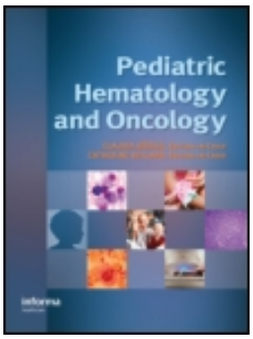

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Letter

RENAL CELL CARCINOMA IN CHILDHOOD: A Case Report

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Renal cell carcinoma (RCC) is the most common primary malignancy of the adult kidney, accounting for 2% of adult cancer cases, but it occurs only rarely in children [1–6]. For a number of years, RCC was thought to have originated in adrenal rests within the kidney, but immunohistologic and ultrastructural analysis has now established that the proximal renal tubular epithelium is the tissue of origin [7]. This paper presents a pediatric case of renal cell carcinoma.

CASE REPORT

A 12-year-old boy presented to a local hospital with right loin pain of 7 months' duration, an abdominal mass of 4 months' duration, and gross hematuria of 2 months' duration. The performance status was graded as 3 according to World Health Organization criteria [8]. He was cachectic, and his abdomen was distended. A hard, fixed mass measuring 14 × 14 cm that crossed the midline was palpated in the right upper and lower quadrants. Dyspnea, tachypnea, and tachycardia were present. On auscultation, breath sounds were inaudible on the lower half of the right lung, with dullness on percussion.

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The erythrocyte sedimentation rate (77 mm/h) and the lactate dehydrogenase level (1,562 U/L) were found to be elevated. The complete blood count and other biochemistry were within normal limits. Macroscopic and microscopic hematuria was present, but the urinalysis was otherwise normal.

Computed tomography (CT) of the abdomen demonstrated a 12 × 12 × 20 cm mass in the upper pole of the right kidney. Bilateral coin lesions were seen both on chest radiography and CT of the thorax. The patient underwent an exploratory laparotomy. The mass was found to have infiltrated the liver and was rendered unresectable. Only a biopsy was performed. Histopathologic evaluation revealed an RCC (clear cell papillar adenocarcinoma). The patient was listed as stage IV-B according to the staging system of Robson et al. [9]. A chemotherapy regimen consisting of 5-fluorouracil (5-FU), 750 mg/m^2/24 h infusion for 5 days, and Interferon-α, 10 million U/d subcutaneously for 5 days, 1 hour before 5-FU administration. Interferon-α, 10 million U subcutaneously three times per week was administered between courses. After the first course, a decrease in pain was reported. There was no objective response with either physical examination or radiologic evaluation [CT]. One week after the second course, the patient's respiratory difficulty increased and he died with progressive disease.

DISCUSSION

Renal cell carcinoma is extremely rare in children. Data from the third National Cancer Survey indicate that renal malignancies rank sixth in frequency among all childhood cancers, with Wilms' tumor outnumbering RCC by a ratio of nearly 30 to 1 [1]. In our series of 339 childhood malignancies seen between September 1989 and December 1993, this is the only RCC case (0.3%) encountered; the ratio of Wilms' tumor cases have outnumbered RCC cases by a ratio of 22 to 1.

The clinical manifestations of childhood RCC appear identical to those seen in adults. The presence of the classic triad of flank pain, gross hematuria, and a palpable mass usually indicates a more advanced stage of the disease [2]. Wilms' tumor is often considered in the differential diagnosis, but the older age (more than 10 years) at presentation should alert the clinician to the possibility of RCC [2, 5]. Other space-occupying lesions of the pediatric kidney, including multilocular cysts, nephroblastomatosis, angiomyolipoma, congenital mesoblastic nephroma, benign stromal tumors, intrarenal neuroblastoma, renal teratoma, malignant rhabdoid tumor, clear cell sarcoma, lymphoma (especially Burkitt's), and

crenal sarcoma (eg, rhabdomyosarcoma, liposarcoma) should be considered in the differential diagnosis.

The overall prognosis for children with RCC appears to be similar to that in adults. Of the 37 children reported in two series, the long-term survival rates were 11 of 11 (100%) for stage I, 6 of 11 (66%) for stage II, 3 of 7 (43%) for stage III, and 1 of 8 (12%) for stage IV [2, 5]. Castellanos et al. [4] have reported actuarial survival rates of 60% at 2 years, 56% at 5 years, and 50% at 10 years in 84 children. Most recurrences and deaths occur within the first 2 years after diagnosis, but later recurrences are not infrequent [6].

The primary therapy of localized RCC is radical nephrectomy with resection of the kidney and tumor, the adrenal gland, the surrounding perinephric fat, Gerota's fascia, and the regional lymph nodes. The role of preoperative or postoperative radiotherapy is uncertain. No single agent or combination of agents has yet been proven to be of significant benefit. However, RCC may prove amenable to therapy with biologic response modifiers. Numerous trials have demonstrated a complete or partial response to interferon-α and to interleukin-2 with lymphokine-activated killer (LAK) cells [1]. In our patient, we used 5-FU and interferon-α, which is reported to be an effective regimen in attempting to achieve a response [10]. Unfortunately, there was no objective response.

The prognosis in advanced stages of RCC is poor. In the series reported by Lack et al. [2], all patients with stage IV disease have died. For patients with stage IV disease therapeutic strategies currently are inadequate, with no proven curative regimens.

In conclusion, although RCC is rare in childhood, it should be kept in mind in childhood renal malignancies of older age. Making the diagnosis in an earlier stage may result in a better prognosis.

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