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Case report

Primitive neuroectodermal tumor of kidney mimicking as an inflammatory renal mass



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KEYWORDS

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Abstract

Introduction: Renal primitive neuroectodermal tumor (PNET) is a rare and aggressive renal tumor with few reported cases in the literature.

Observations: We report a case of a 23-year-old male patient who initially presented with features of an inflammatory renal space occupying lesion (SOL) on clinical evaluation and imaging. Guided fine-needle aspiration cytology from renal mass revealed poorly differentiated neoplasm. Left open radical nephrectomy was performed. Final histopathology examination, despite the absence of clinical, radiological and gross features was consistent with a diagnosis of renal PNET. Such uncommon presentation of renal PNET has been rarely reported in the literature. Our patient then received six cycles of adjuvant chemotherapy (vincristine 1.5 mg/m² on day 1, doxorubicin 20 mg/m² on days 1–3, etoposide 150 mg/m² on days 1–3, and ifosfamide 3 g/m² on days 1–3 with mesna every 21 days). The patient developed multiorgan metastasis and progressive disease after remaining disease-free for 14 months.

Conclusion: Renal PNET should be kept in the differentials of a renal SOL presenting in adolescents and young adults. All diagnostic modalities concerning SOL of the kidney must be interpreted with caution in order for the appropriate management. Punctures for cytology can be indicated in select cases. Histopathology, immuno histochemistry supported by cytogenetic studies are required for the exact diagnosis of renal PNET. Multidisciplinary approach consisting of surgery, chemotherapy, and radiotherapy is recommended to manage this condition in view of its aggressive nature and poor prognosis.

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Introduction

Renal primitive neuroectodermal tumor (PNET) is a rare renal tumor with few reported cases in the literature [1–3]. Since this entity has no characteristic features on clinical examination and imaging it may present with confusing features. A 23-year-old man presented with left flank pain and fever for one month. Imaging revealed a large mildly enhancing mixed density space occupying lesion (SOL) in left kidney with adjacent fat stranding suggesting an inflammatory mass. Guided fine-needle aspiration cytology from renal mass was done and revealed poorly differentiated neoplasm. Left open radical nephrectomy was performed and the final histopathology examination, despite the absence of clinical, radiological and gross features were consistent with a diagnosis of renal PNET.

Case presentation

A 23-year-old male patient presented with discomfort in the left loin with a history of fever for last one month. On examination, he looked pale. A lump was palpable in the left upper quadrant and lumbar region measuring around 10 cm × 8 cm × 6 cm. His pulse rate was 125/min, BP 120/85 mmHg and temp 39.2 °C. Except for mild anemia and leucocytosis, his routine blood tests were normal (Hb 9.2 g/dL, TLC $13.6 \times 10^9/l$ and Platelets $156 \times 10^9/l$; blood urea 4.9 mmol/l and creatinine 70 $\mu\text{m/l}$). Chest X-ray did not reveal any abnormality. An ultrasound (US) examination of the abdomen showed a voluminous cystic SOL in the mid pole of the left kidney. To better evaluate the US findings, a contrast-enhanced computed tomography (CECT) scan of the abdomen was performed that revealed a large (14 cm × 12 cm × 8 cm) roundish mixed density solid cystic lesion with interrupted peripheral rim calcifications predominantly involving upper and mid-pole of the left kidney with minimal enhancement and adjacent perinephric fat plain stranding (Fig. 1). In view of atypical presentation and radiologic features image-guided FNAB was performed which revealed poorly differentiated neoplasm. The patient was planned for surgery and he underwent left radical nephrectomy with open access that showed a friable yellow lesion, partially cystic (Fig. 2); it was difficult to

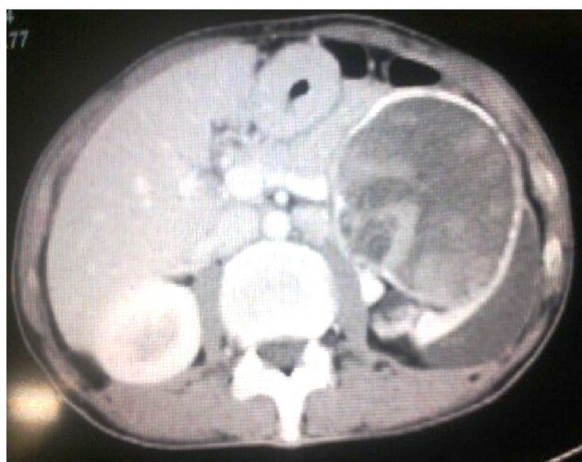


Figure 1 Computerized tomography scan image of abdomen depicting mildly enhancing large roundish mixed density solid cystic lesion in left kidney with interrupted peripheral rim calcifications. The lesion also shows internal non-enhancing necrotic areas and adjacent perinephric fat stranding.



Figure 2 Image showing surgically excised left kidney depicting large encapsulated lesion with solid cystic areas.

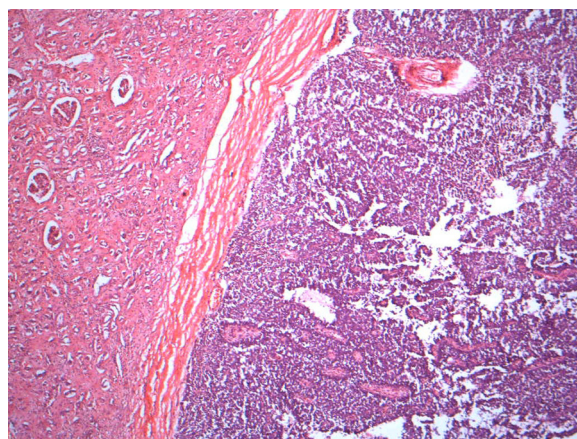


Figure 3 Scanner view showing an encapsulated tumor tissue showing small round cells.

dissociate the SOL from adjacent organs because of many adhesions. However, the final histological report came as a surprise to us and revealed PNET limited to the left kidney. Microscopy revealed the presence of small round cells (Fig. 3). Immuno-histochemistry revealed strong immunostaining for CD99 (Fig. 4), S100, and neuron-specific enolase. Vimentin staining was focally positive and CK20 and WT-1 were negative. Our patient then received six cycles of adjuvant chemotherapy (VIDE; vincristine, ifosfamide, doxorubicin, and etoposide) consisting of vincristine 1.5 mg/m² on day 1, doxorubicin 20 mg/m² on days 1–3, etoposide 150 mg/m² on days 1–3, and ifosfamide 3 g/m² on days 1–3 with mesna every 21 days, all intravenously (I.V.) with granulocyte-colony stimulating factor. The patient developed multiorgan metastasis and progressive disease after remaining disease-free for 14 months.

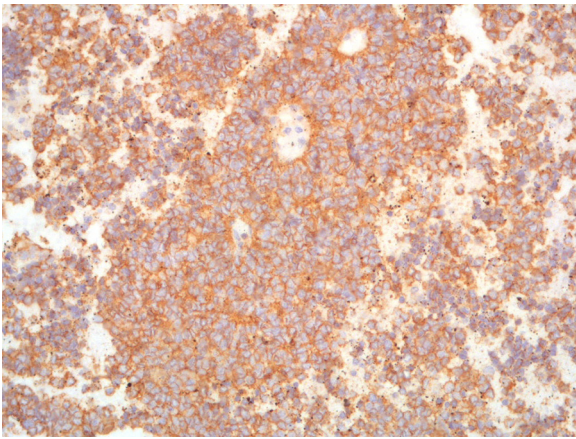


Figure 4 Tumor cells showing CD99 positivity.

Discussion

Renal PNET is a rare renal tumor first described by Seemayer et al. in 1975 [1]. Around 100 case reports have been so far published with few cases reported from India [2,3]. The largest case series of 34 cases were described by Karpate et al. in 2012 [3]. It mainly occurs in adolescents and young adults, with an average presentation age of 27 years and has a slight male predominance [4,5]. Renal PNET usually presents with nonspecific symptoms, including flank pain, hematuria and symptoms related to genitourinary infections [2–5]. CT scan images have no characteristic features to differentiate it from other tumors, most commonly there are large, heterogenous masses with variable areas of hemorrhage or necrosis manifesting as areas of enhancement with diffuse calcifications [6]. Atypical solid kidney masses such as in present case present a challenge for definitive radiologic characterization. In the evaluation of renal masses, renal fine needle aspiration biopsy (FNAB) has become an exceptional procedure in this era of renal helical computed tomography (CT), which has a high diagnostic accuracy in the characterization of renal cortical lesions. There are mainly four distinct indications for renal FNAB: (a) solid renal masses with atypical radiological features or poorly characterized on imaging studies, (b) confirmation of radiological suspected renal malignancy in inoperable patients (advanced stage disease or poor surgical candidate status), (c) kidney mass in a patient with a prior history of other malignancy, and (d) miscellaneous (drainage of abscess, indeterminate cystic lesion, urothelial carcinoma [7]. An image-guided renal mass biopsy is safe, reliable and accurate, and it changes clinical management in many cases by avoiding nephrectomy or other surgical options [8]. Microscopically renal PNET is characterized by primitive appearing round cells with the high nucleo-cytoplasmic ratio, perivascular pseudo-rosettes, typical Homer-Wright rosettes [4]. Immunohistochemically, renal PNET shows positivity for CD 99 and FLI-1 with variable positivity for neuroendocrine markers like NSE, vimentin and S 100 with no expression of WT-1 [9]. A combination of markers is generally helpful in arriving the correct diagnosis. Cytogenetic study of renal PNET is a supportive method to make the final diagnosis. Most of the tumors show chromosomal translocation $t(11;22)(q24;q12)$ [10]. Hence the diagnosis of renal PNET is confirmed by histopathology and immunohistochemistry in nephrectomy specimens and is supported by the cytogenetic study (not done in our case). Multi-modal treatment approach including surgery (radical nephrectomy), adjuvant chemotherapy

(vincristine, adriamycin, cyclophosphamide with alternating ifosfamide and etoposide every 21 days – VAC/IE) and radiotherapy is the cornerstone for the management of this aggressive renal malignancy. Adjuvant radiation therapy is recommended for patients with incomplete resection, positive resection margins, or recurrence [9]. The overall prognosis of this condition is poor due to lack of discrete clinical and radiological findings which make preoperative diagnosis difficult, the strong tendency of local recurrence and remote metastases to organs such as lungs, liver, and bone.

Conclusions

Renal PNET is a rare renal malignancy that should be kept in the differentials of a renal SOL presenting in adolescents and young adults. Histopathology, immuno histochemistry supported by cytogenetic studies are required for the exact diagnosis. Multidisciplinary approach consisting of surgery, chemotherapy, and radiotherapy is recommended to manage this condition in view of its aggressive nature and poor prognosis. All diagnostic modalities concerning space-occupying lesions (SOL) of the kidney must be interpreted with caution in order for the appropriate management. Punctures for cytology can be indicated in select cases.

Authors contribution

1. Dr. G. Garg – Concept, design, supervision, processing, writing manuscript and critical analysis.
2. Dr. R. Aeron – Concept, design, supervision, processing, writing manuscript and critical analysis.
3. Prof. B. P. Singh – Supervision, processing, writing manuscript and critical analysis.
4. Dr. S. Agarwal – Concept, supervision, writing manuscript and critical analysis.

Informed consent

Written informed consent was obtained from the patient.

Conflict of interest

No conflict of interest was declared by the authors.

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