Primitive Neuroectodermal Tumor of the Kidney: a case report and literature review

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Primitive neuroectodermal tumors (PNET) are sarcomas that arise most commonly in the central nervous system. The much less common peripheral PNETs have been reported in many body sites, but renal PNET is extremely rare. Most renal PNETs occur in children or adolescents. We report a 40-year-old man presenting with an enlarging right flank mass for one month. Computed tomography (CT) demonstrated a large exophytic soft tissue mass with heterogeneous enhancement originating from the right kidney and containing areas of necrosis and hemorrhage. The tumor was completely resected, and the pathological examination revealed a renal PNET. The imaging findings of this tumor were discussed, and the literatures were reviewed.

Primitive neuroectodermal tumors (PNET) was first recognized by Stout in 1918 [1-2] comprising a group of soft tissue sarcomas with neuroepithelial differentiation, which may arise in various parts of the body. These tumors are very similar to Ewing’s sarcoma. Both PNET and Ewing’s sarcoma belong to small-round-cell tumors. They share the same histochemical staining profile and a unique characteristic translocation t(11;22) or a variation of the same within the tumor cell [3]. Different terms have been used depending on their location and extent of neural differentiation [4]. Peripheral PNET is uncommon, accounting for only about 1% of all sarcomas [5]. The incidence of peripheral PNET in the abdomen and pelvis, including the retroperitoneum, is about 14% of all peripheral PNETs [5]. It has rarely been described in the genitourinary system, including kidney, bladder, prostate, testis, ovary, and uterus. Their typical radiological appearance resembled large non-calcified, soft tissue masses with cystic or necrotic areas; heterogeneous contrast enhancement was also evident on CT [6]. The peak incidence of PNET is in adolescence and young adulthood, with most reported patients younger than 35[5, 7]. We report a case of peripheral renal PNET in a 40-year-old man.

CASE REPORT

A 40-year-old man complained of a progressively enlarging right flank mass for about 1 month. He was a non-smoker with an unremarkable medical history. He denied any other symptoms in association with the mass. No body weight loss or hematuria was noted. He was referred from a local clinic to our urology clinic for further evaluation and management.

On initial physical examination, the right flank mass was movable and no tenderness felt during palpation. A plain abdominal film revealed increased soft tissue density in the right upper abdomen with downward displacement of the hepatic flexure.
of the colon, consistent with a space-occupying lesion. CT scan showed a large soft tissue mass measuring about 15 x 15 x 10-cm arising from the anterior aspect of the right kidney. On unenhanced images, the tumor appeared heterogeneous density with irregular areas of low, iso-, and slightly high attenuation, suggesting areas of necrosis and hemorrhage (Fig. 1). Few small calcifications were also identified. The mass enhanced heterogeneously in contrast enhanced scans (Fig. 2). The tumor caused anterior and left-sided displacement of the uncinate process of the pancreas and left-sided displacement as well as extrinsic compression of the inferior vena cava and right psoas muscle. The margins were well-defined, and there was no evidence of invasion of adjacent structures, vascular involvement or lymphadenopathy. The patient received radical right nephrectomy for a presumed malignant renal neoplasm.

On gross examination, the tumor was measured about 16 x 13 x 9-cm in size. The renal capsule was intact and the tumor was chiefly located within the renal parenchyma, effacing the renal cortex and pelvis. The renal vessels, ureter, perirenal fat, renal hilar and retroperitoneal lymph nodes were uninvolved. On section, the tumor contained areas of necrosis, hemorrhage, and cystic change. Microscopically, it was composed of small blue cells with frequent mitoses and nuclear pleomorphism, along with massive areas of necrosis and hyalinization. Immunohistochemical stains of the tumor cells were strongly positive for CD99 and vimentin and

Figure 1. a. b. c. Non-contrast computed tomography (CT) of the upper abdomen reveals a large soft tissue mass arising from the anterior aspect of the right kidney, which shows heterogeneity of the tumor with iso-attenuation areas (tumor), low attenuation areas (possible tumor necrosis), and slightly high attenuation areas (possible hemorrhage).

Figure 2. a. b. c. Contrast-enhanced CT images of the same axial sections as in figure 1 demonstrate heterogeneous enhancement of the tumor. The tumor mass causes displacement of adjacent organs (arrow in a: uncinate process of pancreas, dotted arrow in c: right psoas muscle) and compression of IVC (arrow in c) without evidence of direct invasions or retroperitoneal lymphadenopathy.
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diffusely and weakly positive for synaptophysin and chromogranin. The final pathological diagnosis was PNET of the kidney. The patient’s post-operative course was uneventful, and he had no evidence of recurrence on out-patient department follow-up.

**DISCUSSION**

Primary PNET of the kidney is extremely rare, and the clinical distinction from other primary renal malignancies is difficult [1-2, 8]. Renal PNET mainly affects adolescents and young adults with 75% of patients aged 10 to 39 years. The average age at diagnosis was around 27-year-old in several series [1-2, 5, 8-11]. Our patient was 40-year-old, therefore, was older than those in most renal PNET case reports. The clinical presentation of most patients is non-specific, including abdominal or flank pain, a palpable mass, and hematuria [1, 9, 11]. When renal PNET was diagnosed, it was often quite large and had already invaded adjacent organs. The mean tumor size was nearly 12 cm in a series of 11 cases [11]. The prognosis is poor, with a 5-year-survival rate of 45% to 55% [2, 5, 8-9]. Nearly 50% to 60% of patients were in advanced stage when diagnosis. The tumor most commonly metastasizes to lymph node, lung, bone, and liver [1-2, 5, 8-9]. These clinical features, however, do not help to distinguish PNET from other renal tumors.

Despite current advances in imaging modalities, no specific signs or characteristics for renal PNET have been described on ultrasound, CT, or MRI. Renal PNET often presents as a large, ill-defined mass with areas of necrosis and hemorrhage. On unenhanced CT images, the tumor appears as a mixed density with solid and cystic areas. Multifocal low density necrotic areas are often seen throughout the tumor as well as high density foci indicating hemorrhages. The tumors show a variable degree of heterogeneous contrast enhancement. Internal septation and calcifications are rarely reported [5, 9, 12]. On ultrasonography, echogenicity in comparison to normal renal parenchyma varies considerably in different reports, from hypoechoic to isoechoic or hyperechoic. On MRI, the tumor appears iso- to hypointense on T1-weighted images, but heterogeneous on T2-weighted images [9]. Most importantly, imaging modalities provide accurate assessments of local tumor extent, as the tumor often shows aggressive behaviors. Renal PNET often directly invade adjacent organs such as the pancreas, spleen, stomach, and psoas muscles. Vascular involvement is rare, venous thrombi in the renal vein and inferior vena cava was reported in only a few cases [1, 5, 9]. Different from most renal PNET cases in the literature review, our patient’s tumor, though quite large and exhibiting areas of necrosis and hemorrhage on CT, was not locally invasive.

A definite diagnosis of a large aggressive renal tumor with necrosis and hemorrhage is difficult basely on image studies, since nearly all aggressive renal malignancies demonstrate this uncharacteristic image finding. For children and adolescence, distinction from other primary malignancies of kidneys is crucial for prognosis. The differential diagnosis includes extraosseous Ewing’s sarcoma, rhabdomyosarcoma, Wilms’ tumor, neuroblastoma, carcinoid, clear cell sarcoma of the kidney, lymphoma, small cell variant of osteosarcoma and dysplastic small round cell tumor [1-2, 8].

In adults, such as our patient, renal cell carcinoma should still be the primary consideration when patients present with a large palpable flank mass with necrosis and hemorrhage. Calcification is also occasionally identified in renal cell carcinoma. Other possibilities including transitional cell carcinoma, lymphoma, and angiomylipoma complicated with rupture and hemorrhage should also figure into the differential diagnosis. Tumors arising from the adjacent retroperitoneum, such as neuroblastoma and rhabdomyosarcoma, may also have similar appearances on image studies, although these usually displace the kidney [2, 12], which distinguishes them from a primary renal tumor.

The diagnosis of PNET is thus based on the pathological examination. Microscopically, the tumor is composed of small undifferentiated neuroectodermal cells, which appear as small round blue cells on hematoxylin and eosin staining; the cells may form rosettes or pseudorosettes [1, 8-10, 13]. On immunohistochemical stains, the tumor cells express features of neuronal differentiation, such as neuron-specific enolase, synaptophysin, chromogranin, and vimentin, but these findings are not pathognomonic. More specifically, PNET cells expresses high amounts of surface membrane protein MIC2 antigen (CD99) and exhibit chromosomal translocation between chromosome 11 and 22 (t(11,22)(q24;q12)). Our patient’s tumor cells had neuronal differentiation, as they were diffusely positive for vimentin, synaptophysin, and chromogranin. The strong and diffuse positivity for CD99 of the small round blue tumor cells makes the diagnosis of renal PNET in this case.
CONCLUSION

Although advances in imaging have greatly improved our diagnostic ability in many diseases, it is well to remember that limitations remain. Operation is still the first choice in the treatment of a large renal tumor, and the definite diagnosis can be made by pathological examination. The differential diagnosis of a large renal tumor should include PNET.

REFERENCES

腎臟原始神經外胚層腫瘤：病例報告與文獻回顧

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原始神經外胚層腫瘤是一種罕見的惡性肉瘤，大多數來自中樞神經系統。發源於身體周邊並不常見，而源自於腎臟的原始神經外胚層腫瘤更是少之又少。大多數文獻指出，原發性的腎臟原始神經外胚層腫瘤多發生於小孩或年輕人身上。我們報告了一位40歲男性病患，因右腰部無痛腫塊求診，影像上顯示一個巨大的右腎腫瘤，注射顯影劑後，腫瘤呈現不均勻的顯影，並夾有壞死及出血。最終開刀後的病理結果為罕見的腎臟原始神經外胚層腫瘤。文中將描述影像上的發現並作相關文獻的回顧。