Case Report

Primary small cell carcinoma of kidney in a young female: A rare case report

Ajay Kumar Pal, Sudheer Rathi, Krishna Murty, Nishant Misra

Department of Surgery, LLRM Medical College, Meerut, Uttar Pradesh, India

ABSTRACT

Small cell carcinomas (SCCs) occur mainly in pulmonary tract while extrapulmonary SCCs (EPSCCs) are rare neoplasms comprising approximately 2.5% of all SCCs. A 23-year-old female presented with complaints of hematuria and a mass in right flank region. She had no other complaints. Examination of the patient revealed lump in the right lumbar region with no other remarkable findings. Contrast-enhanced computed tomography showed a large heterogeneously enhancing necrotic mass involving the right entire kidney. A right radical nephrectomy was performed, and a histological examination revealed mass involving whole right kidney with capsular and lymphatic invasion along with small cell histology. The patient was given adjuvant chemotherapy of etoposide and carboplatin, every 3 weeks. The patient started complaining of bony pain after 1 month of operation, and bone scanning revealed of multiple osteoblastic lesions. Zoledronic acid (4 mg) had been started for bony metastasis. SCC of the kidney is a rare neoplasm of the elderly age group that has aggressive behavior with an increased tendency for locoregional and distant dissemination. Our patient represents the first case of SCC of the kidney to be reported in a young female.

Key words: Chemotherapy, female, metastasis, nephrectomy, small cell carcinoma

INTRODUCTION

Small cell carcinomas (SCCs) occur mainly in pulmonary tract while lung extrapulmonary SCCs (EPSCCs) are rare neoplasms comprising approximately 2.5% of all SCCs.[1] Among EPSCCs, urinary bladder prostate and are most notable sites in genitourinary tract.[1] Kidney as a Primary site of SCC is exceedingly rare with approximately fifty cases published till date[2,3] and most of these cases patient were above 60 years of age (median age = 62 years).[4] Since it is infrequent, the natural course of renal SCC is not recognized very well. The clinical course of this disease is highly aggressive and frequently associated with recurrence. Surgical resection may not suffice, and adjuvant chemotherapy forms an important pillar of therapy. We, therefore, present a case report of this rare malignancy in a young female along with management challenges faced by us.

Address for correspondence: Dr. Ajay Kumar Pal, Department of Surgery, King George Medical University, Lucknow - 226 003, Uttar Pradesh, India.
E-mail: akpal.jnmc@yahoo.com

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.200109

CASE REPORT

We report a case of 23-year-old female who presented with complaints of hematuria and a mass in the right flank region. She had no other complaints and gave a normal past, menstrual, family, and medical history. Examination of the patient revealed lump in right lumbar region with no other remarkable findings.

Contrast-enhanced computed tomography [Figures 1 and 2] abdomen was performed and it showed a large heterogeneously enhancing necrotic mass of 14 cm × 9 cm involving the right entire kidney with minimal sparing of the right upper pole with distortion of the

Cite this article as: Pal AK, Rathi S, Murty K, Misra N. Primary small cell carcinoma of kidney in a young female: A rare case report. Clin Cancer Investig J 2016;5:540-3.

pelvicalyceal system and thinning of renal parenchyma and extension into renal pelvis with intra-tumoral thrombus in right renal vein extending up to its confluence with inferior vena cava with no contrast excretion. Other radiological investigation (chest X-ray, pelvis, and spine) were normal.

A right radical nephrectomy was performed, and a histological examination revealed 15 cm × 10 cm × 10 cm mass involving whole right kidney with capsular and lymphatic invasion along with small cell histology with no lymphatic involvement. Neuroendocrine differentiation was proven by a positive immunoreaction to synaptophysin and neuron-specific enolase [Figures 3-5].

DISCUSSION

SCC is a highly malignant and aggressive tumor with usual site in lung. EPSCC is a relatively rare and was first reported in 1930 by Duguid and Kennedy.[5] SCCs are uncommon in the urinary tract and mostly they are associated with other components such as transitional cell carcinoma, adenocarcinoma, squamous cell carcinoma, and sarcomatoid squamous cell carcinoma. Most commonly urinary bladder is the site of urinary tract SCC, with the primary renal pelvis, renal parenchyma, and ureteral SCC occurrence is distinctively rare. In 1998, Mackey *et al*.[4] reported 180 cases of urinary tract SCC out of which 106 cases were located in the urinary bladder, sixty in the prostate, eight in the kidney and six in the ureter. In a study by Majhail *et al*.[6] (2003) of 22 patients of SCC of kidney and renal pelvis, 62 years was the median age with a female preponderance (male:female ratio, 1:3.4). Among symptoms, abdominal pain (70%) was most commonly reported. Thirty-two percent had distant metastases at the time of diagnosis. Therapy was constituted by surgery and/or systemic chemotherapy (nephrectomy alone: Nine patients; nephrectomy and chemotherapy: Ten patients; chemotherapy alone: Three patients) with median survival being 8 months (range, - 1–101 months). Improved overall survival was present with the use of

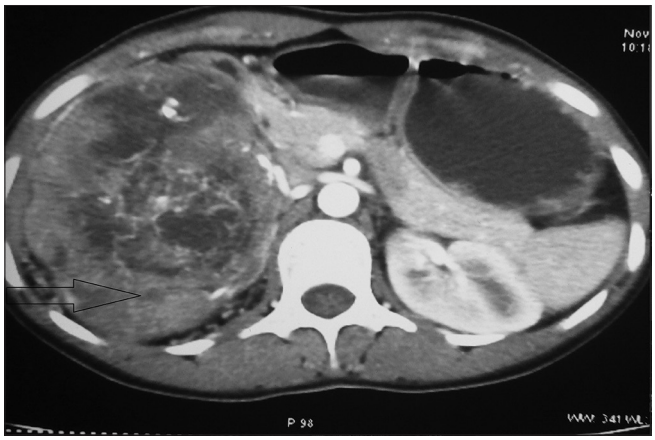

Figure 1: Arrow showing large heterogeneously enhancing necrotic mass of involving the right entire kidney

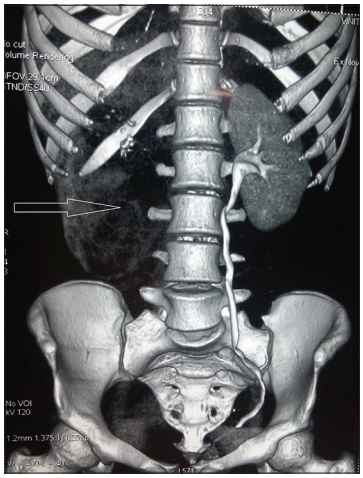

Figure 2: Three-dimensional reconstruction image. Arrow showing 14 cm × 9 cm mass involving the right entire kidney with minimal sparing of the right upper pole with distortion of the pelvicalyceal system and thinning of renal parenchyma and extension into renal pelvis

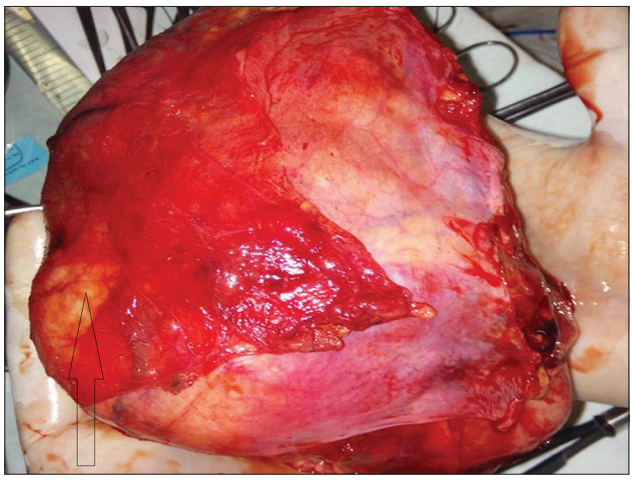

Figure 3: Gross specimen of enlarged right kidney

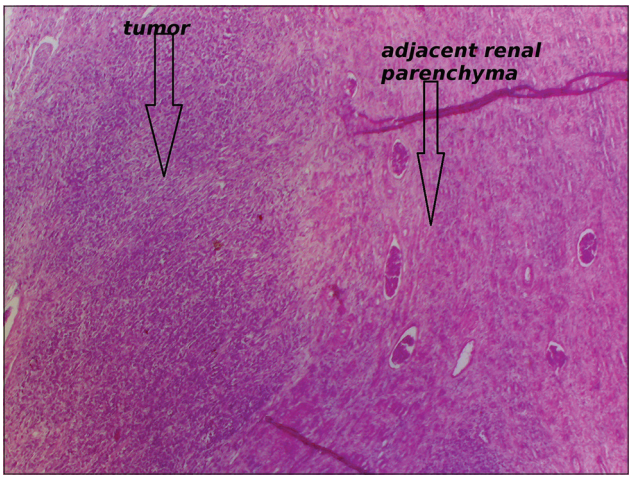

Figure 4: Histopathological examination showing tumor and adjacent normal renal parenchyma

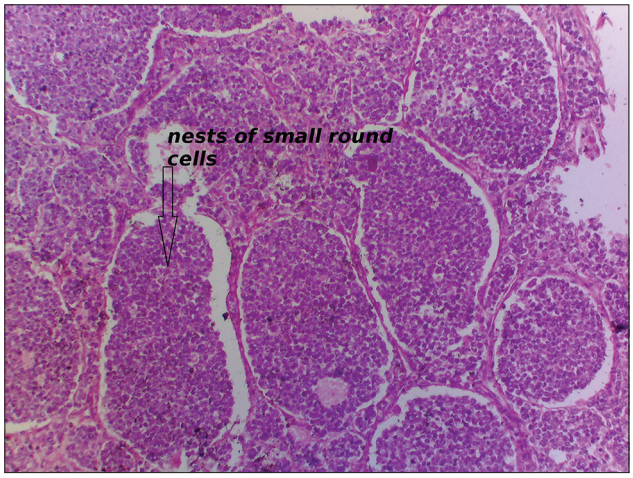

Figure 5: Histopathological examination showing small round cells distributed in a nested pattern

platinum-based chemotherapy (median survival was 20 months in patients receiving a platinum-containing regimen compared with 8 months in those who did not receive platinum; $P = 0.02$).

Regarding histopathogenesis of urinary tract SCC, two theories have been put forward. One suggests that it originates from intrinsic neuroendocrine cells within the normal genitourinary tract, derived from the neural crest[7] during embryogenesis, whereas the other indicate that it transforms from pluripotent epithelial reserve[8] cells in the genitourinary tract with the ability to generate any cell type. Transitional cell carcinoma or other histology exists as frequent combinations with SCC and thus lending support to the latter theory. Urinary tract SCC shares the same histologic features of lung SCC, which are round to fusiform shaped small cells, inconspicuous or absent nucleoli, scanty cytoplasm, and high mitotic activity. Tumor cells have positivity for neuroendocrine markers such as chromogranin, synaptophysin, and neuron-specific enolase.[8] The clinical features of primary urinary tract SCC are indistinguishable from those of renal clear cell carcinoma, with hematuria and flank pain being the most frequently reported symptoms. Gross hematuria is due to vascular invasion, while pain is secondary to hydronephrosis following obstruction of the ureter. These symptoms are hallmark of an advanced stage.

The staging of urinary tract SCC is in accordance with SCC of the lung, that is, whether or not the primary tumor can be encompassed within a tolerable radiation therapy port. Thus, a tumor confined to the primary site, with or without regional lymph node involvement, is classified as limited disease, whereas spread of the disease beyond locoregional boundaries is considered an extensive disease.

In our case, it was limited disease at the time of surgery.

What constitutes optimal treatment of urinary tract SCC is not well-established. Multimodality therapy being advocated by many includes surgery, chemotherapy, and radiation.[4,6] The presently recommended combination of a platinum-based chemotherapeutic agent and etoposide is the most commonly used regimen because of its better response rate compared with other regimens.[4]

The patient was given adjuvant chemotherapy. Four cycles of adjuvant combination chemotherapy were scheduled, including etoposide 80 mg/m^2 on days 1, 2, and 3 with carboplatin 80 mg/m^2 on day 1, every 3 weeks.

The patient started complaining of bony pain after 1 month of operation and bone scanning revealed of multiple osteoblastic lesions in the region of left parietal region in skull, right head of humerous, sternum, multiple cervical vertebra, left side multiple ribs and left head of femur and acetabulum.

CONCLUSION

SCC of the kidney is a rare neoplasm of the elderly age group that bears the features of its equivalent arising from the tracheobronchial and other extrapulmonary sites in its aggressive behavior with an increased tendency for locoregional and distant dissemination. Clinical presentation is usually delayed with high likelihood of occult/disseminated metastasis. Our patient represents the first case of SCC of the kidney to be reported in a young female. Limited improvement in survival offered by platinum-based chemotherapy in these cancers should be explored with further research.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Remick SC, Ruckdeschel JC. Extrapulmonary and pulmonary small-cell carcinoma: Tumor biology, therapy, and outcome. Med Pediatr Oncol 1992;20:89-99.
2. Trivedi PP, Kriplani D, Shah A, Sen S, Baxi H, Shah MJ. Primary small cell carcinoma of the renal pelvis: A case report with review of literature. World J Pathol 2013;4:100-6.
3. Patil S, Kaza RC, Kakkar AK, Chamberlain RS. Small cell carcinoma of the renal pelvis: A case report and review of the literature. ISRN Urol 2011;2011:786505.
4. Mackey JR, Au HJ, Hugh J, Venner P. Genitourinary small cell carcinoma: Determination of clinical and therapeutic factors associated with survival. J Urol 1998;159:1624-9.

5. Duguid JB, Kennedy AM. Oat-cell tumours of mediastinal glands. J Pathol Bacteriol 1930;33:93-9.
6. Majhail NS, Elson P, Bukowski RM. Therapy and outcome of small cell carcinoma of the kidney: Report of two cases and a systematic review of the literature. Cancer 2003;97:1436-41.
7. Fetissof F, Dubois MP, Lanson Y, Jobard P. Endocrine cells in renal pelvis and ureter, an immunohistochemical analysis. J Urol 1986;135:420-1.
8. Christopher ME, Seftel AD, Sorenson K, Resnick MI. Small cell carcinoma of the genitourinary tract: An immunohistochemical, electron microscopic and clinicopathological study. J Urol 1991;146:382-8.