

## Sarcomatoid Carcinoma of the Urinary Bladder

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**Abstract:** Sarcomatoid carcinoma of the urinary bladder is a rare aggressive tumour. This rare biphasic malignant lesion shows morphological and immune-histochemical evidence of both epithelial and mesenchymal differentiation with poor prognosis. Aggressive therapy with radical cystectomy followed by adjuvant chemotherapy and radiotherapy is the preferred mode of treatment. Here we report a case of sarcomatoid carcinoma of urinary bladder in a 58-year-old male along with review of relevant literatures.

**Keywords:** Sarcomatoid carcinoma, urinary bladder, malignant lesion, cystectomy.

### INTRODUCTION

Sarcomatoid urothelial carcinoma is a rare variety of urothelial carcinoma with aggressive behavior[1-5]. Though radical surgery is the mainstay of treatment[2] but surgery with adjuvant chemotherapy or radiotherapy also gives encouraging results[1,3,4]. Due to rarity of cases definite treatment strategies has not been established till now[5]. Here we report a case of sarcomatoid urothelial carcinoma with unfavorable outcome.

### CASE REPORT

A 58-years-old male presented with frequency, urgency, dysuria and multiple episodes of painless, gross hematuria. On Ultrasonography a large growth was found in the right lateral wall of the urinary bladder. CECT revealed a polypoidal mass lesion in right lateral wall occupying the whole of the bladder with multiple regional lymph nodes and right sided hydroureteronephrosis(Fig.-1).



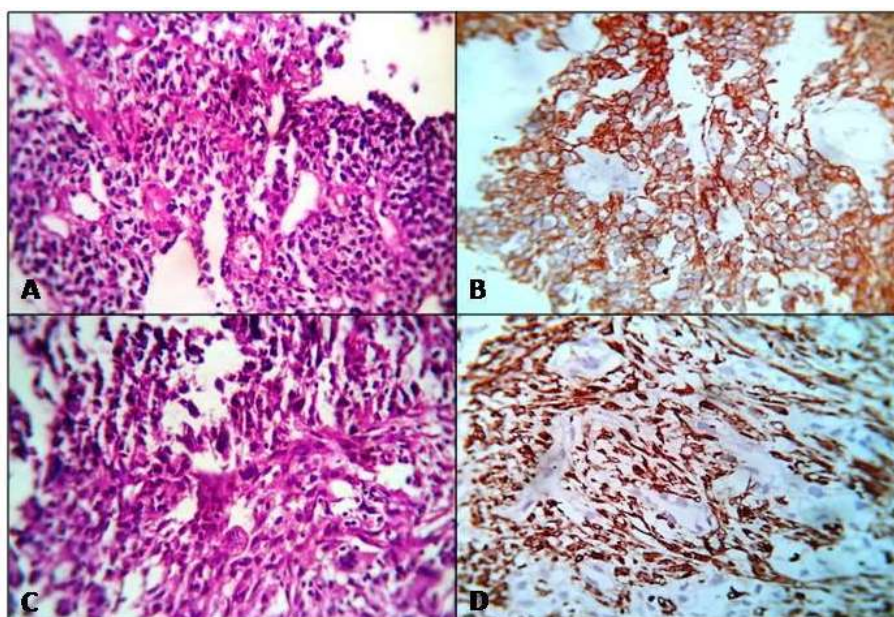
**Fig-1. CECT showing a large intravesical growth occupying most of the bladder with multiple lymph nodes and right sided hydroureter.**

Metastatic work up revealed no distant metastasis. Cystoscopy showed a large broad based growth arising from the right lateral and posterior wall

of the bladder. Transurethral resection of bladder tumour was attempted with incomplete resection. Histopathological examination revealed the tumour

composed of predominantly pleomorphic spindle cells with high nuclear cytoplasmic ratio infiltrating all the layers of muscles with areas of necrosis and foci of transitional cell carcinoma (Fig.-2 A&C). Immuno-

histochemistry showed positivity with Cytokeratin 7 and Vimentin (Fig.-2 B&D) consistent with the histological diagnosis of sarcomatoid (SC) variant of transitional cell carcinoma (TCC).



**Fig-2: Histopathological picture showing-**

- A. Foci of conventional papillary TCC (H&E 40X).**
- B. Immuno-histochemistry with Cytokeratin-7 showing positivity in the TCC component (40X).**
- C. Pleomorphic spindle cells in the sarcomatoid component (H&E 40X).**
- D. Immuno-histochemistry with vimentin showing positivity in the sarcomatoid component (40 X)**

Radical cystectomy with ileal conduit was planned. But the patient refused any further surgical treatment. Chemotherapy was started with gemcetabine and cisplatin, but he was lost for follow up after a single dose of chemotherapy. He was again admitted within six months with pathological fracture of L2 vertebrae and bilateral plural effusion due to metastasis and died within 4 days.

## DISCUSSION

Sarcomatoid carcinoma (SC) of the urinary bladder is an aggressive rare biphasic malignancy with poor prognosis [1, 2, 3]. Its rarity may be known on an analysis of Surveillance, Epidemiology, and End Result (SEER) database mentioning the incidences of this tumour 0.2% to 4.3% [4]. Though smoking has been said to be associated with urothelial carcinoma but definite association of smoking with sarcomatoid TCC is still lacking. Genetic and molecular studies suggested for a common monoclonal cell origin for the epithelial and mesenchymal component of this rare malignant tumour [4, 6]. Some investigators suggested that these tumors develop as a result of undifferentiated, totipotent neoplastic cells that undergo multiple pathways of terminal differentiation into either mesenchymal or epithelial elements[4,6]. Previous history of radiation and intravesical cyclophosphamide chemotherapy may be associated with the development of sarcomatoid

urothelial carcinoma [1,4,5,6]. The disease normally presents itself in younger age group[4] with advanced stage and more aggressive nature than usual urothelial carcinoma[4,6]. Haematuria, dysuria, nocturia, urinary retention and lower abdominal pain are the most commonly reported signs and symptoms [4, 5]. In most of the reported cases tumours are found mainly in the base and trigonal area.

Macroscopically they are intraluminal polypoidal lesion. Microscopically the sarcomatous component is composed of haphazardly arranged sheets of pleomorphic spindle cells exhibiting hyperchromatic nuclei with moderate amount of cytoplasm infiltrating into the muscle layers. The spindle cell component is usually of high grade with nondescript architecture (often resembling malignant fibrous histiocytoma) [6]. Heterologous differentiation may be in the form of osteosarcoma, rhabdomyosarcoma, chondrosarcoma or leiomyosarcoma and the epithelial component may be urothelial, glandular, or small cell component [6]. 30% SC may harbor carcinoma in situ [4]. In our case also the tumour was composed of pleomorphic spindle cells with focal areas of conventional TCC without any heterogeneous differentiation. By immuno-histochemistry, the carcinomatous component is universally positive for epithelial markers EMA and pan-keratin. In contrast, the sarcomatous component

reacts with vimentin only. In the present case cytokeratin-7 and vimentin was positive. It may express specific markers for specific mesenchymal differentiation, if present [6]. The histologic differential diagnosis includes various types of sarcoma, reactive pseudosarcomatous mesenchymal proliferations, and malignant melanoma. Distinguishing these masses from sarcomas as leiomyosarcoma, pleomorphic fibrosarcoma, or malignant fibrous histiocytoma may be difficult but sarcomas usually do not stain for epithelial markers and lack desmosomes or tonofilaments on electron microscopy [2]. Benign pseudosarcomatous mesenchymal proliferations in the bladder include pseudosarcomatous myofibroblastic proliferations, postoperative spindle cell nodules, inflammatory pseudotumors, and pseudosarcomatous fibromyxoid tumors. These pseudosarcomatous tumors are immunoreactive for vimentin and muscle actin, although aberrant expressions of cytokeratins and EMA have been reported. Histologically, these reactive lesions show neither atypical mitosis nor significant cytologic atypia, whereas sarcomatoid carcinoma exhibits these atypical features with tumor necrosis, invasive tumor margins, and high cellularity [3, 4].

Still now due to lack of randomized controlled trial no definite mode of treatment has been formulated. Radical cystectomy has been the preferred treatment in most of the reported cases. Margin negativity and absence of metastasis at presentation may be associated with better prognosis. Cancer-specific survival was significantly better for those who underwent cystectomy instead of transurethral resection[1]. Most patients died as in our case due to rapid metastasis in lymph nodes, lung, pleura, brain, liver and bones[6]. Sarcomatoid carcinomas of the urinary bladder are rare and highly aggressive neoplasm usually diagnosed at advanced stage with a median survival of 10 months[3]. Though radical cystectomy is the treatment of choice [6] but now chemotherapy or chemo radiation have been shown to give favorable tumour response[1,2,3,4,5] in a native bladder leading to improved quality of life[7]. Chemo radiation is a reasonable alternative showing promising results and cystectomy should be reserved

for salvage operation[7]. Two reported case showed a complete durable remission with cisplatin and gemcitabine[5,7] and another report showed a complete remission using neoadjuvant chemotherapy with carboplatin and gemcitabine followed by partial cystectomy[3]. A multimodality approach with radical cystectomy followed by adjuvant chemotherapy and radiation should be considered for better oncological outcome[1,3,4,5]. A multicentre trial of treatment option of this rare tumour is needed to arrive at a conclusive opinion for the best treatment option[4,5].

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