemistry the cells were positive for vimentin: it was a high grade round cell sarcoma of the bladder according to FNCLCC, the surgical limits were microscopically positive (Fig. 2). The patient was referred to radiotherapy, where she had 5 adjuvant radiotherapy sessions and to oncology where she started cisplatin-based chemotherapy. 3 months later, a CT scan showed multiple metastases in the lungs, liver and lymph nodes with a large local recurrence. Unfortunately, the histological type of these secondary lesions is not known since they were not biopsied, but based on the chronology and the fact that the uterine sarcoma has been in remission for 20 years, these lesions were probably secondary localizations of bladder sarcoma. The patient died 1 month later.

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<https://doi.org/10.1016/j.eucr.2023.102402>

Received 19 February 2023; Received in revised form 5 April 2023; Accepted 12 April 2023

Available online 13 April 2023

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3. Discussion

Endometrial stromal sarcoma (ESS) is part of a rare and complex group of mesenchymal uterine neoplasms and represents 1% of all uterine malignancies.

ESS is the second most common mesenchymal uterine tumor after leiomyo-sarcomas and Figures 15% of malignant mesenchymal neoplasms of the uterus. It has been categorized into low-grade, high-grade, and undifferentiated uterine sarcoma by the World Health Organization.¹

In the case of undifferentiated uterine sarcoma, such as in our case, the standard treatment provides hysterectomy and bilateral salpingo-oophorectomy. Adjuvant external pelvic radiation seems to decrease loco-regional recurrence,² but it causes many early side effects such as radiation cystitis, and late ones: radiotherapy-associated tumors like bladder sarcoma. Bladder sarcomas are rare, accounting for 1–2% of all malignant genitourinary tumors.³ Round cell sarcoma of the bladder is an exceedingly rare neoplasm, in our knowledge this case is among the rare cases described in the literature.

In 19 patients with primary bladder sarcomas reported by Spiess et al., leiomyo-sarcoma was the most frequent histology.⁴ Bladder sarcoma usually arises in adult patients with a history of radiation therapy, cigarette smoking, or exposure to chemical agents (vinyl chloride).

Our patient was exposed to wood smoke for 30 years and had undergone pelvic radiotherapy 20 years ago.

Major presenting symptoms observed are hematuria, dysuria, fatigue, lymphedema, abdominal lump, and frequency. Diagnostic confirmation is based on cystoscopy with histological examination: endoscopically the sarcoma has no particular appearance but histopathologic examination revealed a malignant small round cell tumor with a typical immunoprofile and genetic profile in most cases.

A round cell sarcoma must be distinguished from other primary and metastatic tumors with similar morphology, including other types of high-grade neuro-endocrine carcinomas (small cell carcinoma) that are rare in the bladder.⁵

Given to the rarity of these tumors, no consensus on optimal treatment has been reached. Sarcomas are usually treated with a multimodal approach combining surgery, radiotherapy, and chemotherapy. Regardless of histologic subtype, the scientific literature on bladder

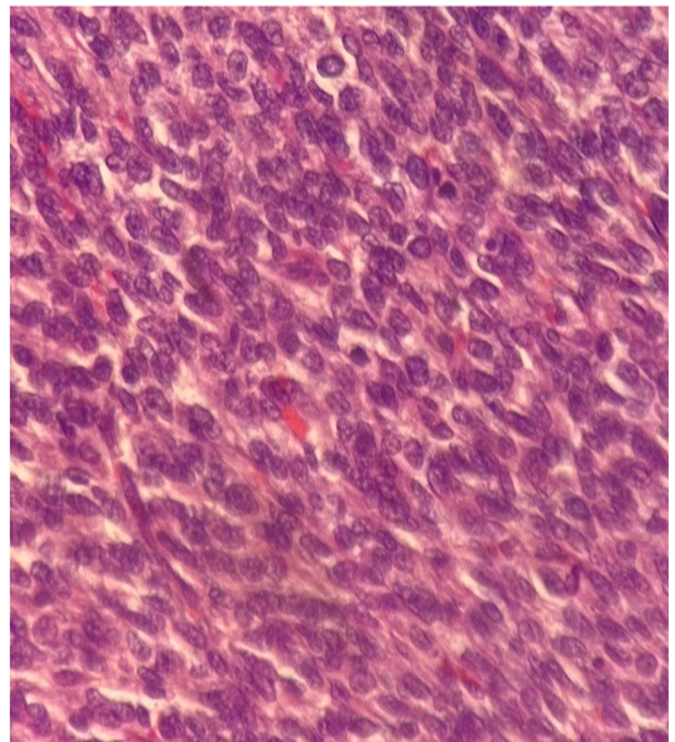


Fig. 2. Histological examination showed an undifferentiated tumoral proliferation organized in clusters with large cells and on immunohistochemistry the cells were positive for vimentin: it is a high grade round cell sarcoma of the bladder according to FNCLCC, the surgical limits were microscopically positive.

sarcoma has underlined the poor prognosis associated with these tumors.

In the series reported by Spiess et al., of ten patients undergoing therapy, nine died with a median survival of 20 months.⁴

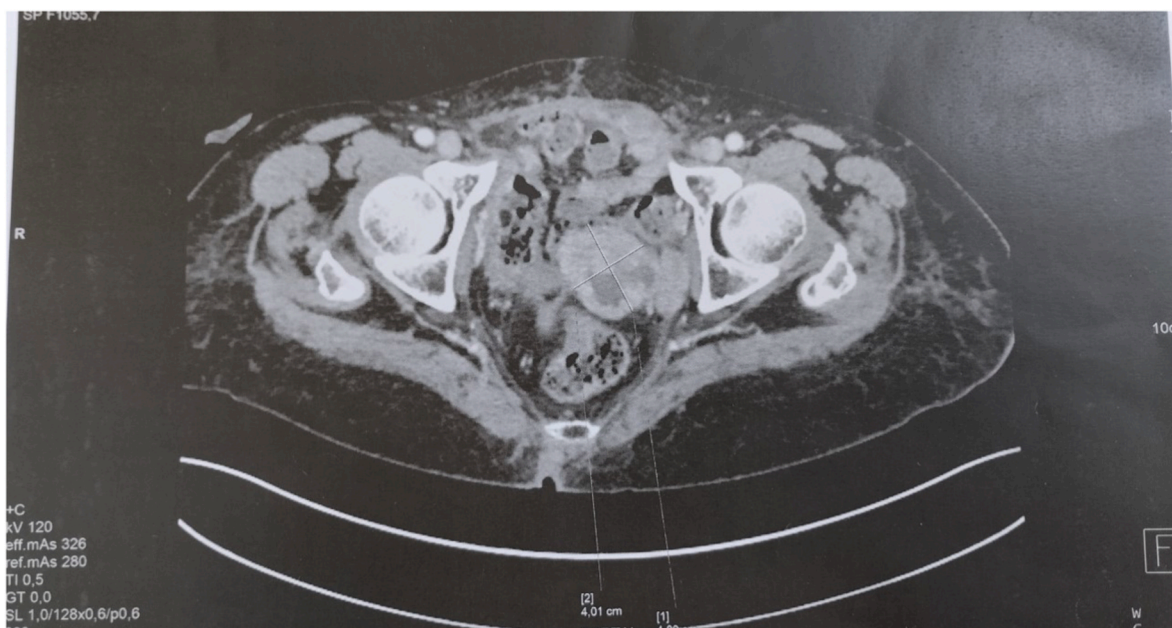


Fig. 1. An abdomino-pelvic Computed Tomography (CT) Scan was done showing a 5 cm budding lesion of the bladder floor which invaded the left ureteral meatus and left uretero-hydronephrosis without secondary lesions.

4. Conclusion

Round cell sarcoma of the bladder is a very rare but serious neoplasm with poor prognosis. It can be primitive or radiotherapy-associated tumors. We reported the case of a round cell sarcoma of the bladder which occurred in a patient already operated for a uterine sarcoma and who had external radiotherapy.

The diagnostic can only be established by pathological examination. The treatment must be multimodal.

Declaration of competing interest

The authors declare that they have no competing interests.

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