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Sarcomatoid Urothelial Carcinoma of Urinary Bladder – A Rare Case Report

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ABSTRACT

Sarcomatoid variant of urothelial carcinoma of urinary bladder is a rare tumor which exhibits epithelial and mesenchymal differentiation of tumor cells both morphologically and immunohistochemically. Cyclophosphamide treatment and radiation therapy are associated with this tumors. This tumor is considered to be the aggressive variant of transitional cell carcinomas of the bladder. Treatment for this aggressive tumor is radical surgical excision with chemoradiation. We report a case of 47-year-female presenting with dysuria and hematuria. On imaging, heterogeneously enhancing hyperdense lesion in bladder was noted which was diagnosed as sarcomatoid urothelial carcinoma on histopathological examination.

Key words: Carcinoma, Sarcomatoid variant, Tumour, Urinary bladder

Sarcomatoid urothelial carcinoma of urinary bladder accounts for approximately 0.3% of the urothelial tumors [1]. It is biphasic malignant neoplasm with both epithelial and mesenchymal components. Though sarcomatoid urothelial carcinoma is rare, it is more common than the primary sarcoma of the bladder [2]. These tumors are associated with history of intravesical cyclophosphamide therapy and radiation therapy. Grossly, they present as polypoidal large intraluminal masses. Microscopically, epithelial component of these tumors is represented by transitional cell carcinoma, adenocarcinoma, squamous cell carcinoma or small cell carcinoma. Mesenchymal component is represented by high grade spindle cell neoplasm.

Immunohistochemically, these tumors are positive for both epithelial and mesenchymal markers. Though the definitive treatment modality has not been described due to availability of small number of cases, the preferred treatment is radical cystectomy with adjuvant chemotherapy and radiation. We, hereby, report a case of sarcomatoid urothelial carcinoma due its rarity.

CASE PRESENTATION

A 47-year-old woman presented with history of painful micturition and blood mixed urine for last 2 months. There was no other significant present or past history. On examination, she was afebrile, conscious with stable vital signs. General and systemic examination did not reveal any abnormality. All the biochemical parameters were within the normal limits.

Ultrasound examination revealed echogenic material measuring 4.6X2.8X3.2cm with few calcifications, arising from posterior right lateral wall of the base of the bladder and infiltrating the right vesicoureteric junction and moderate right hydroureteronephrosis secondary to

infiltration of right vesicoureteric junction. Contrast enhanced computed tomography (CECT) abdomen revealed heterogenously enhancing hyperdense lesion measuring 5X3.2X4.2cm along the right lateral wall of urinary bladder involving vesicoureteric junction on right side. Mild perivesical fat stranding was noted above the mass causing right hydroureteronephrosis. Cystoscopy revealed single, solitary growth of size around 4X3cms over right lateral wall extending on to right ureteric orifice.

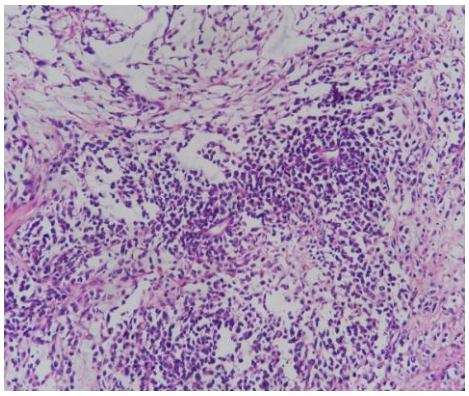

Figure 1 – Tumor showing myxoid change (upper portion) and carcinomatous cells (lower portion) (H&E, X100)

Radical cystectomy with ureterosigmoidostomy was performed and cystectomy specimen was sent for histopathological examination. Patient was healthy post operatively. Grossly cystectomy specimen was measuring 9X8X4cms. Cut section showed nodular, firm tumor in the right lateral wall measuring 4.5X3.5X2cm. Cut section of the tumor was gray white with myxoid areas. Histopathological examination of the tumor revealed tumors cells arranged in glandular pattern, alveolar pattern and nesting pattern. Many foci shows spindle shaped tumor cells having oval elongated nuclei. Mononucleated and multinucleated tumor giant cells were seen. Extensive areas of myxoid change (**Figure 1**) and areas of necrosis were noted. On the basis of these features, the diagnosis of transitional cell carcinoma with sarcomatoid differentiation was made.

DISCUSSION

Sarcomatoid variant of urothelial carcinoma of urinary bladder is an uncommon form of malignancy where tumor cells exhibit histological, cytological and molecular features of both mesenchymal tumors (sarcoma) and epithelial tumors (carcinomas). The incidence of sarcomatoid urothelial carcinoma ranges from 0.2% to 4.3% [3]. Mean age of presentation for these tumors is 66 years (range 50-77 years). These tumors are associated with history of previous treatment with intravesical cyclophosphamide therapy or radiotherapy [4].

The most common presenting features of these tumors are painful micturition, hematuria, lower abdominal pain and acute retention of urine. The most frequent location of sarcomatoid urothelial carcinoma is the lateral wall of the bladder. However, previous studies have shown the commonest location to be trigone of the bladder which gives evidence for the origin of the tumor from wolffian body [5].

Macroscopic appearance of the sarcomatoid variant of urothelial carcinoma is large polypoidal intraluminal mass. Microscopically, the tumor is composed of epithelial and mesenchymal components. Epithelial component is represented by urothelial carcinoma, squamous cell carcinoma, adenocarcinoma and small cell carcinoma. Mesenchymal component is represented by high grade spindle cell neoplasms which can be malignant fibrous histiocytoma or leiomyosarcoma. Osteosarcoma and chondrosarcoma are the most common heterologous elements found in them.

Some of these tumors exhibit prominent myxoid stroma as seen in our case. Due to this feature it can be misdiagnosed as inflammatory pseudotumor. But the inflammatory pseudotumors are positive for anaplastic lymphoma kinase stains, where as the sarcomatoid urothelial carcinoma is negative. On immunohistochemistry, epithelial component of sarcomatoid urothelial carcinoma stains positive for cytokeratins and stromal elements react with markers of mesenchymal differentiation (vimentin) [6]. Though the histogenesis of the tumor is not clear, molecular studies have shown that both epithelial and mesenchymal component of these tumors have monoclonal cell origin.

Sarcomatoid urothelial carcinoma must be differentiated from other benign and locally aggressive lesions such as pseudosarcomatous myofibroblastic proliferations or post operative spindle cell nodules and pseudotumors of the urinary bladder (inflammatory myofibroblastic tumors). Pseudosarcomatous myofibroblastic proliferations also form large polypoidal mass which has high cellularity and increased mitosis but without atypical mitotic figures. This lesion has zonal

pattern of distribution with base showing increased cellularity and towards the surface are hypocellular and myxoid regions [7]. Inflammatory myofibroblastic tumors also show myxoid areas but they lack the carcinomatous elements.

Other tumors like primary sarcomas (especially Leiomyosarcomas) and urothelial carcinoma with chondroid or osseous metaplasia should be distinguished. In the primary sarcomas, epithelial component will be absent. In the urothelial carcinoma with metaplastic components, there will be absence of atypia in the cartilage and bone [8]. These tumors are associated with poor prognosis and multimodality therapy is recommended. After surgical resection of the tumor of the urinary bladder, chemotherapy and chemoradiotherapy is recommended [9].

CONCLUSION

Sarcomatoid urothelial carcinoma of urinary bladder is rare and aggressive neoplasm that has similar clinical presentation as that of conventional high grade urothelial carcinoma, but has worse prognosis. They present at younger age and at an advanced stage. Radical surgery along with chemoradiation improves the prognosis of the tumor.

REFERENCES

1. Zhai QJ, Black J, Ayala AG, Ro JY. Histological variants of infiltrating urothelial carcinoma. Arch Pathol Lab Med. 2007; 131: 1244-56.
2. Wright JL, Black PC, Brown GA et al. Differences in survival among patients with sarcomatoid carcinoma, carcinosarcoma and urothelial carcinoma of the bladder. J Urol. 2007; 178(6): 2302-2307.
3. Black PC, Brown GA, Dinney CPN. The impact of variant histology on the outcome of bladder cancer treated with curative intent. Urologic Oncol. 2009; 27(1): 3-7.
4. Lopez – Betran A, Pacelli A, Rothenberg HJ, et al. Carcinosarcoma and sarcomatoid variant of bladder: clinicopathological study of 41 cases. J Urol. 1998; 159(5): 1497-1503.
5. Atigan D, Gencten Y. Carcinosarcoma of the bladder. A case report and Review of literature. Case reports in Urology. 2013; Article ID – 716704.
6. Sung MJ, Wang M, Mac Lennan, et al. Histogenesis of sarcomatoid Urothelial carcinoma of the urinary bladder: evidence for a common clonal origin with divergent differentiation. J Pathol. 2007; 211(4): 420-430.
7. Harik LR, Merino C, Coindre JM, Amin MB, Pedeutour F, Weiss SW. Pseudosarcomatous myofibroblastic proliferations of the bladder: a clinicopathologic study of 42 cases. Am J Surg Pathol. 2006; 30(7): 787-794.
8. Amin MB. Histological variants of Urothelial carcinoma: diagnostic, therapeutic and prognostic implications. Modern Pathology. 2009; 22(S2): S96-S118
9. Wang J, Gilespie C, Kunadharaju R, Taimon GA, Enke C. Sarcomatoid urothelial carcinoma: a single centre experience. World J Oncol. 2011; 2(4): 175-1180.

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