The Role of Surgery for Local Recurrence of Renal Ewing's Sarcoma
A Case Report

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INTRODUCTION

Primitive neuroectodermal tumor (PNET) is a rare malignancy mainly developed in the central nervous system and soft tissues of the children; however, primary occurrence of this tumor in the kidney has been recently reported.[1,2] Histological diagnosis of such cases is challenging. We report a case with recurrence of left renal PNET that was treated successfully by mere tumor resection.

CASE REPORT

In February 2003, a 22-year-old man presented to our center with the chief complaint of gross hematuria and pain in the left flank. Medical history and physical examination were unremarkable. Except for a microscopic hematuria, all laboratory tests had normal results. Ultrasonography revealed a 145 × 106-mm heterogeneous mass in the upper pole of the left kidney without the evidence of the metastatic disease. The findings were confirmed by CT scan with mild enhancement after contrast injection (Figure 1A). No tumoral tissue was detected in the inferior vena cava and the renal veins by magnetic resonance imaging. Chest radiography was also normal.

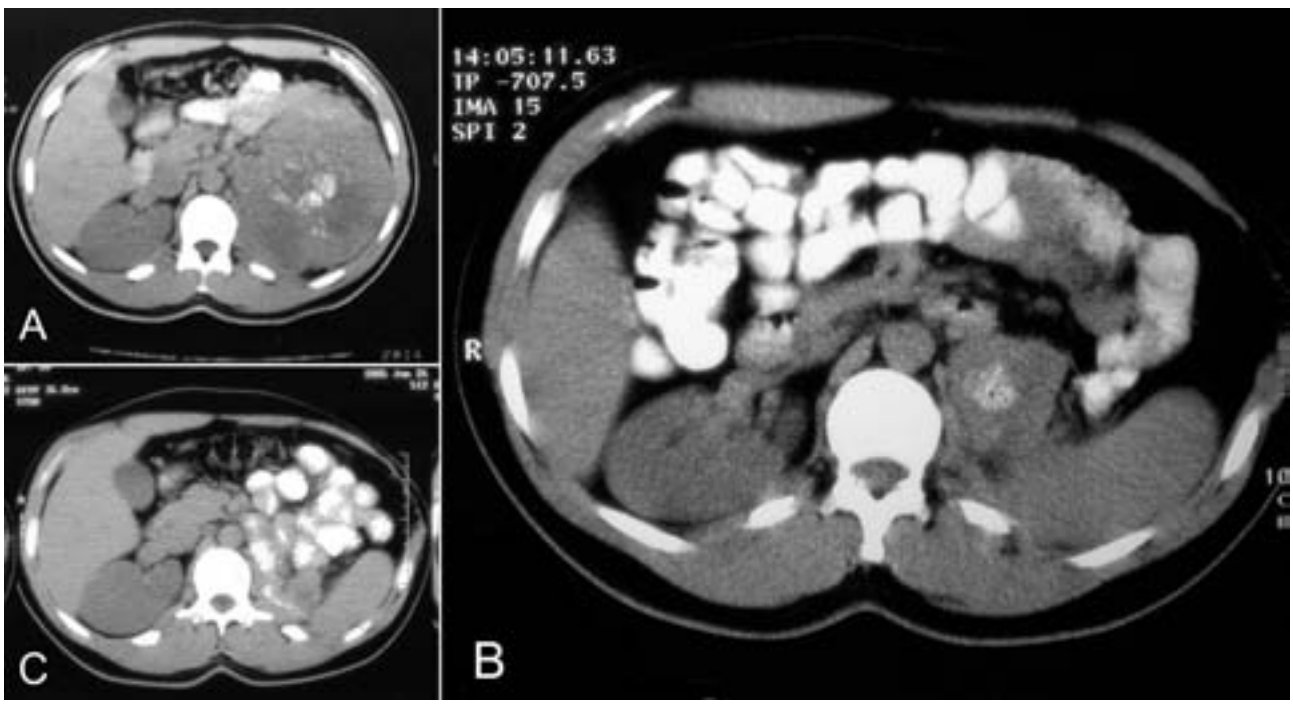

Figure 1. Diagnostic and follow-up CT scans. **A**, Primary renal tumor. **B**, Local recurrence. **C**, Last follow-up.

With the probable diagnosis of primary left kidney tumor, radical nephrectomy was performed. Histopathological assessment demonstrated uniform, small, round cells compatible with a stage II blastemal-type Wilms tumor. The surgical margins were free of tumor.

The patient underwent chemotherapy afterwards. But due to its complications, he refused to continue the treatment after 3 courses of chemotherapy. He was only followed with biochemistry, chest radiography, and abdominal and pelvic CT scan every 3 months. At the 12th postoperative month, CT scan revealed a 40 × 50-mm mass with central calcification in the left renal fossa (Figure 1B). Since chemotherapy was refused by the patient, resection of the mass was performed. It was excised with a margin of the psoas muscle and the colon mesentery to which it was adhered (Figure 2A).

Pathologic findings were identical to the primary kidney tumors such as a small round-cell sarcoma with extensive necrosis. Although all margins were tumor-free, immunohistochemical assessment of the specimen was performed due to the rosette formation (Figure 2B). The patient had a periodic acid-Schiff staining as focal fine granular cytoplasmic depositions. On immunohistochemistry, the neoplastic cells showed cytoplasmic staining of synaptophysin and cell membrane staining for microneme protein 2, but no reactivity was noted for desmin, actin, terminal deoxynucleotidyl transferase, and Wilms tumor suppressor (WT1). We additionally performed fluorescence in situ hybridization analysis which showed a split signal with DNA probes against the *EWS* gene, indicating *EWS* rearrangement.

Overall, histology and immunohistochemistry studies confirmed the diagnosis of Ewing's sarcoma (Figure 2C). The tumor was qualified as a renal PNET. Again, the patient refused further chemotherapy and he remained disease-free (within 16 months of follow-up after the recurrence). On the last follow-up visit, CT scan of the abdomen and pelvis was negative for metastatic disease or local recurrence (Figure 1C) and the patient was in a good general condition.

DISCUSSION

Primitive neuroectodermal tumor should be differentiated from other small round cell tumors of the kidney including lymphoma, small cell carcinoma, renal carcinoid tumor, neuroblastoma, rhabdomyosarcoma, blastema-predominant Wilms tumor, synovial sarcoma, and desmoplastic round cell tumor.[3]

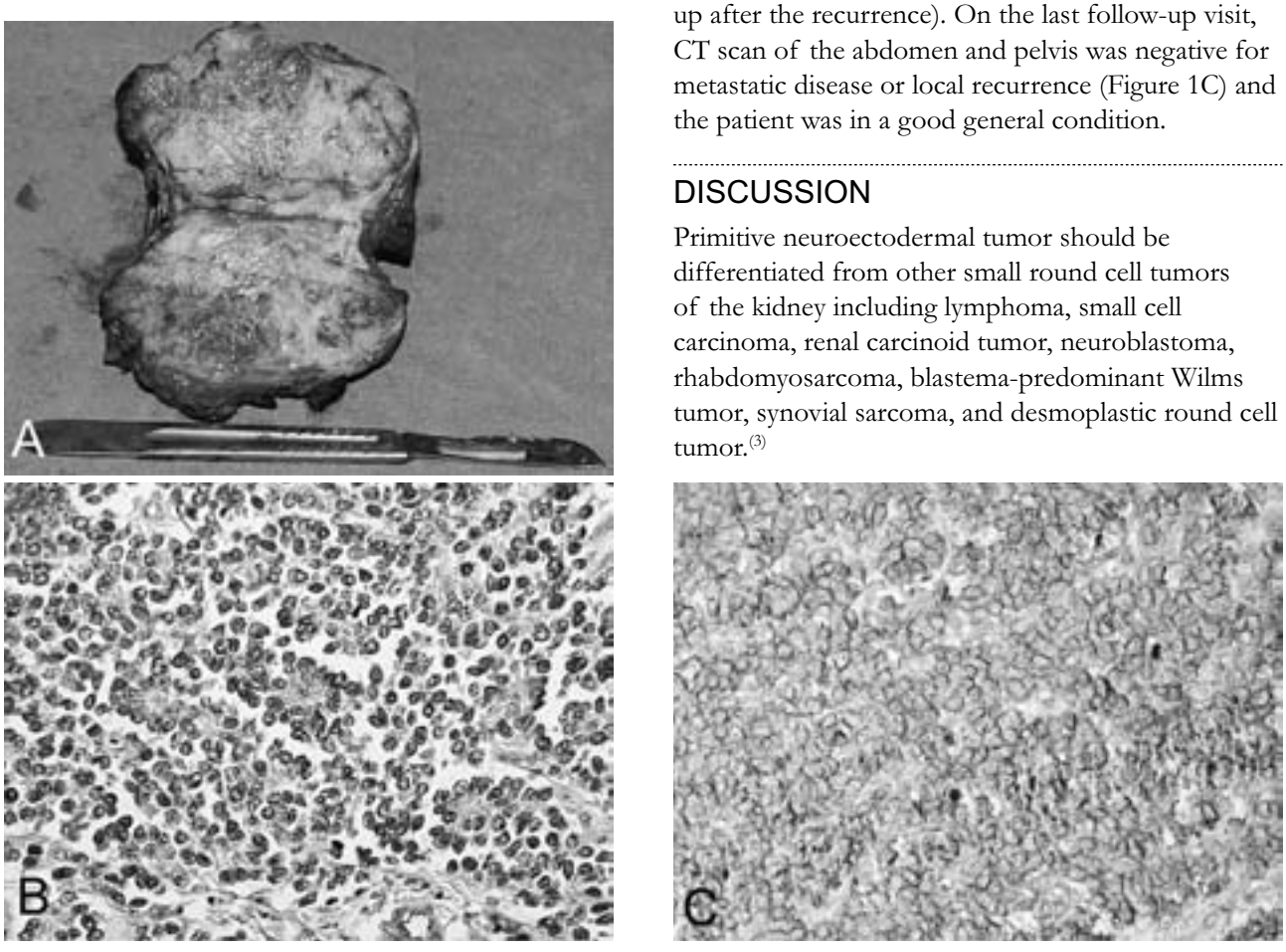

Figure 2. A, Excised recurrent tumor. **B**, Sheets of small cells with rosette formation (hematoxylin-eosin, × 400). **C**, The cell membrane staining of MIC2 in the neoplastic cells (× 400)

Renal PNET was first reported by Mor and colleagues.[4] Thereafter, a large number of renal Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) cases have been reported in the literature.[1,2,5,6] Most of the reported cases have been found among the young adults (average age, 28 years) with a slight male predominance (male-female ratio, 1.5 to 1).[6]

Radiotherapy or chemotherapy is not effective and the prognosis is poor in most cases with a reported 5-year survival rate of 45% to 55%.[7] Current chemotherapy protocols used in the treatment of the children with ES/PNET is composed of induction and consolidation cycles with vincristine, doxorubicin, cyclophosphamide, ifosfamide, and etoposide.[6] Further investigation of these regimens in patients with renal ES/PNET is warranted to determine their precise impact on the survival.

These tumors have a great tendency to develop metastases and subsequently a poorer prognosis is expected.[2,5,6] Although there is a report of spontaneous regression of pulmonary metastasis following nephrectomy,[8] most patients die within an average duration of 16 months despite chemotherapy.[6]

To our best knowledge, this is the first report in which mere resection of the recurrent tumor in the kidney could yield at least 16 months of being free of tumor or metastasis. Thus, re-exploration and removal of the recurrent tumor in cases of renal PNET may have a decisive role in the treatment of local recurrences of renal Ewing's sarcoma.

CONFLICT OF INTEREST

None declared.

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