



BMH Med. J. 2017;4(4):126-130 **Case Report**

## **A Rare Tumour With A Rare Presentation: A Case Of Sarcomatoid Variant Of Urothelial Carcinoma Arising From Ureter**

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### **Abstract**

Sarcomatoid variant of urothelial carcinoma is a very rare tumor, and this tumour arising in a ureteric location is even rarer still. We report a case of such a tumor with this rare presentation.

**Keywords:** urothelial carcinoma, sarcomatoid variant, ureter

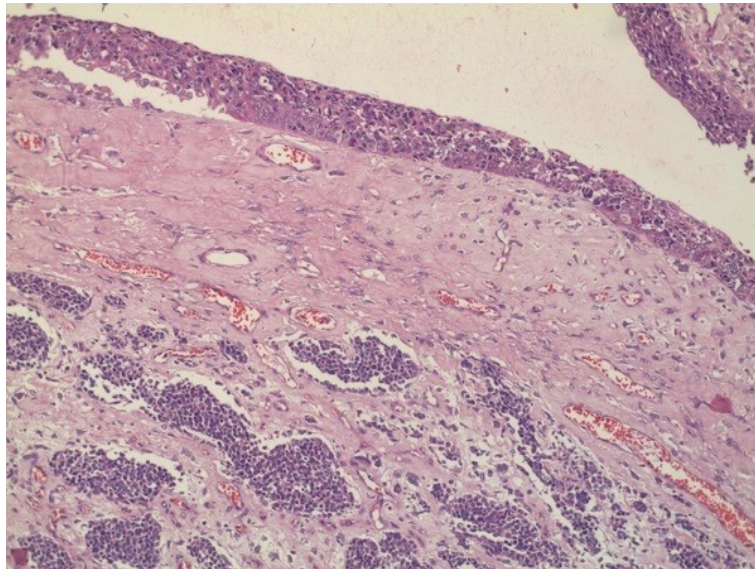
### **Case presentation**

A 51 year old, post-menopausal lady, with no co-morbidities, had presented with lower abdominal pain, loss of appetite and loss of weight. She had also noticed progressive abdominal distension. There was no history of vomiting, constipation, malena, bleeding per vaginum or urinary symptoms.

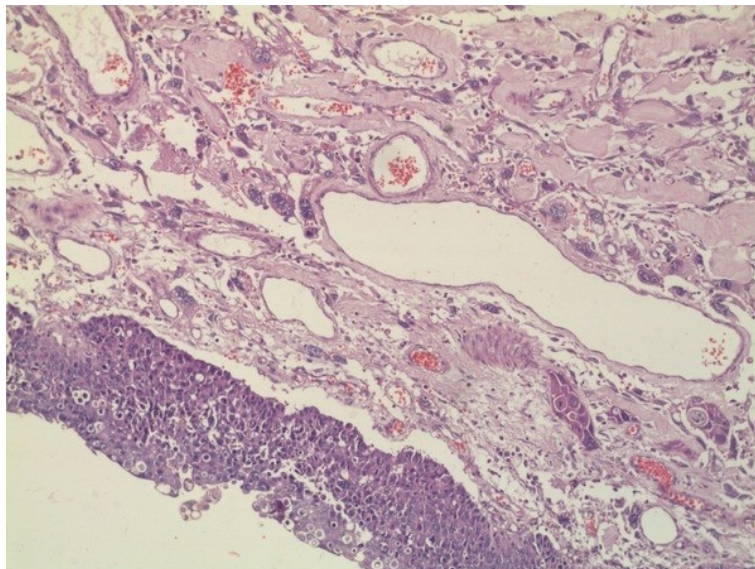
MRI of the abdomen and pelvis, showed a large, heterogeneous, lobulated, enhancing, predominantly solid and partially-cystic, mass lesion in the pelvis (mostly extending to the right half); with multiple focal lesions along the wall of the dilated right ureter and pelvicalyceal system. Differential diagnoses considered radiologically were (1) Multicentric Transitional Cell Carcinoma (TCC) (2) Ovarian carcinoma with invasion of the collecting system, causing grade IV hydronephrosis. USG guided pelvic biopsy was done which was reported as undifferentiated sarcoma.

She then underwent surgery [Anterior Resection, small bowel resection, Total abdominal hysterectomy (TAH) with bilateral salpingo-oophorectomy (BSO) and right nephro-ureterectomy.

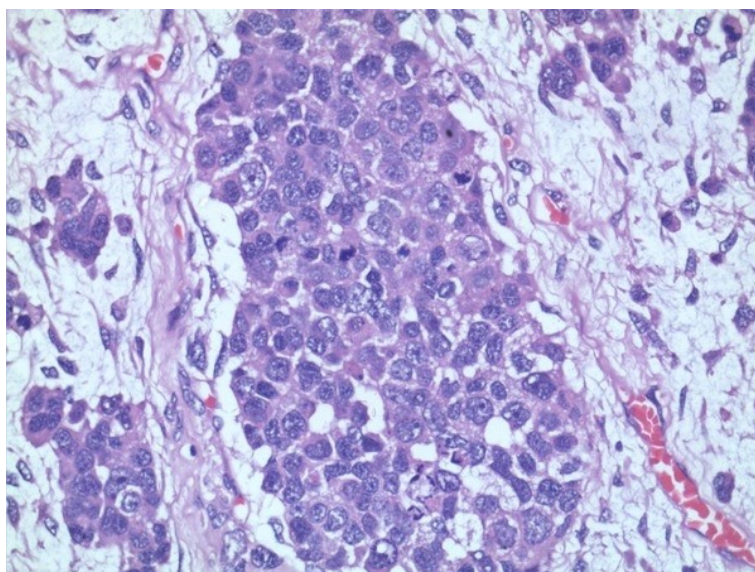
Histopathological examination of surgical specimen was reported as sarcomatoid urothelial carcinoma of lower ureter. The lower ureter also showed carcinoma in situ changes. The largest diameter of the tumor was 19 cm, and it was extending into the renal pelvis. The kidney showed lymphatic tumor emboli. Tumor was adherent to the colon and ovary. Uterus and fallopian tubes were free of disease. Cells were diffusely positive for vimentin and focally for cytokeratin, CD 8/18, HMWCK and CD117 and they are negative for CK 7, CK20, ChromograninA, SMA, DOG-1, S100p, desmin and uroplakin III.



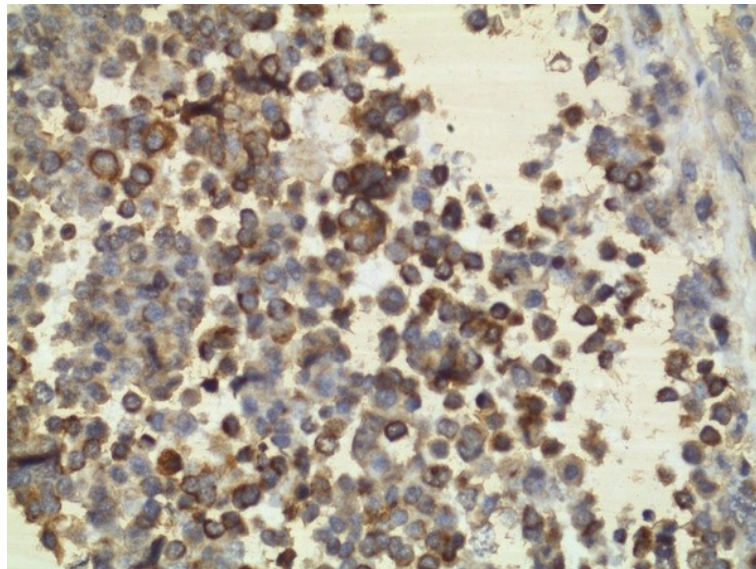
**Figure 1:** Section showing dysplastic neoplasm and submucosa showing lobules of cells having small hyperchromatic nuclei.



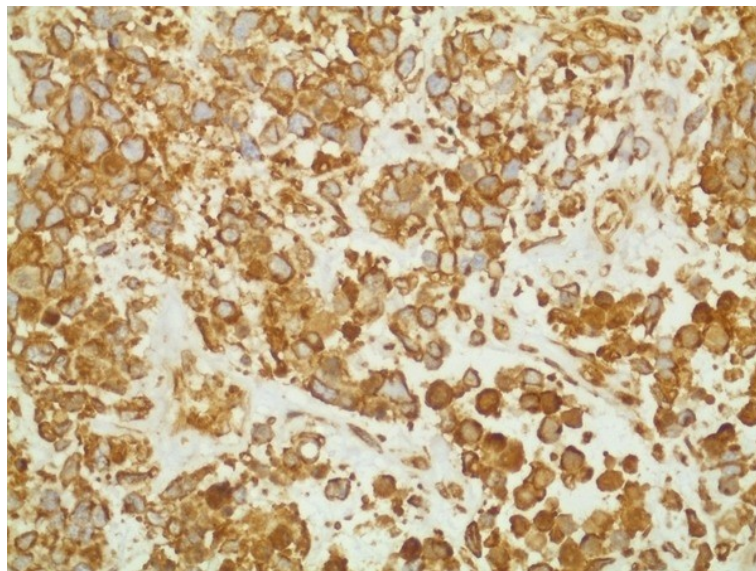
**Figure 2:** Pleomorphic nuclei in a loose stroma showing foci of spindly and bizarre cells with highly atypical and bizarre nuclei.



**Figure 3:** Nests of pleomorphic cells with plenty of mitosis.



**Figure 4:** Shows focal positivity of cytokeratin.



**Figure 5:** Fiffuse positivity for vimentin.

Post-surgery, the patient received adjuvant external beam radiation therapy (5040cGy in 28 fractions, 180cGy/fraction, 1 fraction daily), using IMRT technique, to the post-operative surgical bed.

After completing radiation, she was started on adjuvant chemotherapy with gemcitabine and carboplatin. While on chemotherapy, she was detected to have a lump in the left breast, and an excision biopsy of the same was done. Histopathological examination was suggestive of a metastatic lesion from the urothelial carcinoma, sarcomatoid variant. Subsequently, second line chemotherapy, with liposomal doxorubicin, was initiated. During the course of second line chemotherapy, patient had disease progression, with deterioration of general condition, and expired, 11 months after diagnosis.

### **Discussion**

Ureteric malignancies are rare, with primary tumors of ureter being only half as frequent as tumors of the renal pelvis. Transitional cell carcinoma (TCC), the most common histology of upper urinary tract malignancies, occur two to three times more frequently in men, peaking in the 7th and 8th decades of life. [1] .Sarcomatoid urothelial carcinoma usually presents between the ages of 50 years

and 77 years and the mean age of presentation is 66 years [2].

Women are more likely to have a more advanced and higher grade tumor, than men, at nephro-ureterectomy. [3] Even among transitional cell carcinomas (TCC), the most common urological histology, tumours arising from the ureter, account for only 1% of all urinary tract malignancies, while TCC of the bladder is a hundred times more frequent. [1] The incidence of sarcomatoid urothelial carcinoma ranges from 0.2% to 4.3%, in reported literature. [3]

In an analysis of the Surveillance, Epidemiology, and End Results (SEER) database, Wright et al. reported that a total of 135 sarcomatoid urothelial carcinomas and 166 cases of carcinosarcoma were identified from among 1,82,283 patients with primary bladder cancer. [4] Sarcomatoid carcinoma is an uncommon form of carcinoma whose malignant cells have histological, cytological, or molecular properties of both epithelial tumours (carcinoma) and mesenchymal tumours. [5] Immunohistochemical staining had revealed that epithelial elements react with cytokeratins, and stromal elements react with specific markers which correspond with the type of mesenchymal differentiation showing positive immunohistochemical staining with cytokeratin (MNF16) and Vimentin. [6,7] Sarcomatoid variant of urothelial carcinoma is the terminology that is used for all biphasic malignant neoplasms, which exhibit a morphologic and/or immunohistochemical picture of epithelial and mesenchymal differentiation, with the presence or absence of heterologous elements. [5] The terminology carcinosarcoma has been seen to be used as a synonym for sarcomatoid carcinoma but they are recognized as different pathological entities [8]. Though histological distinction of sarcomatoid carcinomas from carcinosarcomas is often difficult, immunohistochemistry is a helpful diagnostic adjunct in the correct diagnosis. [8-11] This rare biphasic variant of urothelial carcinoma had, only been described in small case series, which had suggested a poor outcome for patients with this variant of urothelial carcinoma [1]. Patients with sarcomatoid variant of urothelial carcinoma have worse disease-specific and overall survival, even after adjusting for stage of tumour, in comparison with patients with high-grade pure urothelial cancer [3,4] There is no definite treatment protocol for this sarcomatoid variant of urothelial carcinoma. [3,4,12] Information gained from a number of publications reveal that an estimated 70% of patients with sarcomatoid variant of urothelial carcinoma die within 2 years of diagnosis. [9,13-16]

## Conclusion

Sarcomatoid variant of urothelial carcinoma is a rare, aggressive type of tumour which tends to present at a younger age and at a higher grade and stage, and is associated with a poor outcome in comparison with conventional urothelial carcinoma. The rarity of this histology, and specifically, its ureteric location, makes this an unusual case, that we would like to report. Data from few, short, case series suggest that chemo-radiation, in addition to radical surgery, may improve outcomes in this tumour. Pooled, multi-institutional data needs to be harvested, to better evaluate the various treatment options, for this rare malignancy, and study outcomes, to help in prognostication and optimization of therapy.

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