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Clear-cell sarcoma of the kidney in a child – case report

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Summary

Background: Kidney tumours other than nephroblastoma (Wilms' tumours) occur relatively rarely in the pediatric population. They include but are not limited to: nephroma mesoblasticum, rhabdoid tumour of the kidney and clear-cell sarcoma of the kidney.

Case Report: In this article we presented a case of a 20-month-old boy with clear-cell sarcoma of the kidney.

Conclusions: Imaging studies – including abdominal and chest CT examinations – allow to determine the extent of the disease and help in the initial assessment of the tumour character, as well as in planning the treatment, thus having a positive prognostic impact on the pediatric patients.

MeSH Keywords: Acute Kidney Injury • Child • Diagnostic Imaging

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Background

Nephroblastoma, or Wilms' tumour, is the most common malignant renal neoplasm in children. In Poland there are approximately 80 new cases diagnosed per year [1].

Apart from nephroblastoma, renal tumours are relatively rare in children, which is unanimously emphasized by pediatric oncologists who belong to NWTS (National Wilms Tumor Study Group), SIOP (International Society of Paediatric Oncology) and other research groups.

It should be noted here that there is a great variety of renal tumours in children that used to be classified in the past as Wilms' tumour. In the recent years a few of these tumours have been carefully described and recognized as separate pathological entities. A correct diagnosis can be suggested by clinical presentation, child's age at tumour occurrence and distinctive features in imaging studies.

These tumours include but are not limited to: nephroma mesoblasticum, rhabdoid tumour of the kidney and clear-cell sarcoma of the kidney.

The knowledge about the nature of these lesions can suggest their genetic background, which affects treatment planning and further prognosis for the patients. However,

even with use of the state-of-the-art imaging techniques (such as advanced magnetic resonance sequences), it is impossible to assess tumour type accurately without a histopathological examination.

Case Report

A 20-month-old boy was admitted to our center due to a massive hematuria observed for several days. On physical examination on admission he had a palpable large abdominal mass, crossing the midline from the right to the left. In additional examinations the following findings were noted: elevated inflammatory markers, reduced cell counts, elevated level of lactate dehydrogenase and massive hematuria. In the abdominal CT scan there was a huge, heterogenous, hypodense tumour mass with calcifications, located in the right mid-abdomen, measuring approximately 115×100×127 mm, originating from the lower part of the right kidney (Figure 1). Based on the radiological findings a right renal nephroblastoma was suspected. The child was referred to Children's Memorial Health Institute in Warsaw. A right nephrectomy was performed and a diagnosis of clear-cell sarcoma of the kidney was made based on histopathological examination. The patient underwent subsequent chemotherapy. In the further course of the disease a relapse occurred after almost three years, in the form of metastasis to the right femur complicated by its fracture (Figure 2) – at that moment the patient completed a 2-year chemotherapy course.



Figure 1. An abdominal CT scan. **(A)** MPR reconstruction. **(B)** Axial plane. A 20-month-old boy. Clinical observations – a large abdominal tumour, massive hematuria in laboratory examinations. A giant heterogeneous, hypodense mass with calcifications – clear-cell sarcoma of the right kidney (asterisk).

Discussion

Clear-cell sarcoma of the kidney (CCSK) used to be regarded as one of the variants of Wilms' tumour. It accounts for 4% [2] of renal tumours diagnosed during childhood. It is considered to be an extremely aggressive lesion with poor clinical outcome. The tumour occurs in children aged 1–4, the highest incidence is between 2 and 3 years of age [3] and there is a slight male predominance [4]. On the other hand, the incidence peak of Wilms' tumour is between 2 and 5 years of age, on average at 3.7 years. In all the cases described in the literature CCSK was unilateral, unlike Wilms' tumour. The latter can occur bilaterally in up to 5% of cases and in such case it coexists with nephroblastomatosis (precancerous lesions). Both clinical symptoms and



Figure 2. X-ray of the femur. **(A)** Anteroposterior projection. **(B)** Lateral projection. A metastatic lesion of the femoral shaft. A pathological fracture.

imaging findings are similar in the patients with CCSK and Wilms' tumour. However, clear-cell sarcoma of the kidney is far more aggressive than nephroblastoma, and is associated with higher relapse and mortality rate. One of the most typical features of CCSK is its propensity to metastasize to bone (40–60%), which accounts for much higher rate of bone metastases compared to the patients with Wilms' tumour (approximately 2%) [5]. Bone metastases can present as both lytic and sclerotic lesions. They can also mimic other lesions – including benign ones, which can lead to a delay in diagnosis and treatment [4]. Additionally, CCSK can metastasize to atypical sites, such as scalp, extradural space, nasopharynx, neck, paraspinal area, abdominal wall, axilla and orbita [6]. This can result from its tendency to infiltrate and spread through the lymphatic vessels. Making early and correct diagnosis is prevented by high degree of tumour malignancy, high risk of metastasizing to bones, tendency to late relapse (up to 4 years from the primary diagnosis [6]) and relatively poor treatment results compared to Wilms' tumours [2].

In the imaging studies CCSK usually presents as a large, well-circumscribed mass, which distorts or replaces a kidney. Hemorrhagic and necrotic components can be observed in 70% of these lesions. In rare cases multiple cysts can be a dominant feature [7]. In 70% of patients there is a renal capsule infiltration present at first examination. Infiltration of the renal vein is very rare.

References:

1. Sawicz-Birkowska K: Nerczak płodowy u dzieci (guz Wilmsa, nephroblastoma). Nowotwory wieku dziecięcego. *Onkol Prakt Klin*, 2007; 3: 673–79 [in Polish]
2. Boo YJ, Fisher JC, Haley MJ et al: Vascular characterization of clear cell sarcoma of the kidney in a child: a case report and review. *J Pediatr Surg*, 2009; 44: 2031–36
3. Sebire NJ, Vujanic GM: Paediatric renal tumours: recent developments, new entities, and pathological features. *Histopathology*, 2009; 54: 516–28
4. Lowe LH, Isuani BH, Heller RM et al: Pediatric renal masses: Wilms tumor and beyond. *Radiographics*, 2000; 20: 1585–603
5. Miniati D, Gay AN, Parks KV et al: Imaging accuracy and incidence of Wilms and non-Wilms renal tumors in children. *J Pediatr Surg*, 2008; 43: 1301–7
6. Zhuge Y, Cheung MC, Yang R: Pediatric on-Wilms renal tumors: subtypes, survival, and prognostic indicators. *J Surg Res*, 2010; 156(2): 257–63
7. Prasad SR, Humphrey PA, Menias CO et al: Neoplasms of the renal medulla: radiologic-pathologic correlation. *Radiographics*, 2005; 25: 369–80
8. Kural AR, Onal B, Ozkara H et al: Adult clear cell sarcoma of the kidney: a case report. *BMC Urol*, 2006; 6: 11
9. Bonadio J, Perlman EJ: Immunohistochemical analysis of 61 clear cell sarcomas of the kidney for a panel including NGFR and CD99. *Mod Pathol*, 2008; 21: 218A
10. Balarezo FS, Joshi VV: Clear cell sarcoma of the pediatric kidney: detailed description and analysis of variant histologic patterns of a tumor with many faces. *Adv Anat Pathol*, 2001; 8(2): 98–108
11. Argani P, Perlman EJ, Breslow NE et al: Clear cell sarcoma of the kidney: a review of 351 cases from the National Wilms Tumor Study Group Pathology Center. *Am J Surg Pathol*, 2000; 24(1): 4–18

Immunohistochemical examinations are useful in differentiating CCSK from other childhood tumours. In case of CCSK there were positive results reported for vimentin and CD-99 [8–11]. Bonadio and Perlman [9] analyzed 61 cases of CCSK and showed in all the examined cases the presence of nerve growth factor receptor (NGFR). It is also well documented that CCSK is negative to cytokeratin, Mic-2, S100, neural markers, desmin and WT-1 [10,11].

Treatment consists of nephrectomy and subsequent chemotherapy.

Long-term survival rate of patients with diagnosed CCSK is between 60 and 70%. Unlike CCSK, prognosis in the patients with Wilms' tumour is very good – a 2-year survival rate reaches 90% and it usually means a complete cure.

Conclusions

CCSK should be taken into account in differential diagnosis of renal tumours in children.

Final diagnosis requires a histopathological confirmation.

Imaging studies – including CT scan of the chest and abdomen – allow to determine the extent of the disease and help in the initial assessment of the tumour character, as well as in planning the treatment regimen, thus having a positive prognostic impact on the pediatric patients.