PNET/EWING’S SARCOMA OF THE KIDNEY: IMAGING FINDINGS IN TWO CASES

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The CT-imaging findings of primary renal PN.EWING’s sarcoma in two patients were retrospectively assessed. A large renal mass with heterogenous contrast enhancement and necrotic and hemorrhagic areas were the predominant characteristics. In adolescents or young adults presenting with a large renal mass, PN.EWING’s sarcoma may be included in the differential diagnosis.

Primitive Neuroectodermal Tumor (PNET) or EWING’s sarcoma of primary renal origin is a very rare entity with aggressive behavior. Affected patients are adolescents or young adults, with variable and aspecific clinical presentation. In most cases the presence of a renal tumoral mass is assessed with ultrasound or computed tomography, but the diagnosis of PN.EWING’s sarcoma is usually made only on histopathology after nephrectomy, based on immunohistochemical and cytogenetic tests. We report on the CT-imaging findings in two cases of renal PN.EWING’s sarcoma.

Case presentations

In 2011 two new cases of PN.EWING’s sarcoma of primary renal origin were diagnosed at our institutions and treated with radical nephrectomy including regional lymph node dissection and adjuvant chemotherapy. The patients were a 24-year-old female (Fig. 1) and a 26-year-old male (Fig. 2) presenting with vague abdominal pain and without remarkable clinical records. The histopathology reports revealed small round blue cells with Homer-Wright type rosette formation on hematoxylin and eosin staining, and showed no vascular tumor thrombus. Both tumors consisted of solid components with hemorrhagic, cystic and necrotic areas, resulting in heterogeneous attenuation numbers on unenhanced and contrast-enhanced CT scans. The solid components showed moderate enhancement. The tumors were at least partially surrounded by a small rim of healthy renal cortex. In one case, the tumor showed sparse and diffuse microcalcifications, histologically confirmed as dystrophic calcifications in necrosis. Enlarged retroperitoneal lymph nodes were noted in one patient, proven non-metastatic following regional lymphadenectomy.

Discussion

PN.EWING’s sarcoma of the kidney is an exceedingly rare entity, only about 50 cases were reported in literature (1, 2). The true incidence however, may have been underestimated as the number of case reports is increasing in recent years, probably due to advanced immunohistochemistry, enabling better characterization of renal tumors (1). PN.EWING’s sarcoma of the kidney typically occurs in adolescents and young adults with a reported median age of 24-27 years (1, 3). The clinical presentation is variable and aspecific with patients complaining of flank pain (85%), palpable abdominal mass (60%), hematuria (37%) or weight loss (8%) (1, 4).

Histopathology is essential to the diagnosis of renal PN.EWING’s sarcoma as demonstrated by the expression of neural immunohistochemical markers such as NSE, vimentin, synaptophysin and S-100 (1). Imaging findings in PN.EWING’s sarcoma are variable and nonspecific. The CT-appearance of PN.EWING’s sarcoma is characterized by a large renal mass with heterogeneous contrast enhancement, necrotic or hemorrhagic areas and sometimes calcifications (1). However, the radiological features are indistinguishable from other primary, malignant renal parenchymal tumors or urorheal cancer (1, 2, 4). Other tumors that may be considered in the differential diagnosis of PN.EWING’s sarcoma included in the differential diagnosis.
diagnosis include rhabdomyosarcoma, Wilms’ tumor, carcinoid tumor, neuroblastoma, lymphoma, desmoplastic small round cell tumor and nephroblastoma (1, 3, 5). Since no pathognomonic features of PNET/Ewing’s sarcoma have been described, it is difficult to preoperatively diagnose this entity (1, 4). Nevertheless, in adolescents or young adults presenting with a renal mass, PNET/Ewing’s sarcoma may be included in the differential diagnosis and a preoperative fine needle aspiration or core needle biopsy can be taken into consideration (3).

Most PNET/Ewing’s sarcomas have an aggressive behavior and the majority of patients present at advanced stage disease (57.6%), including lymph node invasion (25%), pulmonary metastases (20%) and liver metastases (14%) (1-5). One third of patients present with tumor thrombi in the renal vein or inferior vena cava at the time of diagnosis (1).

The management principles of renal PNET/Ewing’s sarcoma have been extrapolated from osseous Ewing’s sarcoma, although the sequence of neoadjuvant chemotheraphy followed by surgery, which is the standard of care in osseous Ewing’s sarcoma, is usually not relevant in renal PNET/Ewing’s sarcoma since surgery is for the majority of cases the initial and necessary step to the diagnosis (2). The treatment options should be further investigated, but regarding the small number of patients with renal PNET/Ewing’s sarcoma this is hard to achieve. Therefore radical nephrectomy still remains the most important modality of treatment (1, 3). Since most patients with apparently localized disease do have occult metastasis, additional polychemotherapy is usually warranted. The standard chemotherapeutical agents currently used are vincristine, ifosfamide, doxorubicin, etoposide, etoposide, Adriamycin and ifosfamide (1, 2). The role of radiotherapy is not clear, but it may be indicated in case of positive surgical margins or involvement of Gerota’s fascia (3).

The prognosis of patients with renal PNET/Ewing’s sarcoma is poor, with high local recurrence rates and only a minority of patients experiencing long disease-free survival (1-4).
Conclusions

The CT-appearance of PNET/Ewing’s sarcoma of primary renal origin is that of a large renal mass with heterogeneous contrast enhancement, necrotic or hemorrhagic areas and occasional calcifications. It is virtually indistinguishable from other renal tumors such as renal cell carcinoma. Nevertheless, in adolescents or young adults presenting with a large renal mass the diagnosis of PNET/Ewing’s sarcoma may be suggested and a preoperative fine needle aspiration or core needle biopsy can be taken into consideration. Radical nephrectomy remains the most important modality of treatment, although neoadjuvant chemotherapy preceding nephrectomy might be a possible treatment alternative.

References