

Sarcomatoid Carcinoma of the Renal Pelvis with Prominent Chondrosarcomatous and Osteosarcomatous Elements: A Case Report

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Sarcomatoid carcinoma (SC) is a high-grade malignant neoplasm that exhibits morphological and immunoprofile of epithelial and mesenchymal differentiation. This tumour can arise in various organs including breast, larynx, oral cavity, oesophagus, genital tract and bladder [1]. SCs of renal pelvis are very rare and even more uncommon are the tumours showing foci of heterologous differentiation. To the best of our knowledge, around 25 cases of such neoplasms have been reported in the literature to date.

The present case is a histologically proven SC involving the renal pelvis and upper ureter with malignant chondrogenic and osteogenic elements. Only one previous case report has been published with chondrosarcomatous areas [2] and two cases with osteosarcomatous foci [3,4]. Our case is the first example comprised of heterologous differentiation showing both chondrosarcomatous and osteosarcomatous elements.

Our patient was an 80 years old gentleman with known type II diabetes mellitus and hypertension, whose presenting complaint was haematuria. He was detected to have a left renal pelvic mass on ultrasound scan. The following CT abdomen and pelvis demonstrated the left pelvicalyceal system to be dilated to the level of the proximal ureter, with internal enhancing component suggestive of a renal pelvic neoplasm (Figure 1A). The rest of the kidney showed cortical thinning with renal cysts.

A laparoscopic nephroureterectomy was performed and showed marked hydronephrosis with a large friable grey-white to haemorrhagic mass filling the pelvicalyceal system (Figure 1B). The vessels at the hilum were not involved. The remaining renal parenchyma showed cortical atrophy with no discernible cortico-medullary differentiation. A cortical cyst was also noted. Two lymph nodes were sampled from the renal hilum. Adrenal gland was not identified.

Microscopic examination of the renal pelvic mass showed a malignant neoplasm centred within the pelvicalyceal system with mixed carcinomatous and sarcomatous elements, with the latter component predominating. The epithelial components (40%) included urothelial carcinoma in situ and high grade conventional urothelial carcinoma

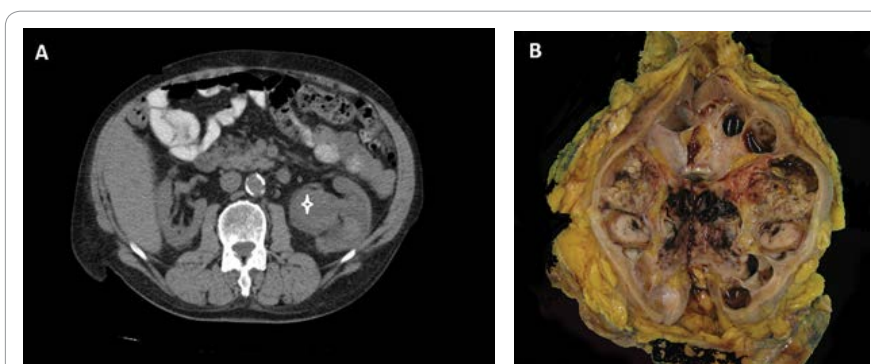


Figure 1. (A) CT scan showing large heterogeneously enhancing mass lesion in the pelvicalyceal system of the left kidney. **(B)** Gross specimen of radical nephroureterectomy with variegated haemorrhagic solid mass in renal pelvis.

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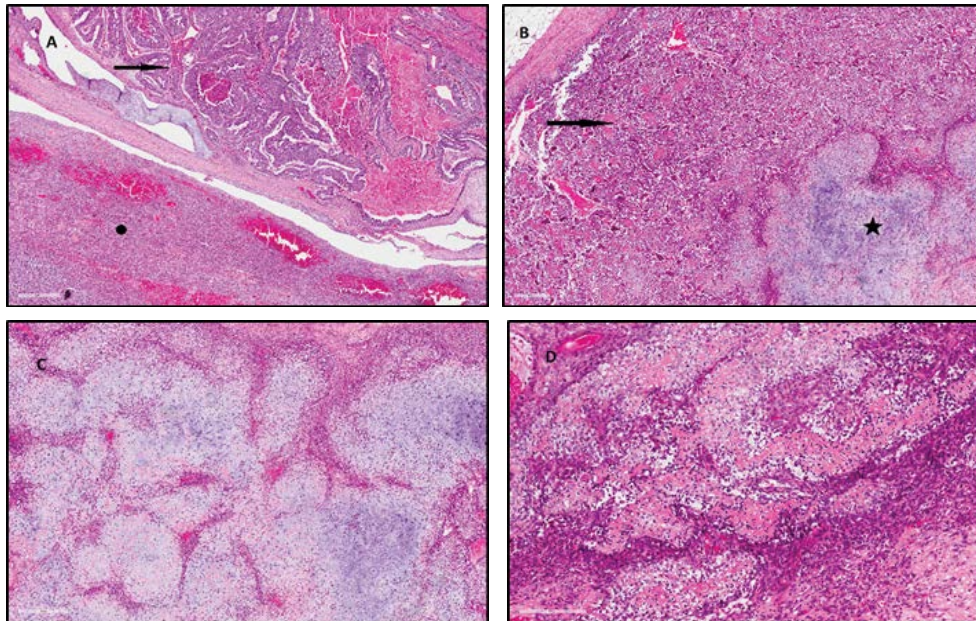


Figure 2: (A) Sarcomatoid carcinoma with carcinomatous component (arrow) and sarcomatous spindle cell elements (circle). (B) Admixed chondrosarcomatous (star) and osteosarcomatous (arrow) components. (C) Chondrosarcomatous and (D) Osteosarcomatous components.

(Figure 2A). The sarcomatous elements encompassed plump spindle cells admixed with osteoclastic giant cells (Figure 2A). Heterologous differentiation with admixed chondrosarcomatous and osteosarcomatous elements were identified (Figure 2B-2D). Necrosis and haemorrhage accounted for 30% of the overall tumour. Small vessel invasion was also present. The proximal ureter was invaded by the tumour with infiltration into the inner layers of muscularis propria. The distal ureter was dilated but not involved by the tumour.

The tumour had infiltrated into the renal parenchyma, renal sinus fat with focal perinephric fat involvement. The uninvolved parenchyma showed changes of end stage renal disease secondary to diabetes and obstructive uropathy. A benign cortical cyst and two uninvolved lymph nodes were also identified.

The postoperative period was uneventful. The patient had routine follow up with the renal physician for haemodialysis. Six months later, he was detected to have metastatic disease involving abdominopelvic lymph nodes, vertebra, clavicle and iliac bone. He underwent radiotherapy for metastatic disease. Due to lack of treatment progress, radiotherapy was ceased. His general condition deteriorated, and he subsequently passed away.

In the WHO classification of tumours [5], the term sarcomatoid variant of urothelial carcinoma is defined by the presence of histological features that are morphologically indistinguishable from those of sarcoma. The first case of SC of the renal pelvis was reported by Pisciole, *et al.* in 1984 [6].

Clinically, these are large and bulky tumours. There is a male gender predilection with mean age of patients ranging from 50-77 years [7,8]. Tobacco smoking and irradiation of the genital tract in gynaecological patients have been considered risk factors for the development of SC [9]. The presenting symptoms are non-specific, including gross haematuria, flank pain, abdominal mass

like conventional urothelial tumours. On radiological examination, CT images are informative in depicting the origin and nature of the tumour, showing the components of changing densities.

Little is known about the pathogenesis of SCs. Two opposing theories have been proposed. The monoclonal theory states that the carcinomatous and sarcomatous tumour cells are both derived from a single pluripotent stem cell [10]. The multiclonal theory, on the other hand, states that the SC is a collision tumour composed of the derivatives of two or more stem cells of separate epithelial and mesenchymal origin [11].

SC has a worse prognosis than the more common urinary tract carcinomas, and recurrence and metastasis are frequent, as seen in our case. Limited data suggest that tumours with heterologous elements have much worse prognosis than tumours without them [5]. Hence, extensive sampling is required to identify the biphasic elements and carcinoma in situ.

This case was exceptional in the fact that it showed divergent chondrosarcomatous and osteosarcomatous components. SC of the renal pelvis with both these components has not been described previously in the literature. Most of the SCs described were mixed transitional carcinoma with spindle cell carcinoma. The histological differential diagnosis includes various types of sarcomas, carcinomas with metaplastic benign-appearing bone or cartilage in the stroma and reactive pseudosarcomatous mesenchymal proliferations. True sarcomas do not show epithelial elements or carcinoma in situ. The pseudosarcomatous proliferations mostly would have a prior history of urological or gynaecological surgery and would show a diploid pattern in DNA ploidy analysis, in contrast to aneuploid pattern in SC [12]. Our case was unequivocal for SC, clearly showing malignant epithelial and sarcomatous differentiation, the latter showing the rare combination of osteosarcomatous and chondrosarcomatous heterologous elements.

Overall, SCs behave badly as compared to the usual type of carcinoma of the urinary tract. Metastatic disease and advance tumour involvement of the renal parenchyma was present in all the previously reported cases of the renal pelvic SCs. In summary, we present a case of sarcomatoid carcinoma involving the renal pelvis and ureter, showing osteosarcomatous and chondrosarcomatous differentiation, a combination not previously described in the literature.

Conflicts of interest and sources of funding

The authors state that there are no conflicts of interest to disclose.

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