Iranian Journal of Pathology | ISSN: 2345-3656

Sarcomatoid Carcinoma of Renal Pelvis Involving Ureter and Renal Parenchyma with Heterologous Osteosarcomatous Differentiation: A Case Report and Review of Literature

Bhavya P. Mohan[1]*, Jayalakshmy PL[1], Letha V[1], Suresh Bhat[2]

1. *Dept. of Pathology, Government Medical College, Kottayam, Kerala, India*
2. *Dept. of Genitourinary Surgery, Government Medical College, Kottayam, Kerala, India*

KEYWORDS

Sarcomatoid Carcinoma
Renal Pelvis
Osteosarcoma

Article Info

Received 31 Aug 2016;
Accepted 10 Oct 2017;
Published Online 2017;

ABSTRACT

Sarcomatoid carcinoma is a high-grade rare malignant tumor with both epithelial and mesenchymal components. Sarcomatoid carcinoma in the upper urinary tract is very rare. We reported here a case of sarcomatoid carcinoma of renal pelvis with osteosarcomatous differentiation, with involvement of the ureter and renal parenchyma in a 68-year-old female. Histologically, predominant pleomorphic spindle cell sarcoma component with osteoid production and urothelial carcinoma component with in situ areas were identified. Immunohistochemical analysis showed vimentin positivity in sarcomatous component and cytokeratin positivity in carcinomatous component.

Corresponding Information:Dr.Bhavya P.Mohan, Assistant Professor, Pathology, Medical college, Kottayam, E-mail: bhavyasharath80@gmail.com

Introduction

Sarcomatoid carcinoma is a rare high-grade malignant neoplasm with morphological and/or immunohistochemical evidence of both epithelial and mesenchymal differentiation. Sarcomatoid carcinoma can occur in many organs including the bladder, breast, female genital tract, oesophagus, kidney and larynx (1, 2). The urinary bladder is the predominant site of its occurrence in the urinary tract (3). Sarcomatoid carcinoma of renal pelvis (SCRP) is extremely rare (4). Surgical resection is the treatment of choice at present. Because of the rare nature of SCRP and its poor response to adjuvant therapy, early pathologic diagnosis is crucial to improve patients' survival. Thorough histopathological evaluation together with immunohistochemistry is essential for accurate diagnosis of this biphasic tumor. To the best of our knowledge, only less than 30 cases of sarcomatoid carcinoma of the renal pelvis have been published in English literature and only two of them showed osteosarcomatous differentiation (3). Here, we reported a case of sarcomatoid carcinoma arising in renal pelvis, involving ureter and renal parenchyma, with osteosarcomatous differentiation at all the three sites.

Case report

A 68-year-old female presented with a history of painless haematuria associated with passage of clots for one month. She was catheterised following an episode of acute urinary retention 3 weeks back. She had a history of diabetes and hypertension. There was no abdominal mass on palpation. The haemoglobin level was 11.2g/dL. Other haematological and biochemical parameters were within the normal limits. Urine cytology had negative results regarding malignant cells. Spiral computed tomography scan of abdomen and pelvis with urogram revealed a large heterogeneously enhancing infiltrative lesion with specks of calcification in the pelvicalyceal system of the left kidney infiltrating into the left renal cortex and upper half of the ureter with hydronephrosis and hydroureter (Figure.1).

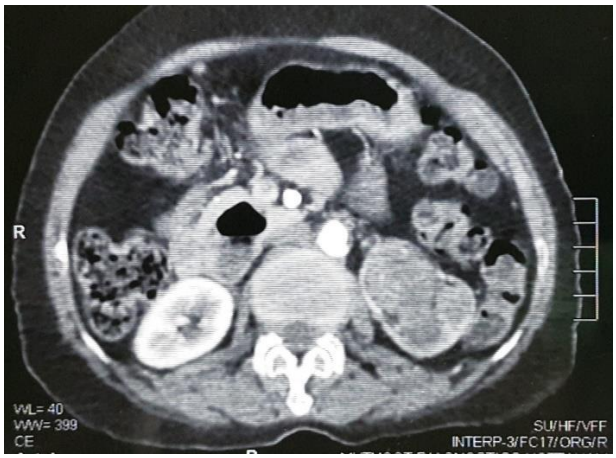

Fig 1. CT scan showing large heterogeneously enhancing mass lesion in the pelvicalyceal system of the left kidney infiltrating into left renal cortex.

Left nephroureterectomy was performed with a preoperative diagnosis of transitional cell carcinoma.

Gross pathology; resected nephrectomy specimen measuring 12 cm × 5 cm × 4 cm with attached ureter measuring 10 cm. External surface of the kidney was covered with perinephric pad of fat and appeared smooth and nonadherent. Cut surface of the kidney showed a polypoidal whitish solid homogeneous growth in the pelvic region measuring 10 cm × 4.5 cm × 3 cm extending to the renal parenchyma. The corticomedullary differentiation was retained in the adjacent renal parenchyma. The tumor also involved the ureter with marked dilatation, irregular nodularities on the surface of upper ureter, wall thickening and intraluminal extension (Figure 2a).

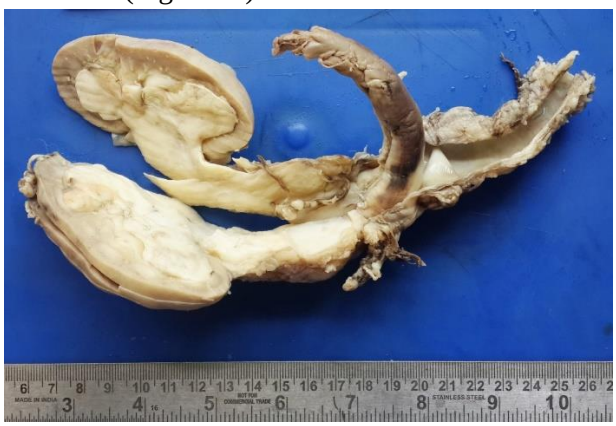

Fig 2a. Gross specimen with whitish solid mass in renal pelvis, ureter and kidney

The resected end of the ureter, renal capsule and vessels were free of tumor.

Microscopy showed a neoplasm composed predominantly of sheets and poorly formed fascicles of pleomorphic spindle cells with vesicular nuclei and nucleoli.

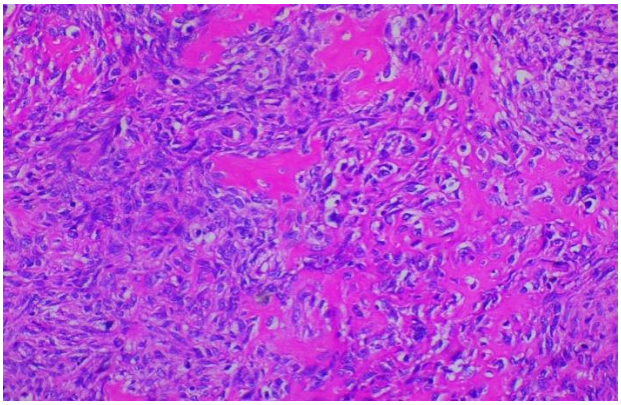

Fig 2b. Section showing sarcomatous area with abundant osteoid production (H&E stain X 200)

Extensive foci of osteosarcomatous differentiation was noted with abundant osteoid matrix production and areas of mineralised osteoid (Figure 2b).

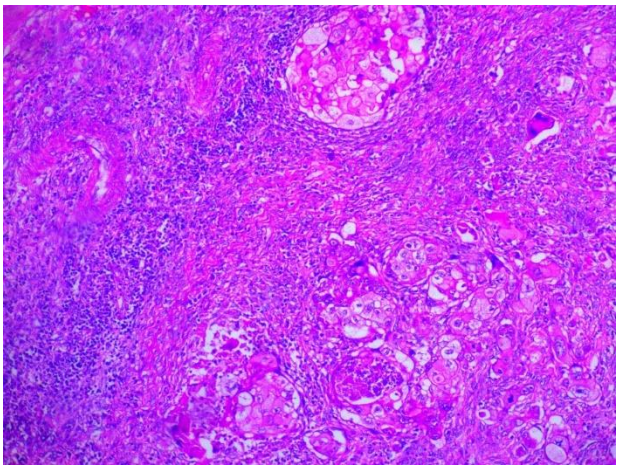

Fig 2c. Section showing areas of urothelial carcinoma forming epithelial component (H&E stain X 100).

6-8 mitotic figures/high power field with atypical mitosis, tumour giant cells, foci of necrosis, calcification and hyalinisation were also noted. The tumour involved renal parenchyma and ureter with osteosarcomatous differentiation at both sites. The focal epithelial component was formed by islands of urothelial carcinoma with carcinoma in situ in the urothelium of renal pelvis (Figure 2c, 2d).

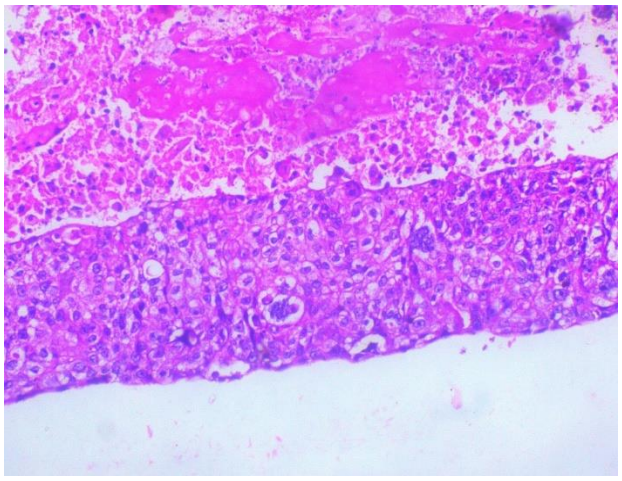

Fig 2d. Section showing focus of urothelial carcinoma in situ with marked pleomorphism (H&E stain X 200)

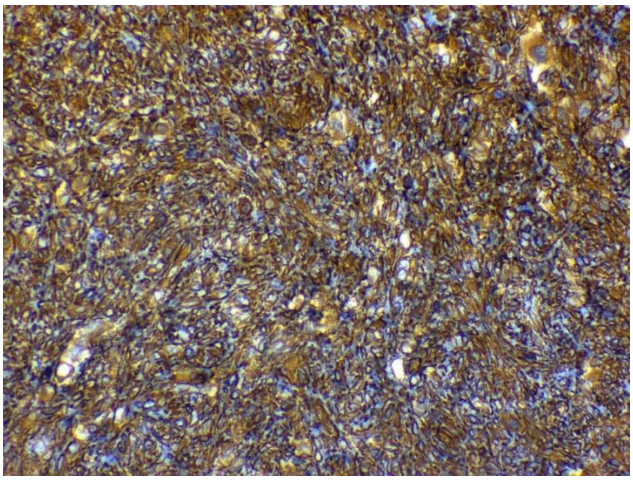

Fig 3b. Immunostain (x 200 magnification) showing vimentin positive sarcomatous areas

Immunohistochemistry was performed and epithelial elements were found to have strongly positive results for cytokeratin and negative for vimentin. The sarcomatous component was found to have positive results for vimentin (Figure 3 a, b).

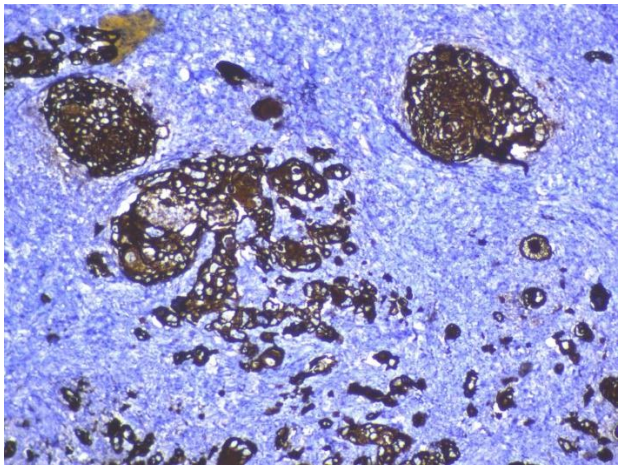

Fig 3a. Immunostain (x 200 magnification) showing strong cytokeratin positivity in epithelial component and negativity in spindle cell areas.

A pathological diagnosis of sarcomatoid carcinoma of renal pelvis with osteosarcomatous differentiation with involvement of the ureter and renal parenchyma was given. The resected end of the ureter, renal capsule and vessels were free of tumor. The postoperative period was uneventful and the patient was discharged 8 days after the operation. Ten months later the patient was receiving regular clinical follow-up and there was no evidence of local or metastatic disease.

Discussion

As per the World Health Organization Classification of Tumours (2016), “sarcomatoid carcinoma” (also called carcinosarcoma) is a biphasic malignant neoplasm exhibiting epithelial and mesenchymal differentiation (1). Immunohistochemically, epithelial elements react with cytokeratins and sarcomatous elements react with vimentin or other mesenchymal specific markers.

SCRP is a very rare entity. The first true case of sarcomatoid carcinoma in the renal pelvis was reported by Piscioli et al. (2). To the best of our knowledge, only less than 30 cases of sarcomatoid carcinoma of the renal pelvis have been reported in English literature (Table 1).

The age range of onset for this tumour is 38-85 years with a male-to-female ratio of 4-5:1. Presenting symptoms include gross haematuria, flank pain, abdominal mass, dysuria and acute urinary retention. Our patient was a 68-year-old female and presented with gross haematuria and acute urinary retention.

Table 1. Characteristics of previous reported cases of SCRP

Authors	Year	Salient features	No. of cases
Piscioli *et al*.	1984	62/M; TCC/rhabdomyosarcomatous tumor-like	1
Wick *et al*.	1985	45/M; TCC/Anaplastic spindle cell sarcoma in 1 case; SCC/spindle in other case	2
Tajima Y et al.	1988	66/M; Renal pelvic tumor containing transitional cell carcinoma, adenocarcinoma and sarcomatoid elements	1
Suster S et al.	1989	85/M; Spindle cell carcinoma of the renal pelvis demonstrating coexpression of keratin and vimentin intermediate filaments	1
Lopez Beltran et al.	1996	Mean age 71.6years; M:F=3:2; mean survival after onset of symptoms 11.2months; Spindle/anaplastic in 3cases; spindle/myxoid in 2 cases	5
Vermeulen P et al.	2000	Biphasic sarcomatoid carcinoma of the right kidney with chondrogenic differentiation	1
Hisataki *et al.*	2001	43/F; TCC/spindle; in a duplicated renal pelvis	1
M. F. Acikalin et al.	2005	66/M; SCRP with Giant Cell Tumour like Features	1
Thiel DD et al.	2006	65/M; Renal pelvis confined sarcomatoid carcinoma of transitional cell origin.	1
Yu-Li Lin, et al.	2008	67/F; TCC in situ and another sarcoma-like area with prominent spindle cells	1
Chen GM et al.	2011	77/M; SCRP in duplex kidney	1
S. Gill Samra et al.	2012	76/F; Sarcomatoid carcinoma involving the renal pelvis and ureter with heterologous osteosarcomatous differentiation	1
Chen S et al.	2013	Age range 38-78 years; All males; Tumor size range 1.5-16cms. A spindle cell carcinoma without apparent epithelial elements in 1 case, spindle and TCC in 4 cases, spindle and SCC in 3cases.	8 (over a 10 year period)
Hye In Ahn et al.	2013	68/M; Sarcoma with exuberant osteosarcomatous element and papillary urothelial carcinoma	1
Xiquan Tian et al.	2014	49/F; Pleomorphic sarcoma with multinucleate giant cells and high grade urothelial carcinoma	1
Ramakrishnan D et al.	2014	49/M; Urothelial carcinoma arising in the renal pelvis with exuberant chondrosarcomatous element associated with adrenal metastasis	1

IHC: Immunohistochemistry; TCC: Transitional cell carcinoma; SCC: Squamous cell carcinoma; M: Male; F: Female; SCRP: Sarcomatoid carcinoma of renal pelvis

In most of the cases, the epithelial component was urothelial carcinoma with squamous or glandular differentiation and the mesenchymal element was poorly or undifferentiated spindle cells with or without heterologous elements (5). The peculiar histologies reported in association with sarcoma component were rhabdomyosarcomatous, giant cell tumour like, myxoid, osteoclast rich and chondrosarcomatous types (3, 4, 6, 7). Our case showed a predominant component of pleomorphic sarcoma with osteosarcomatous differentiation with abundant osteoid matrix production which is extremely rare at this site. Moreover, both ureter and renal parenchyma were involved by osteosarcomatous component. The focal epithelial component was formed by islands of urothelial carcinoma with in situ carcinoma of urothelium of renal pelvis. Immunohistochemical analysis showed vimentin positivity in sarcomatous component and cytokeratin positivity in carcinomatous component. Only two cases of sarcomatoid carcinoma of renal pelvis with osteosarcomatous differentiation have been published so far (3). It is almost impossible to reach a correct preoperative diagnosis of SCRP, due to the lack of distinctive clinical or radiological features. The accurate diagnosis is often made by histological features and immunohistochemical findings, postoperatively.

The exact pathogenesis of sarcomatoid carcinoma has been a matter of controversy (9). The monoclonal theory about its pathogenesis states that both carcinomatous and sarcomatous tumour cells

are derived from a single pluripotent stem cell that undergoes divergent epithelial and mesenchymal differentiation. Sung et al. (9) supported monoclonal cell origin of sarcomatoid carcinoma and suggested that clonal divergence may occur during tumour progression and differentiation by demonstrating identical pattern of non-random X-chromosome inactivation and significant overlap of loss of heterozygosity in both carcinomatous and sarcomatous components.

The differential diagnoses include true sarcomas, sarcomatoid renal cell carcinomas and non-neoplastic lesions such as inflammatory pseudo tumour. Sometimes, differentiating the origin of sarcomatoid carcinoma as either a primary renal or urothelial tumor is difficult. Demonstration of carcinoma in situ change in the urothelium and immunohistochemical staining of PAX8 and the transcription factor GATA3 can be of use in identifying the origin (10).

Sarcomatoid carcinoma is a high grade tumor associated with recurrences and metastasis. A poor prognosis has been reported for most patients with very few exceptions (5, 8, 11). Our patient did not receive any adjuvant therapy. She was on regular follow-up 10 months later and there was no evidence of local recurrence or metastasis.

In conclusion, we presented a case which is extremely rare. Though rare, SCRP should be considered in the differential diagnosis of tumors of renal pelvis. Thorough sampling of renal pelvic tumours to demonstrate biphasic elements and carcinoma in situ component and using appropriate immunohistochemistry panel would help in diagnosis.

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