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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/m^2 on day 1, doxorubicin 20 mg/m^2 on days 1–3, etoposide 150 mg/m^2 on days 1–3, and ifosfamide 3 g/m^2 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 × 10^9/l and Platelets 156 × 10^9/l; blood urea 4.9 mmol/l and creatinine 70 μm/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 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/m^2 on day 1, doxorubicin 20 mg/m^2 on days 1–3, etoposide 150 mg/m^2 on days 1–3, and ifosfamide 3 g/m^2 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.

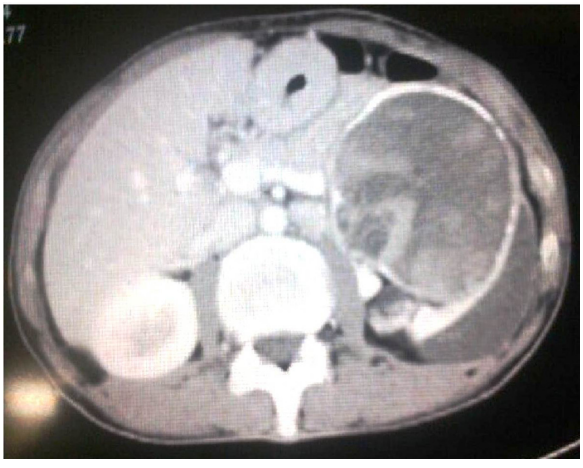

Figure 1 Computerized tomography scan image of abdomen depicting mildly enhancing large roundish mixed density solid cystic lesion in left kidney with interrupted peripheral rim calcifications. The lesion also shows internal non-enhancing necrotic areas and adjacent perinephric fat stranding.

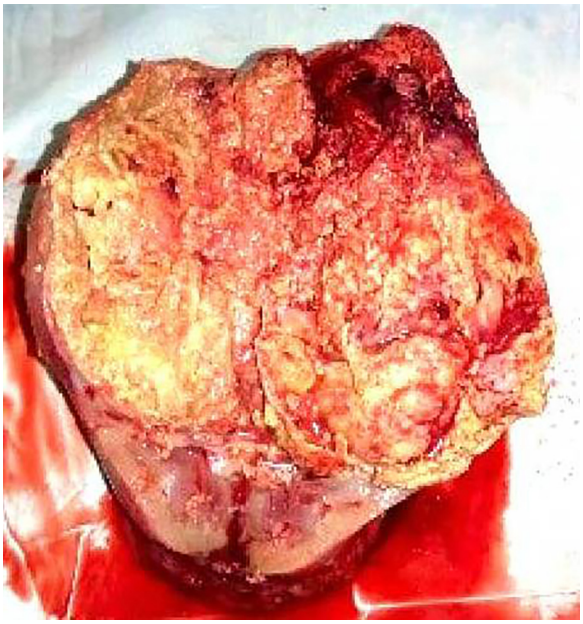

Figure 2 Image showing surgically excised left kidney depicting large encapsulated lesion with solid cystic areas.

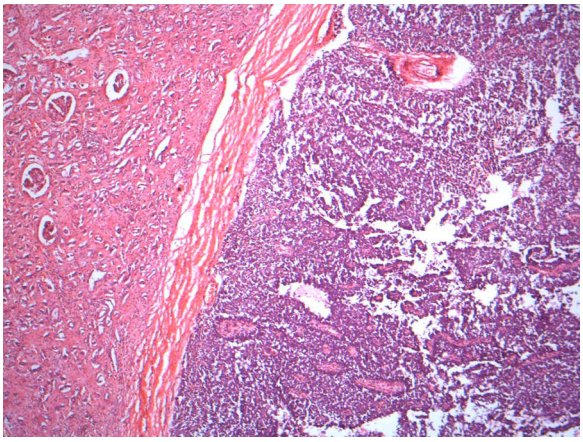

Figure 3 Scanner view showing an encapsulated tumor tissue showing small round cells.

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