Renal Cell Carcinoma in a 3 year-old Girl: a case report

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A 3-year-old girl presented with vomiting for two days. Abdominal ultrasonogram revealed a mass at the lower pole of left kidney. Non-contrast enhanced abdominal computed tomography(CT) scan revealed high attenuation of the tumor and calcified lymphadenopathies in the retroperitoneum. On a contrast-enhanced CT scan, the mass was enhanced heterogenously. The patient underwent left radical nephrectomy and resection of adjacent lymph nodes. The pathology report confirmed the diagnosis of renal cell carcinoma with metastatic lymphadenopathies. Calcified metastatic lymph nodes adjacent to a renal mass may suggest the possibility of renal cell carcinoma.

Renal cell carcinoma (RCC) is predominantly a malignancy of adults. In children, the most common primary renal cancer is Wilms’ tumor, accounting for 90% of pediatric renal malignancies [1]. RCC is extremely rare in patients younger than 20 years, accounting for less than 1% of all cases of RCC [2]. When reported, it has usually been diagnosed in preteens or adolescents [3, 4]. We describe a 3-year-old girl with RCC and the image findings of computed tomography(CT).

CASE REPORT

A 3-year-old girl known to have a type II atrial septal defect was evaluated for vomiting for two days. There was no other significant medical history. Physical examination and blood tests were unremarkable. On urinalysis, there were 2 white cells per high power field but no hematuria. An abdominal ultrasonogram revealed a solid mass in the lower pole of left kidney. Non-contrast enhanced abdominal CT revealed a 5 x 7 x 10-cm, lobulated mass with high attenuation at the lower pole of left kidney, as well as enlarged, calcified, perirenal and para-aortic lymph nodes (Fig. 1a). After contrast administration, the tumor was enhanced heterogenously (Fig. 1b). These CT findings were highly suggestive of a malignant renal tumor with retroperitoneal lymphadenopathies.

The patient underwent left radical nephroureterectomy with lymph node dissection. The gross specimen was grayish white and soft, with areas of hemorrhage (Fig. 2a). Microscopically, the tumor was composed of clear cells growing in a papillary pattern (Fig. 2b, 2c). Immunohistochemical studies revealed neoplastic cells that were positive for cytokeratin (AE1/AE3), vimentin, and CD10, but negative for desmin and HMB-45, consistent with a papillary renal cell carcinoma. All hilar lymph nodes and one para-aortic node showed metastatic tumor cells and psammoma bodies (Fig. 2d). No other treatment was...
Figure 1. **a, b**, Non-contrast enhanced CT revealing a lobulated mass with higher attenuation than adjacent normal renal parenchyma (arrows), as well as enlarged, calcified lymph nodes (arrowheads). **c**, Contrast-enhanced CT scan with heterogenous enhancement of the tumor.

Figure 2. **a**, Gross appearance of the tumor, which is grayish white with areas of hemorrhage in the lower pole of the kidney. **b**, Microscopically, the tumor cells are arranged in a papillary pattern (left) as compared to normal renal tissue (right). (H&E, x 40) **c**, The tumor consists predominantly of clear cells growing in papillary pattern (arrows). (H&E, x 100) **d**, Psammoma bodies (arrows) in a metastatic lymph node (H&E, x 200), indicating round collections of calcium.
given, and the child was well at one-year follow-up.

**DISCUSSION**

The case reported here is of interest both for the young age of the patient at diagnosis, and for the presence of calcified lymph node metastases. In a review of 16 cases of pediatric RCC, including 5 boys and 11 girls, collected over 17 years from 3 centers in Canada, the mean age at diagnosis was 9.6 years (range 3 to 19 years) [3]. A similar series from Boston reviewed 11 cases seen over 38 years. The mean age at presentation was 14.7 years (range 9.3 to 17.6 years), and there was a 2.7:1 female predominance [4]. Our patient, therefore, was diagnosed at a considerably younger age than the above-mentioned reported pediatric patients.

The diagnosis in our patient, in fact, was probably fortuitous. Her presenting symptom of vomiting might have been incidental rather than directly related to the tumor. The most common presentation of RCC in children is a palpable mass, followed by gross hematuria [5], but the complete clinical triad — gross hematuria, palpable mass, and flank pain — is reportedly seen in only 6% to 9% of affected children [2]. Paraneoplastic symptoms such as polythecemia and hypertension, often noted in adults, are uncommon in pediatric RCC [7].

The disease in adults is not uncommonly associated with certain hereditary genetic defects, particularly Von Hippel-Lindau disease, and several children with RCC associated with Von Hippel-Lindau syndrome or tuberous sclerosis have been reported [1]. No hereditary predisposition was evident in our patient. Histologically, adult RCC is predominantly of clear cell type, while a greater proportion of children have papillary histology [3].

In adults, the CT appearance varies according to the cell type of the tumor. RCC is usually seen as an exophytic or infiltrating intrarenal mass that is hypervascular and usually heterogenous in clear cell tumors, homogenous in chromophobe tumors, and hypovascular in papillary tumors [8]. Given the rarity of the disease in children, the CT imaging features of pediatric RCC are not well defined. Our patient’s papillary tumor composed of clear cells did enhance heterogenously but did not appear to be hypervascular. Generally speaking, the CT features of pediatric RCC and the much more common Wilms’ tumor do not differ sufficiently to make a definitive preoperative diagnosis. In adults, calcification is more frequently found in papillary and chromophobe tumors [7], while only 10% to 15% of clear cell RCC is calcified [8]. There is little comment on calcification in pediatric RCC, but what is particularly interesting in our case is the calcified metastatic lymphadenopathies. The only available report of calcified RCC, 2 of 4 children with RCC reported from Japan had calcified metastatic lymph nodes [2], a finding that would be distinctly unusual in Wilms’ tumor. Such an imaging finding in a child with a renal tumor, then, might be more suggestive of RCC.

RCC is treated with radical nephrectomy and regional lymph node dissection. The overall prognosis is dependent upon tumor stage, patient’s age, tumor size, histologic pattern and chromosome abnormality [2, 5]. However, local lymph node involvement does not apparently predict a poor prognosis for children as it does for adults [9]. The roles of adjuvant radiotherapy, chemotherapy, and immunotherapy remain unclear in pediatric RCC [10].

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三歲女童發生腎細胞癌：病例報告

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一位三歲女童因嘔吐兩天而求診。腹部超音波顯示腫瘤位於左腎下級實質部。未顯影電
腦斷層檢查中顯示此腫瘤具有高的密度伴併有鈣化的後腹腔淋巴結腫大。於施打顯影劑後，
腫瘤呈現均勻的顯影。病人之後接受左腎全切除以及周圍淋巴腺廓清術，病理報告證實為腎
細胞癌合併淋巴腺轉移。因此一腎臟腫瘤合併有周圍鈣化淋巴腺轉移提供小兒腎細胞癌的一個
診斷線索。