Bladder preserving approach for liposarcomatoid variant of transitional urothelial carcinoma

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Cite as: *Can Urol Assoc J* 2015;9(11-12):E886-9. http://dx.doi.org/10.5489/cuaj.3167
Published online December 14, 2015.

Abstract

Urothelial carcinoma of the bladder with variant histologies is usually underdiagnosed and understaged. It is found in <25% of cases during transurethral resection of bladder tumours. The majority of cases carry a worse prognosis, with higher rates of recurrence and progression, and should be managed aggressively. Here, we report a rare case of liposarcomatoid variant of urothelial carcinoma managed by partial cystectomy and standard pelvic lymph node dissection.

Introduction

Urothelial carcinoma of the bladder with variant histologies is usually underdiagnosed and understaged. The role of transurethral resection of bladder tumours is crucial for proper staging and grading of urothelial carcinoma. In large case studies, it is shown that around 25% of transurethral resection of bladder cases show some form of variant histology.[1] Identification of variant histology depends on the amount and quality of issue sampled during transurethral resection of a bladder tumour (TURBT), as well as on the experience of the uropathologist.

Most of the variant histology, which includes micropapillary, plasmacytoid, and sarcomatoid, carries poor prognosis[2] with higher rates of recurrence and progression. Sarcomatoid variant of urothelial carcinoma, formerly known as carcinosarcoma , may occur in 0.6% cases of bladder cancer and usually presents in advanced stage. Less than 200 cases of sarcomatoid variant of urothelial carcinoma have been reported in the literature. One common caveat is that early radical cystectomy should be considered in all cases of highly aggressive variant histology, like micropapillary, sarcomatoid, and plasmacytoid carcinoma.

Here, we present a rare case of liposarcomatoid variant of urothelial carcinoma managed by partial cystectomy and extended pelvic lymph node dissection.

Case report

A 72-year-old man with no comorbidity presented with history of intermittent, painless, gross hematuria with clots. He underwent transurethral resection (TUR) biopsy outside the hospital that showed T1 high-grade urothelial carcinoma; deep muscle was not included. General and systemic physical examinations were normal. Routine investigations were within normal limits. The patient underwent abdomen computed tomography (CT), showing residual bladder thickening in the anterior wall with no pelvic lymphadenopathy (Fig. 1). Following that, the patient underwent completion TURBT, which showed 2x2 cm solid growth in the anterior wall and dome region.

Histopathologically, it turned out to be liposarcomatoid variant of transitional carcinoma, a biphasic tumour composed of transitional cell carcinoma and sarcomatous component. The carcinomatous component was present focally and showed features of high-grade transitional cell carcinoma (Fig. 2a). The sarcomatous component was composed of heterologous elements and showed features of liposarcoma. There were univacuolated lipocytes mixed with many multivacuolated lipoblasts (Fig. 2b). The sarcomatous component was sharply demarcated from the carcinomatous area. The carcinomatous component showed strong positivity for cytokeratin (CK) and CK 7, whereas the sarcomatous component was negative (Fig 2c). The sarcomatous component showed positivity for S-100. Alcian blue stain did not reveal any mucin within the tumour cells. The tumour was diffusely infiltrating the lamina propria, but deep muscle was free.

In view of the aggressive histological variant and normal metastatic workup, the option of early radical cystectomy was offered to the patient, but he wanted bladder preservation. Cystoscopy prior to partial cystectomy showed only a

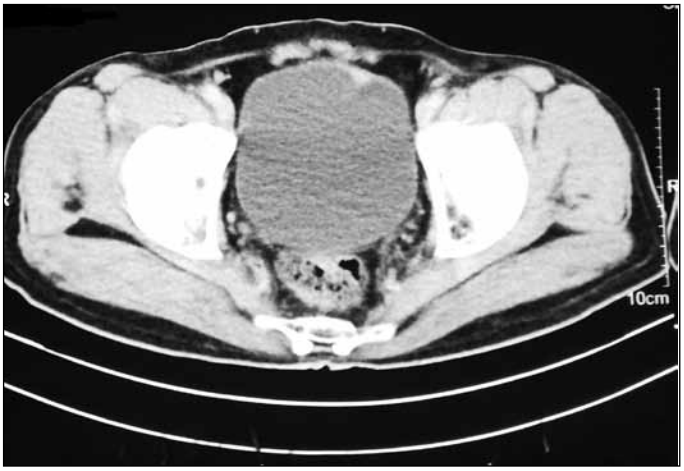

Fig. 1. Abdomen CT showing residual bladder thickening in the anterior wall with no pelvic lymphadenopathy.

scar of 2x2 cm at the dome and anterior wall of bladder. The patient underwent partial cystectomy by taking a 2 cm margin and standard bilateral pelvic lymph node dissection. Postoperative course was uneventful. Final histopathology showed no residual tumour. All lymph nodes were free of tumour (0/18).

The patient is doing well, without any recurrence, for the past year and a half, with regular followup cystoscopy, urine cytology, and CT scan (Fig 3a, 3b).

Discussion

Sarcomatoid variant of urothelial carcinoma is rare and usually presents in people between the ages of 50 and 77.[3] The most common presentation is painless, gross hematuria that may or may not be associated with lower urinary tract symptoms. Generally, it presents at a high-stage and grade with both epithelial and mesenchymal (sarcomatous) elements.[1] The carcinomatous component is predominantly high-grade urothelial carcinoma, followed by glandular and small cell component. The sarcomatous component is usually composed of undifferentiated spindle cell neoplasm.[4] However, in rare cases, areas of osteosarcoma, chondrosarcoma, rhabdomyosarcoma, liposarcoma, angiosarcoma, or a mixture of sarcoma histologies (in decreasing order of frequency) may be seen in sarcomatoid urothelial carcinoma.[5] Histological differentiation from true sarcoma is difficult and immunohistochemistry is helpful in this regard.

The histological differential diagnosis lies between a lipoid cell variant of transitional cell carcinoma (TCC), pure liposarcoma of urinary bladder, and sarcomatoid TCC with liposarcomatous component (Table 1). In a pure liposarcoma, there should not be any epithelial component diagnosed by CK negativity. Another rare variant of urothelial carcinoma is the lipoid cell subtype described in only 33 patients, which has a very poor prognosis.[2,6,7] In lipoid cell variant of urothelial carcinoma, the vacuolated cells show CK and CK7 positivity (Table 1), indicating their epithelial nature.[6,7] The lipoid cell population typically composes 10–50% of the tumour, with the remainder being conventional urothelial carcinoma or another rare subtype, including plasmacytoid and micropapillary carcinoma.[7,8]

In view of the absence of randomized, controlled trials and low incidence, there is no standard treatment protocol for this disease. To our knowledge, no published articles have focused on treatment guidelines for sarcomatoid variant of urothelial carcinoma according to stage, although one group suggested that radical cystectomy rather than intravesical therapy should be the preferred option of treatment for patients with stage T1 disease.[9,10]

Overall, prognosis of sarcomatoid carcinoma is poor, with a five-year survival of 20.3% after radical cystectomy.[11] In their series, Wang et al[12] suggested that although presented with a higher stage and grade (85% muscle invasive and 50% metastatic at presentation), long-term survival is possible in patients treated with multimodality therapy; however, the optimal treatment modality has yet to be defined.

Literature regarding nonsurgical therapy for these bladder tumours is scarce. One of the case reports of metastatic sar-

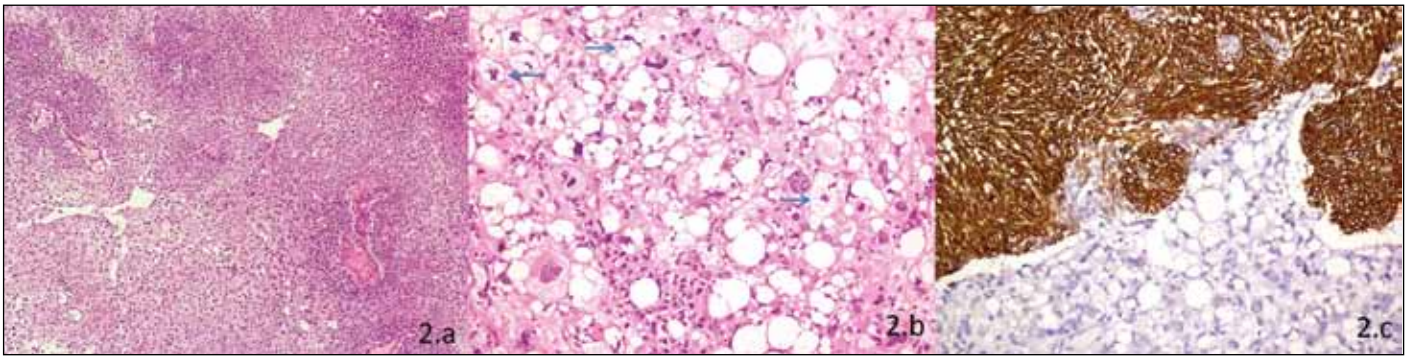

Fig. 2. (a) Photomicrograph showing carcinomatous component of the tumour in form of high-grade urothelial carcinoma arranged in form of fused papillae and sheets (HE, X100); (b) Sarcomatous component of the tumour is composed of lipocytes and many multivacuolated lipoblasts [arrow] (HE, X200); (c) The carcinomatous component shows strong cytokeratin 7 expression (upper part), whereas the liposarcomatous component is completely negative (lower part) (IP, X200).

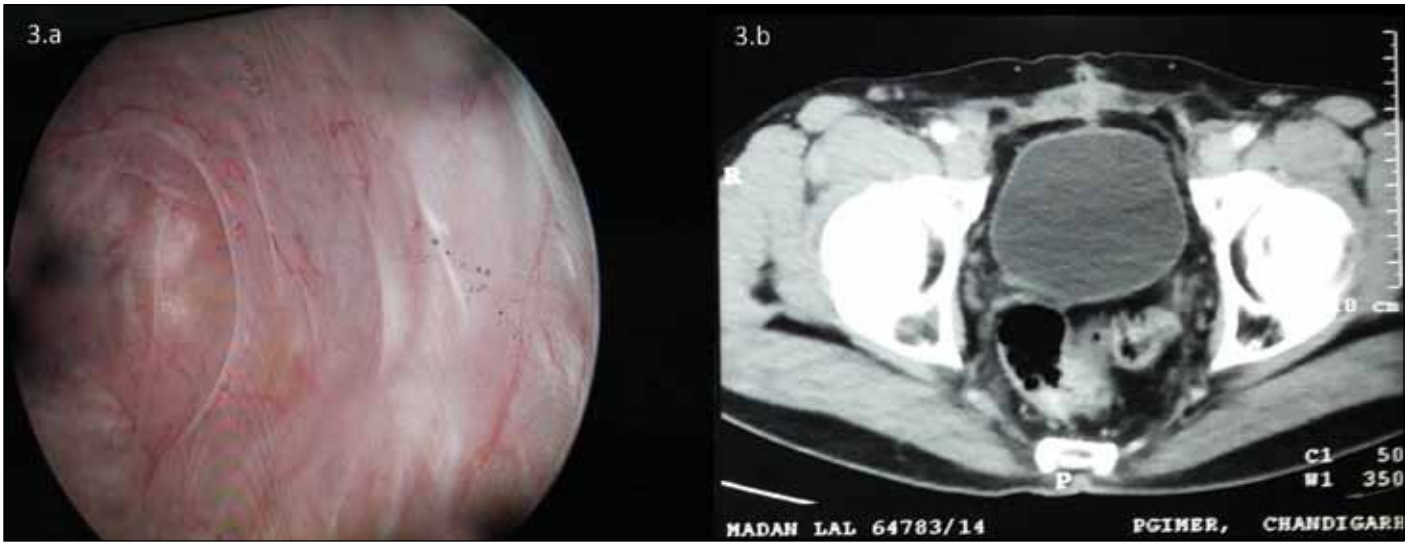

Fig. 3. (a) Followup cystoscopic image showing previous operative scar with no residual tumour; (b) Followup CT image showing normal bladder outline with no residual tumour.

comatoid carcinoma demonstrated complete remission after cisplatin and gemcitabine chemotherapy while the other described complete remission with neoadjuvant chemotherapy followed by partial cystectomy.[13,14]

In our index case, since the patient presented with early-stage, solitary lesion at the dome and refused radical cystectomy; partial cystectomy had been done. In the final histopathology, there was no residual tumour and all lymph nodes were negative. The patient is doing well over one year and a half year of follow, up without any recurrence. To the best of our knowledge, only two previous cases of liposarcomatoid variant of TCC has been reported in the literature.[15,16] Although such variants need aggressive management, in patients with early presentation , a bladder-preserving approach can be considered.

Conclusion

Though sarcomatoid variant of urothelial carcinoma is associated with inferior outcome compared to conventional urothelial carcinoma, our case is one of the rare liposarcomatous variant of urothelial carcinoma with comparatively good prognosis managed by partial cystectomy and standard pelvic lymph node dissection alone. Further cases of this type are required to be reported for better understanding of the biological behaviour of this tumour.

Competing interests: The authors declare no competing financial or personal interests.

This paper has been peer-reviewed.

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Table 1. Immunohistological differentiation between histological variants

	PAN-CYTOKERATIN	ACTIN DESMIN	P63, CK5/6	S-100
Sarcomatoid carcinoma	Positive	Positive, rarely	Positive (up to 40%)	
Leiomyosarcoma	Rarely positive	Positive, extensively	Negative	
Lipoid urothelial carcinoma	CK 7, CK 20 Positive	Negative	Negative	
Our case (Liposarcomatoid variant)	Positive	Negative	Negative	Positive

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