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## High Grade Infiltrative Urothelial Carcinoma- Sarcomatoid Variant, A Rare Entity: Case Report and Review of Literature

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# High Grade Infiltrative Urothelial Carcinoma-Sarcomatoid Variant, A Rare Entity: Case Report and Review of Literature

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**Abstract-** In the urinary bladder, majority of the neoplasms are of pure epithelial origin. In contrast pure mesenchymal tumors and biphasic epithelial-mesenchymal neoplasms, although documented, rarely occur at this site. Heterologous carcinosarcomas (also called metaplastic carcinomas or sarcomatoid carcinomas with heterologous differentiation) are defined as biphasic tumors made up of a varying mixture of carcinomatous and heterologous sarcomatous components with abrupt or gradual transition between one component and the other. These are rare tumors and account for approximately 0.3% of all bladder malignancies with a poor prognosis. Because of its unfavorable histopathologic nature and also its rarity, not much is known about the various treatment options in these tumors. The overall 5 year cancer specific survival rate after cystectomy is only 20.3%.

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## I. INTRODUCTION

Heterologous carcinosarcomas (also called metaplastic carcinomas or sarcomatoid carcinomas with heterologous differentiation) are defined as biphasic tumors made up of a varying mixture of carcinomatous and heterologous sarcomatous components with abrupt or gradual transition between one component and the other <sup>(1)</sup>. These are rare tumors and account for approximately 0.3% of all bladder malignancies with a poor prognosis <sup>(2,3)</sup>.

The histological composition of these tumors is variable but requires the presence of both epithelial and mesenchymal malignant components <sup>(4)</sup>.

Treatment of bladder carcinosarcomas should be aggressive and multimodal but optimal treatment is still unknown <sup>(5)</sup>.

Herein, we report a case of sarcomatous carcinoma with chondrosarcomatous differentiation of the urinary bladder.

## II. CASE HISTORY

A 77 year-old male was admitted to our hospital, who presented with complaints of hematuria and abdominal pain. Ultrasonography (USG) revealed a mass in the bladder involving the posterior wall and projecting into the lumen. Computed Tomography (CT) scan showed a hyper dense mass in the bladder. Transurethral resection of the bladder tumor (TURBT) was done and the biopsy submitted for histopathology.

Histologically, the tumor showed features of infiltrative high grade urothelial carcinoma with tumor cells arranged in sheets, having high nucleocytoplasmic ratio, severe anisonucleosis along with spindling of the nuclei in some of the cells.

In addition, there were also present heterologous elements in the form of groups and islands of cartilaginous tissue. Individual chondrocytes were atypical, pleomorphic and showed binucleation. Areas of necrosis and hemorrhage along with tumor giant cells were also appreciated. The deep muscle included in the biopsy was also involved by the tumor. (Fig 1, 2, 3, 4, 5)

So a diagnosis of infiltrative urothelial carcinoma– high grade, sarcomatoid variant was made.

## III. DISCUSSION

In the urinary bladder, majority of the neoplasms are of pure epithelial origin. In contrast pure mesenchymal tumors and biphasic epithelial-mesenchymal neoplasms, although documented, rarely occur at this site <sup>(4)</sup>.

Carcinosarcomas are defined by the World Health Organization as tumors composed of intimately admixed malignant epithelial and mesenchymal elements <sup>(4,6)</sup>. The epithelial components are squamous, glandular or high grade transitional carcinoma, whereas the mesenchymal components may be chondrosarcoma, malignant fibrous histiocytoma, osteosarcoma, leiomyosarcoma, fibrosarcoma or rhabdomyosarcoma <sup>(2)</sup>. In our case, histopathology showed infiltrative high grade urothelial carcinoma along with areas showing chondrosarcomatous elements.

The exact histogenesis and pathogenesis of these neoplasms remained enigmatic for many decades, and even today remains one of the

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controversial issues in tumor pathology. Previous studies have suggested that the sarcomatous component of the sarcomatoid carcinoma is derived from urothelial carcinoma<sup>(4,7)</sup>. Interestingly, Sung et al. have recently suggested the monoclonal nature of sarcomatoid bladder carcinoma with the use of molecular techniques. This finding strengthens the fact that bladder sarcomatoid carcinoma is a monoclonal lesion derived from urothelial carcinoma<sup>(8)</sup>.

Kikuchi and colleagues suggested that the frequent location of these tumors in the trigone is evidence of origin from the Wolffian body. In recent studies, commonest location of these tumors was the lateral wall<sup>(9)</sup>. In our case, the tumor was located on the posterior wall.

Most commonly, these tumors are found in the female genital tract, where they have historically been termed malignant mixed müllerian tumors<sup>(4)</sup>. These tumors commonly occur in older individuals, display an aggressive biologic behavior with poor prognosis and are at advanced stage disease at initial diagnosis<sup>(2,4)</sup>. The carcinosarcoma of the bladder is more commonly seen in males, and male/female ratio is nearly 2 : 1. The disease appears in the seventh decade of life. The most common symptoms are macroscopic hematuria and dysuria. Generally more than 70% of cases present with advanced stage and have a worse prognosis than conventional urothelial carcinomas<sup>(9)</sup>. The patient in this case was a 77 year old male who presented with complaints of hematuria and abdominal pain.

The mesenchymal element of carcinosarcoma lacks epithelial markers and patients with carcinosarcoma present at a more advanced stage and are at greater risk for death compared to patients with high-grade urothelial carcinoma<sup>(9)</sup>.

Because of its unfavorable histopathologic nature and also its rarity, not much is known about the various treatment options in these tumors. The overall 5 year cancer specific survival rate after cystectomy is only 20.3%<sup>(2)</sup>.

Even though transurethral resection, radical cystectomy, neoadjuvant chemoradiotherapy were the proposed treatment protocols of these tumors, radical cystectomy with adjuvant chemotherapy was the most appropriate treatment option due to the patient's poor prognosis. Nevertheless mean survival rates of these patients are very poor and metastatic disease occurs in 66% of patients within 1 year<sup>(2,4)</sup>.

#### IV. CONCLUSION

Sarcomatoid variant of urothelial carcinoma tends to be high grade, exhibit aggressive biologic behavior, and has a poor prognosis as compared to usual urothelial carcinoma. More studies are needed regarding the exact histogenesis of this tumor to ascertain the radical therapy needed for this rare neoplasm

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Figure Legends

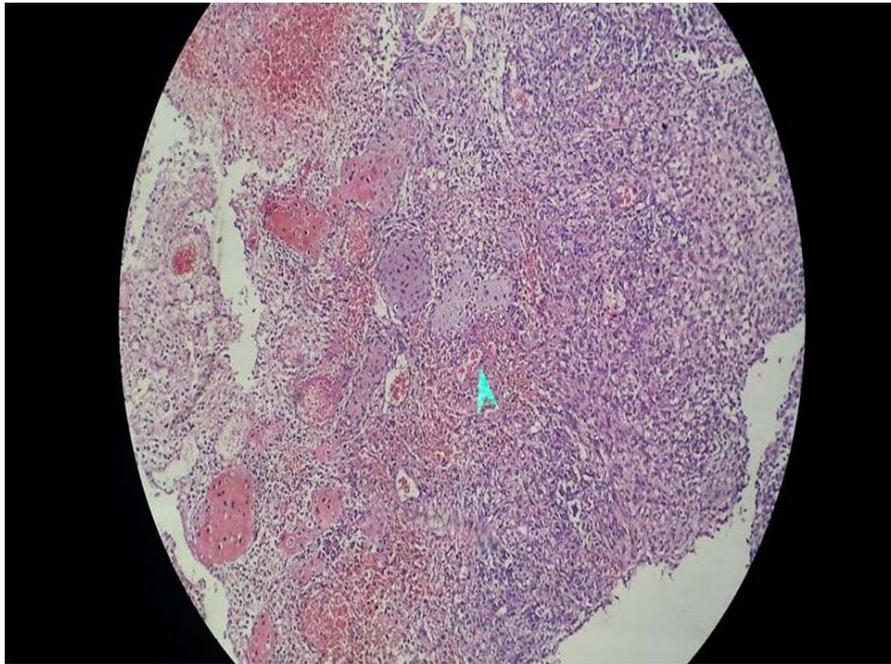


Figure 1 : Scanner view showing Chondrosarcomatous, carcinomatous foci along with hemorrhage (H&E 40X)

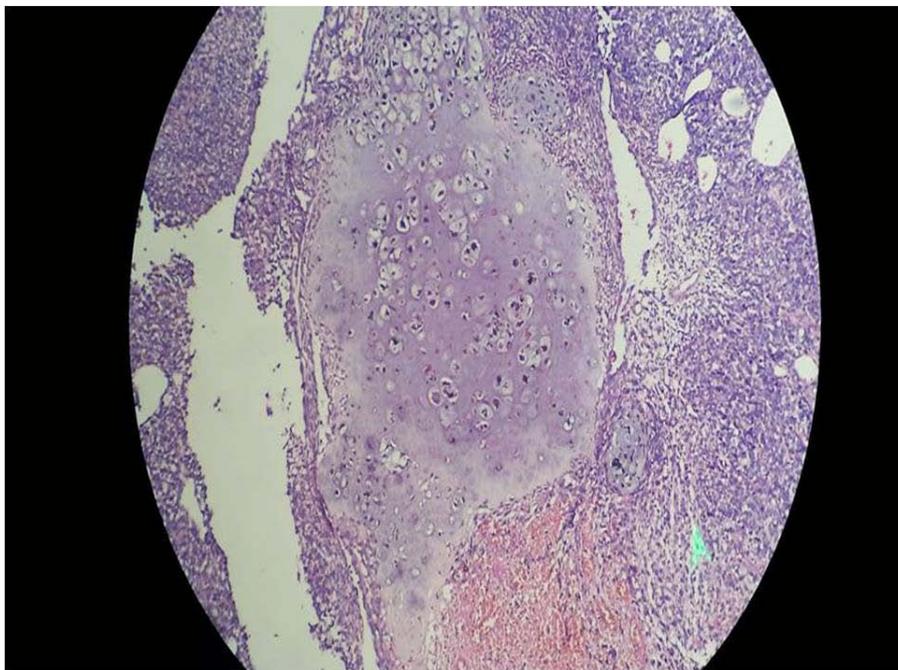
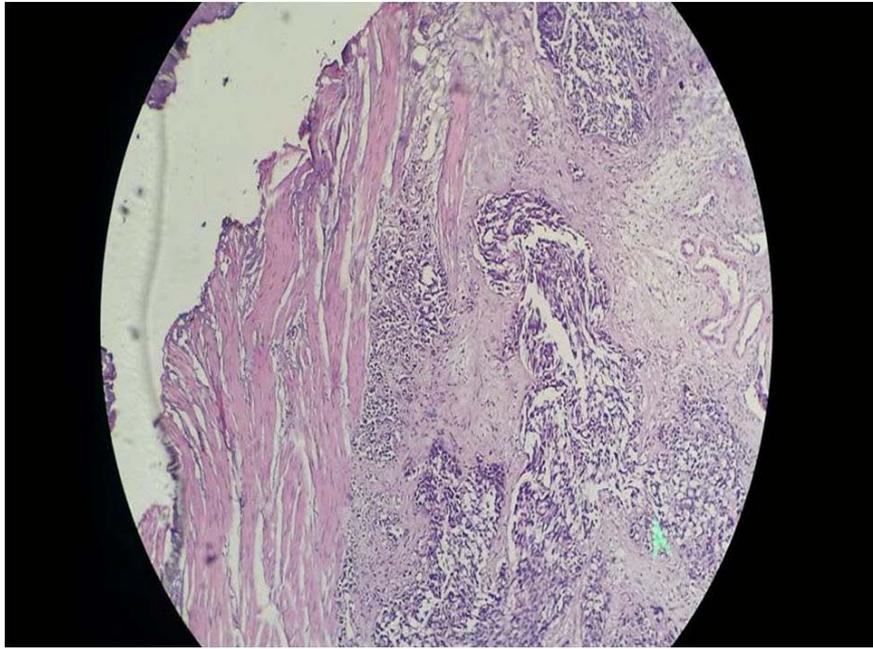
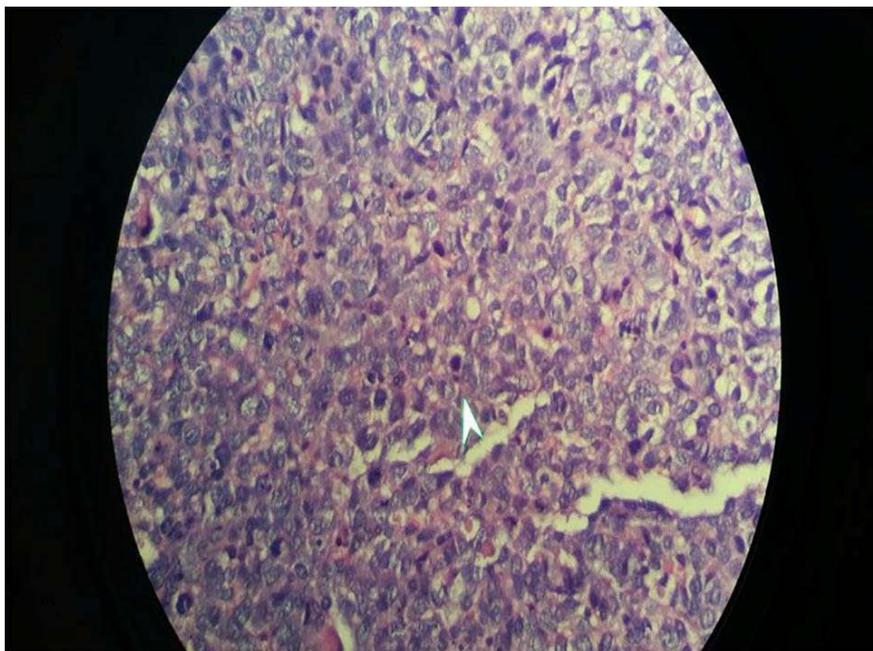


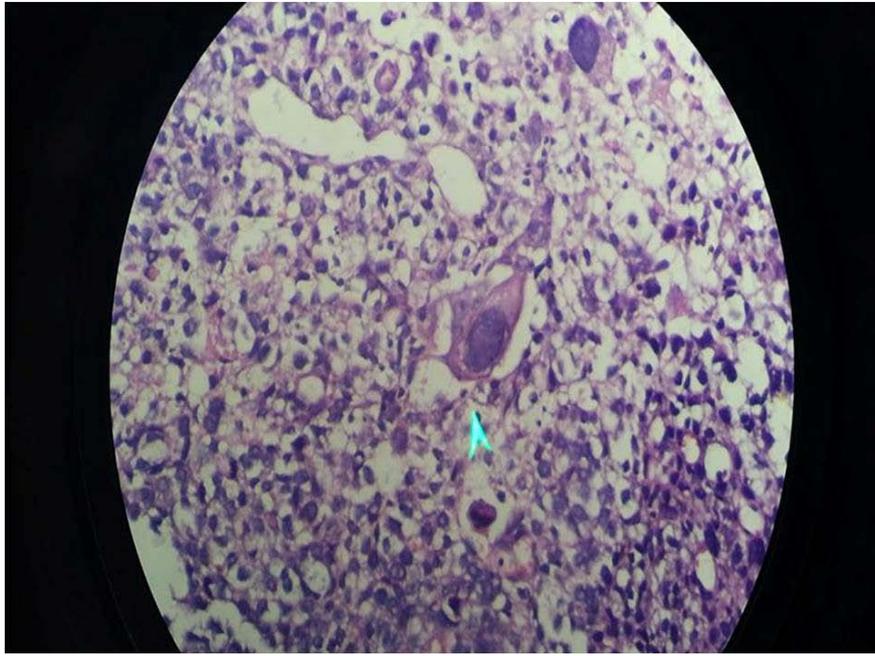
Figure 2 : Low power view showing mixture of chondrosarcomatous and carcinomatous foci (H&E 100X)



*Figure 3 :* Low power view showing Muscularis Propria invasion (H&E 100X)



*Figure 4 :* High power view showing the carcinomatous component (H&E 400X)



*Figure 5 :* High power view showing large bizzare tumor cells (H&E 400X)