

Sarcomatoid Carcinoma of the Urinary Bladder- A Rare Entity

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ABSTRACT:- Sarcomatoid carcinoma of the urinary bladder, a rare neoplasm is composed of adenocarcinoma as well as sarcoma components. Its histogenesis and biological behaviour are controversial. Literature has documented approximately 70 cases in the form of case reports or small series. Herein, we report a case of sarcomatoid carcinoma of the urinary bladder in a 51-year-old male having a history of painless, gross hematuria with clots. The early postoperative period was uneventful and the biochemical and imaging findings were good 7 months postoperatively. However, the patient was lost for follow-up.

Key words:- sarcomatoid, carcinoma, urinary, bladder

I. INTRODUCTION

Sarcomatoid carcinoma, a rare neoplasm of the urinary bladder is defined by the World Health Organization as a biphasic tumor consisting of malignant epithelial and mesenchymal components (1). Literature has documented approximately 70 cases in the form of case reports or small series (1). A series of 221 cases from the Surveillance, Epidemiology and End Results (SEER) Program database has been documented (2). It is frequently observed in males, in the seventh decade, with the male/female ratio being 2:1 (1). Common symptoms are gross hematuria and dysuria. Advanced disease is seen in about 70% of cases and has a bad prognosis compared to regular urothelial carcinoma (3). A monoclonal and a multiclonal stem cell origin for the epithelial and mesenchymal components is hypothesized (4,5). Cyclophosphamide therapy and radiotherapy to the bladder play a role in its development (1,4). Here we report a case of sarcomatoid carcinoma of the urinary bladder in a 51 year old male.

II. CASE REPORT

A 51-year old, male patient was brought to our tertiary care hospital with complaints of painless, gross hematuria with clots since 3 months. He was a known case of carcinoma of bladder for which transurethral resection was done twice and diagnosed to be transitional cell carcinoma only two months back. On admission, all his vitals were stable and no abnormality was detected on systemic examination. Laboratory investigations revealed neutrophilic leucocytosis with moderate anemia (hemoglobin- 8 gm%) and adequate platelets on complete blood count. The renal function tests were impaired with raised values of urea (50 mg %) and creatinine (2.9 mg/ml). However, the liver function tests were normal. Abdomino-pelvic computerized tomography (CT) showed an enhancing lobulated, polypoid bladder mass measuring 6.0cm x 5.6cm, growing along the base and posterior wall, involving both vesico-uretric junctions causing bilateral hydronephrosis & hydroureter. The mass was seen extending to lower part of left ureter. A left nephrectomy with radical cystectomy was done. Additionally, inguinal lymph node dissection was performed.

We received specimen comprised of cystectomy, left nephrectomy and bilateral inguinal lymph nodes. The kidney was contracted and showed dilated pelvicalyceal system. The urinary bladder measured 15cm x 11cm x 7cm. On cutting open, a mass measuring 7cm x 4cm x 3cm was seen. It was polypoidal and ulceroproliferative (Figure 1). Grossly, areas of necrosis, ulceration and muscle invasion were seen. Six right and four left inguinal lymph nodes were dissected. All lymph nodes were sub-centimeter in size. Microscopically, the bladder tumor showed a biphasic pattern, consisting of urothelial carcinoma in-situ and sarcomatous component (spindle-cell) (Figure 2). The epithelial component showed papillary and glandular pattern (Figure 3) whereas the sarcomatous component was made of spindle cells having pleomorphic, vesicular nuclei, prominent nucleoli & eosinophilic cytoplasm, arranged in fascicles and bundles. Few bizarre cells and dedifferentiated cells were

also seen (Figure 4). The tumour was seen invading into deep half of muscularis propria reaching up to the perivesical fat. All surgical margins, both ureters and lymph nodes were free of tumor. A diagnosis of sarcomatoid carcinoma of the urinary bladder was given. Histopathological examination of the left kidney showed features of chronic pyelonephritis with hydronephrosis.

On immunohistochemistry, epithelial membrane antigen (EMA) and pancytokeratin were reactive in epithelial as well as mesenchymal components of the tumor (Figure 5a & 5b). Mesenchymal markers, smooth muscle actin (SMA) and desmin were weakly positive in the spindle-cell component (Figure 5c & 5d).

The early postoperative period was uneventful and the biochemical and imaging findings were good 7 months postoperatively. However, the patient was lost for follow-up.

III. DISCUSSION

Sarcomatoid carcinoma, a rare neoplasm of the urinary bladder, is also known as carcinosarcoma, spindle-cell carcinoma, metaplastic carcinoma, and malignant mixed mesodermal tumour. It is defined by the World Health Organization as a biphasic tumor consisting of malignant epithelial and mesenchymal elements (2). Originally termed carcinosarcoma because of the combination of both epithelial- and mesenchymal-type component, it was initially described by Dent in 1955 (6).

More recently, the term sarcomatoid carcinoma has been used for this neoplasm, since the sarcomatoid features are thought to be derived from dedifferentiation of the carcinomatous component (7).

Literature has documented approximately 70 cases in the form of case reports or small series (1). A series of 221 cases using the Surveillance, Epidemiology and End Results (SEER) Program database has been documented (2). In 1856, Ordonez recorded the first report of a malignant bladder tumour containing elements of cartilage or bone (8).

The histogenesis of carcinosarcomas is controversial [9]. Loss of heterozygosity in stem cells is the main factor in histogenesis (4, 5). Some investigators suggested an origin from undifferentiated, totipotent neoplastic cells that differentiate into mesenchymal or epithelial elements. This is reinforced by the presence of epithelial markers (cytokeratin or EMA) in mesenchymal areas and the presence of ultrastructural features (desmosomes or tonofilaments) of epithelial differentiation in sarcomatoid areas (10). Some believe that they are true "collision" tumors, with malignant mesenchymal and epithelial components arising independently (2). A monoclonal origin has also been suggested (2, 5, 11).

Smoking, previous radiotherapy or chemotherapy are considered etiological (1, 9).

Clinically, they are seen frequently in older males, are rapidly growing and present at advanced stage as polypoid neoplasms (2, 9).

The median age at diagnosis is 75 years, the range being 41 to 96 years. Male to female ratio is 1.9-2:1 (1, 2).

The most common location is the lateral wall of the bladder, with the anterior and posterior walls, dome, and trigone being less common (1, 2). Some authors noted trigone as a frequent location, evidence of Wolffian body origin (1). In our case, the tumour was located on both the lateral walls extending onto the posterior wall of the bladder.

As seen in other bladder cancers, patients present with painless gross hematuria, dysuria, increased micturition, as well as obstructive symptoms (1, 2, 9). In our case, the patient was admitted with painless gross hematuria.

Grossly, the tumors are large, polypoid, or nodular (1, 2). Microscopically, they are biphasic with an intimate mixture of carcinomatous and sarcomatous elements. The epithelial component shows a high-grade urothelial carcinoma with epidermoid and/or glandular differentiation. Common sarcomatous elements are fibrosarcoma, chondrosarcoma, osteosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma or leiomyosarcoma (1, 2, 9). In our case, histopathological examination showed a moderately differentiated urothelial carcinoma with focal glandular differentiation and a spindle-cell sarcomatous component. No heterologous element was seen.

Bladder sarcomatoid carcinomas are aggressive tumors. The main treatment methods are radical cystectomy and lymph node dissection (1, 2, 9). Aggressive surgery during early period is considered curative (9). Recently, radical cystoprostatectomy and lymph node dissection with a combination of neoadjuvant/adjuvant chemotherapy and/or radiotherapy has been advocated for better survival (1, 2, 9) and the one, five, and 10 years survival rates are 53.9%, 28.4%, and 25.8%, respectively (9).

IV. CONCLUSION

In summary, sarcomatoid carcinomas of the urinary bladder are rare, complex, highly malignant and present at an advanced stage in elderly men (2). Large-scale studies will help in understanding their biological behavior, prognosis and to develop novel focussed therapies (1, 2, 9, 12).

REFERENCES

- [1]. DoğanAtılğan and Yusuf Gençten. Carcinosarcoma of the Bladder: A Case Report and Review of the Literature. *Case Reports in Urology*. 2013; 2013:1-3 <https://doi.org/10.1155/2013/716704>.
- [2]. Wang J, Wang FW, LaGrange CA, Hemstreet III GP, Kessinger A. Clinical Features of Sarcomatoid Carcinoma (Carcinosarcoma) of the Urinary Bladder: Analysis of 221 Cases. *Sarcoma*. 2010; 2010:454792. doi:10.1155/2010/454792. 2010. PMID: 20706685 DOI:10.1155/2010/454792
- [3]. Wright JL, Black PC, Brown GA, Porter MP, Kamat AM, Dinney CP, et al . Differences in survival among patients with sarcomatoid carcinoma and urothelial carcinoma of the bladder. *J Urol*. 2007; 178: 2302-2307. PMID:17936803 DOI:10.1016/j.juro.2007.08.038
- [4]. Mukhopadhyay S, Shrimpton AE, Jones LA, Nsouli IS, Abraham NZ Jr. Carcinosarcoma of the urinary bladder following cyclophosphamide therapy: evidence for monoclonal origin and chromosome 9p allelic loss. *Arch Pathol Lab Med*. 2004;128: e8-11. PMID:14692829 DOI:10.1043/1543-2165(2004)128<e8: COTUBF>2.0.CO;2
- [5]. Halachmi S, DeMarzo A M, Chow N H, Halachmi N, Smith A E, Linn JF, et al. Genetic alterations in urinary bladder carcinosarcoma: Evidence of a common clonal origin. *European Urology*. 2000; 37: 350-357. DOI: 10.1159/000052369
- [6]. Dent ED Jr. Carcinosarcoma (collision tumor) of the urinary bladder. *J Urol*. 1955;74:104–108.
- [7]. Sanfrancesco JM, McKenney JK, Leivo MZ, Gupta S, Elson P, Hansel DE. Sarcomatoid Urothelial Carcinoma of the Bladder: Analysis of 28 Cases With Emphasis on Clinicopathologic Features and Markers of Epithelial-to-Mesenchymal Transition. *Arch Pathol Lab Med*. 2016;140:543-51. doi: 10.5858/arpa.2015-0085-OA.
- [8]. M. E. C. Ordonez. 2° transformation cartilagineuse de la vessie chez un vieillard. *Gazette Médicale de Paris*. 1856; series 3, vol. 11, article 824.
- [9]. Basibuyuk I, Ramazan T, Fatih E. Bladder carcinosarcoma: A case report with review of the literature. *Archivio Italiano di Urologia e Andrologia*. 2017;89: 240-242. DOI: 10.4081/aiua.2017.3.24
- [10]. Volker HU, Zettl A, Georg S, Heller V, Heinrich E, Rosenwald A. Molecular genetic findings in two cases of sarcomatoid carcinoma of the ureter: evidence for evolution from a common pluripotent progenitor cell? *Virchows Arch*. 2008; 452: 457–463. PMID: 18266004 DOI: 10.1007/s00428-008-0583-z
- [11]. Armstrong AB, Wang M, Eble JN, MacLennan GT, Montironi R, Tan PH, et al. TP53 mutational analysis supports monoclonal origin of biphasic sarcomatoid urothelial carcinoma (carcinosarcoma) of the urinary bladder. *Mod Pathol*. 2009;22:113–118. PMID:18997737 DOI: 10.1038/modpathol.2008.176
- [12]. Robinson SP, Farooq A, Laniado M, Motiwala H. The demographic features, clinical outcomes, prognosis and treatment options for patients with sarcomatoid carcinoma of the urinary bladder: a single centre experience. *International Brazilian Journal of Urology : official journal of the Brazilian Society of Urology*. 2018;44(1):45-52. doi:10.1590/S1677-5538.IBJU.2016.0347. PMID: 29064650

LEGENDS:



Figure1: A polypoidal, ulcero-proliferative growth measuring 7cmx4cmx3cm seen on opening The urinary bladder.

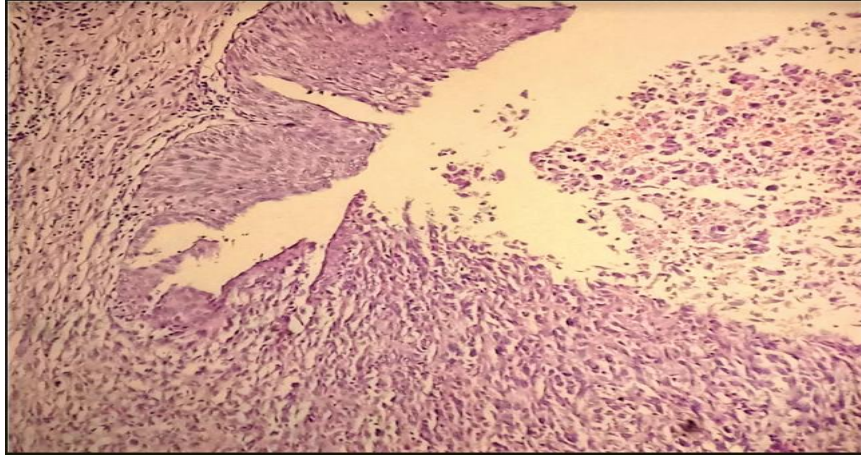


Figure 2: Urothelial carcinoma in-situ and spindle cell component (H&E;100X).

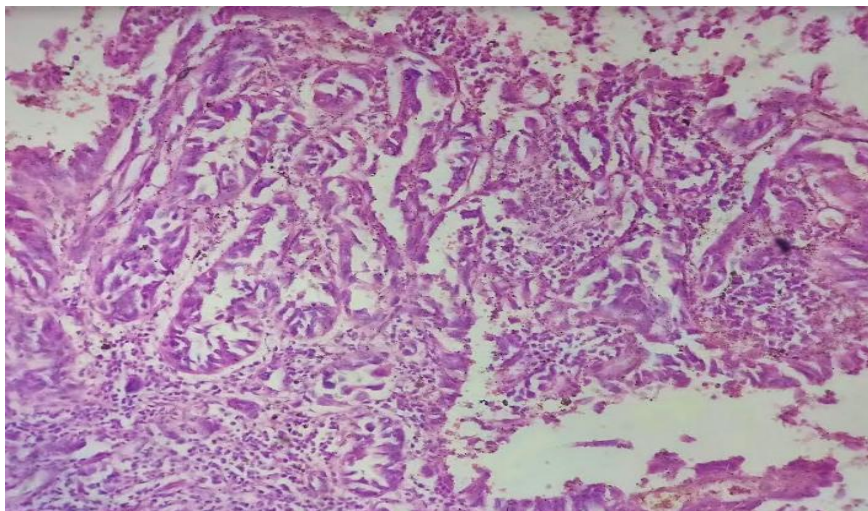


Figure 3: Epithelial component (adenocarcinoma). (H&E;200X)

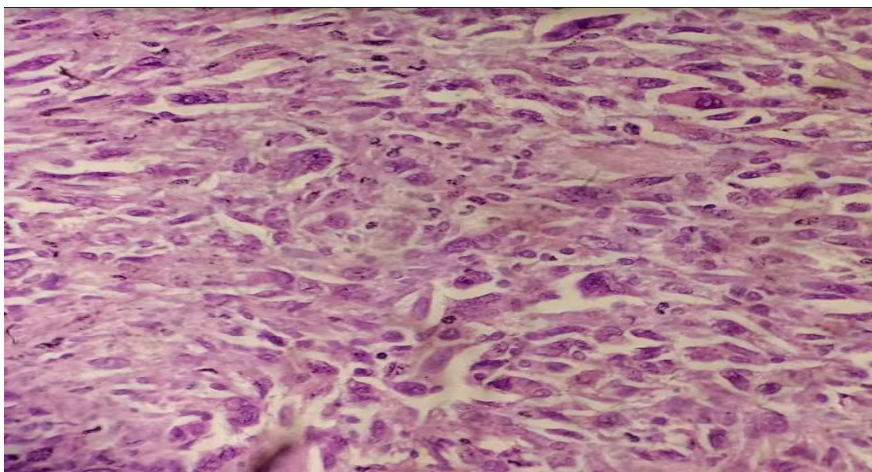


Figure 4: Sarcomatous component with spindle cells and bizarre cells (H&E;400X)

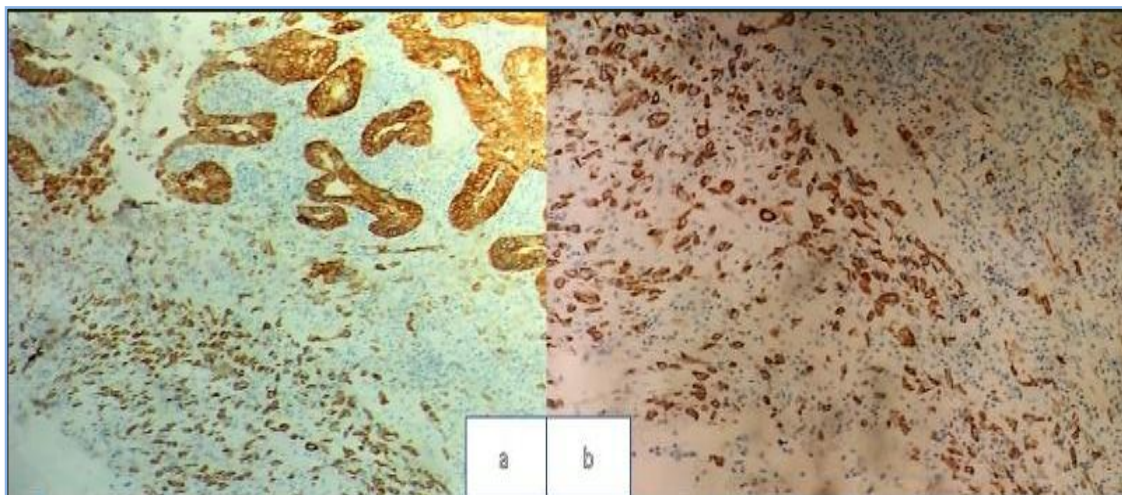


Figure 5 (a & b): Pan-cytokeratin and EMA reactive in epithelial as well as mesenchymal components (Pancytokeratin & EMA;200X).

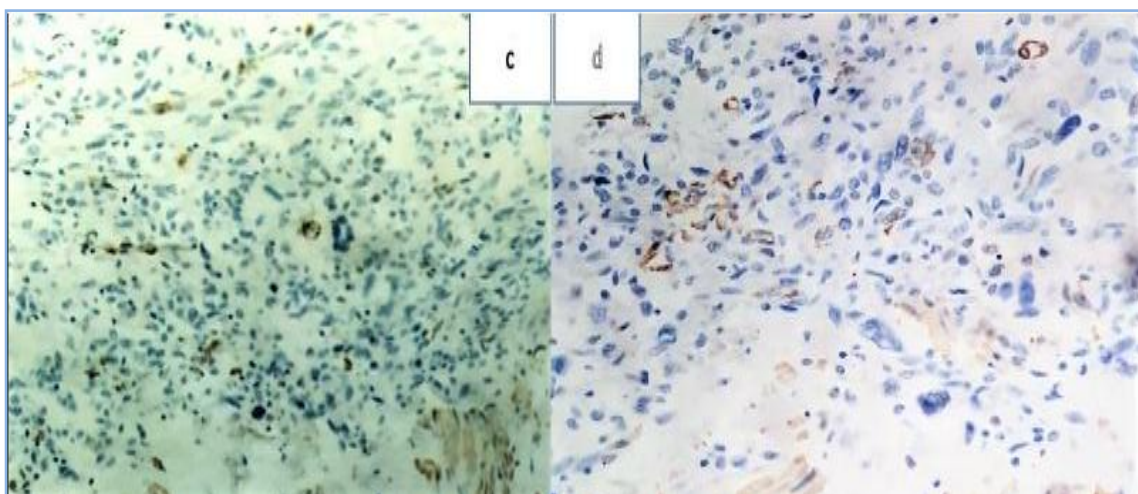


Figure 5 (c & d): Weakly positive SMA and desmin in the spindle-cell component(SMA&Desmin;200X).

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