The Turkish Journal of Pediatrics 31:71 - 77, 1989

RENAL LYMPHOMA*
An Unusual Presentation in a Child

*M. Tezer Kutluk MD**, Münevver Büyükpamukçu MD***
*Safiye Göğüş MD***, Faik Sarıalioğlu MD****, Okan Akhan MD*****
*Nesrin Beşbaş MD*****

Key words: non-Hodgkin's lymphoma, kidney, primary renal, childhood

Renal involvement is commonly seen in lymphoproliferative diseases, especially in non-Hodgkin's lymphoma. This involvement is usually associated with other findings such as lymphadenopathies, hepatosplenomegalia, bone marrow infiltration, and an abdominal or mediastinal mass. In other words, kidney infiltration in non-Hodgkin's lymphoma is a part of the disease[1-6]. Although lymphomatous renal parenchymal infiltration is frequently found at autopsy, it is rarely the cause of renal failure[7-9]. In this report a child with non-Hodgkin's lymphoma presented at the hospital with bilateral renal involvement as a primary localization.

Case Report

A four-year-old boy was admitted to Hacettepe University Children's Hospital with a two-month history of abdominal enlargement. Physical examination of the abdomen revealed a fixed, firm mass in the left lumbar region which was thought to be the left kidney. The liver was enlarged one and a half cm below the right costal margin. Other findings of the physical examination were normal.

Laboratory findings revealed a hemoglobin of 14 g/dl, and white blood cell count of 4600/mm^3 with a distribution of 68% neutrophils and 32% lymphocytes. An intravenous pyelogram showed bilateral enlargement of the kidneys and elongated renal infundibula (Fig. 1). Abdominal ultrasonography revealed bilateral, enlarged, lobulated kidneys with increased echogenicity and parenchymal thickness (right kidney 11.5 cm; left kidney 12 cm on the longitudinal axis). In addition to diffuse parenchymal infiltration there were several hypoechoic nodules representing nodular involvement (Fig. 2).

* From the Departments of Pediatrics and Radiology, Hacettepe University Faculty of Medicine, Ankara.

** Associate Professor of Pediatrics, and Director, Gülveren Health Center, Ankara.

*** Professor of Pediatrics, Hacettepe University Faculty of Medicine.

**** Associate Professor of Pediatrics, Hacettepe University Faculty of Medicine.

***** Assistant Professor of Radiology, Hacettepe University Faculty of Medicine.

Fig. 1: Intravenous pyelogram showing bilateral enlarged kidneys and infundibular elongation at diagnosis.

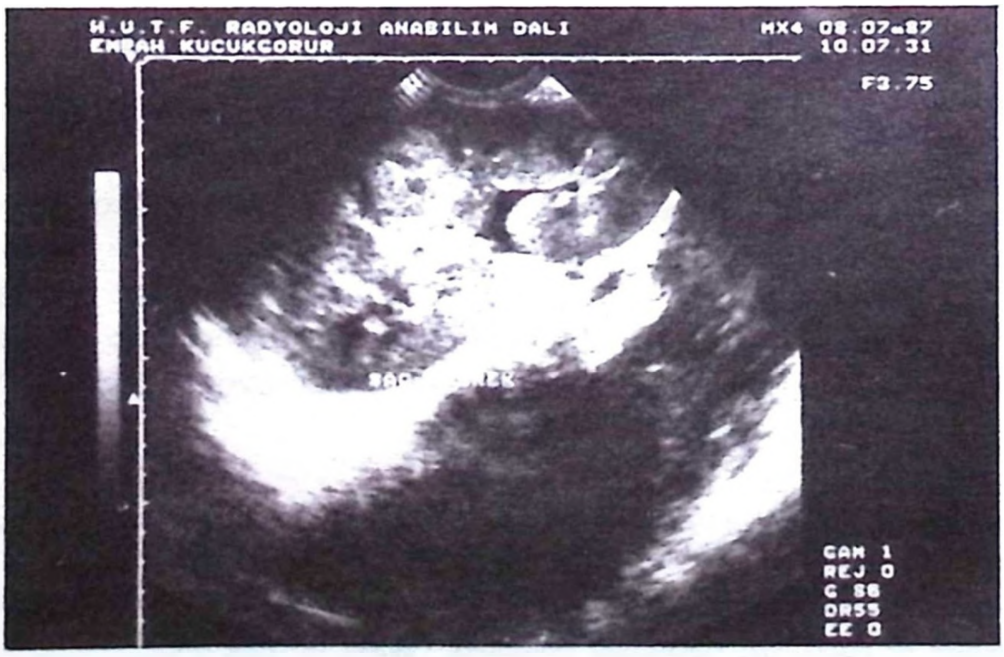

Fig. 2: Ultrasonography showing an enlarged kidney with parenchymal thickening on the right side. There are several hypoechoic nodules and diffuse infiltration.

BUN was 11 mg/dl, serum uric acid 6.9 mg/dl, and creatinine 0.5 mg/dl. Serum calcium, phosphorus, alkaline phosphatase, electrolyte and protein levels were within normal limits. Two-dimensional chest X-rays and the bone marrow smear were normal.

On the basis of clinical and laboratory findings, a malignant, infiltrative process non-Hodgkin's lymphoma was suspected. Due to the rare presentation and bilateral nature of the case, an exploratory laparotomy was performed which revealed a normal liver, spleen, and intraperitoneal and retroperitoneal organs; there was no lymphadenopathy. Both kidneys were enlarged and hard on palpation. A bilateral kidney biopsy was performed. The histological diagnosis was lymphoblastic non-Hodgkin's lymphoma involving both kidneys (Fig. 3).

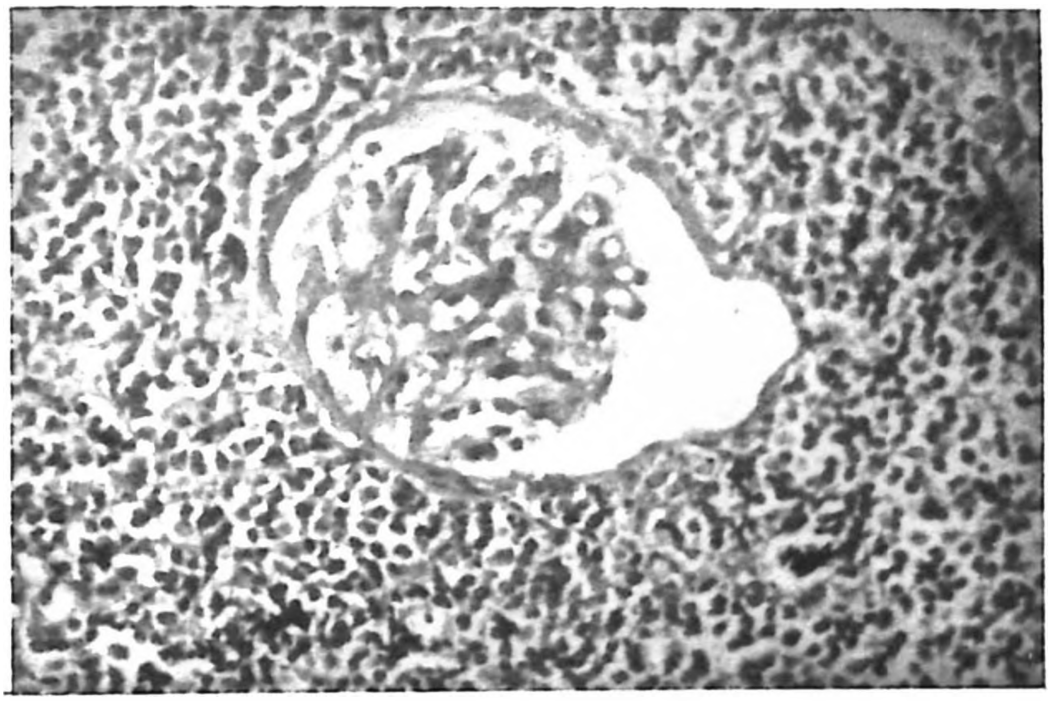

Fig. 3: Diffuse lymphoblastic infiltration in the left kidney (HE × 400).

Treatment with a modified LSA_2-L_2 chemotherapy regimen[10] was intiated and the patient was in complete remission after the induction phase of therapy. He has been in continuous complete remission for 14 months. His latest abdominal ultrasonography revealed normal size kidneys and also other organs which were normal (Fig. 4). An intravenous pyelography done four months later was interpreted as normal (Fig. 5).

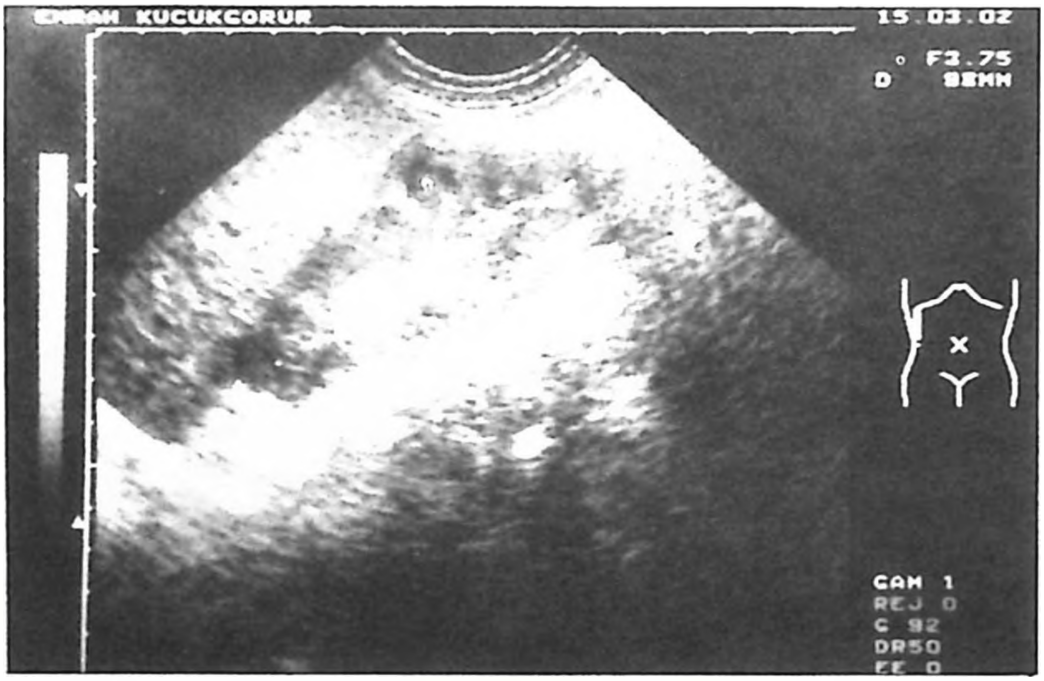

Fig. 4: Ultrasonography showing normal kidney after therapy.

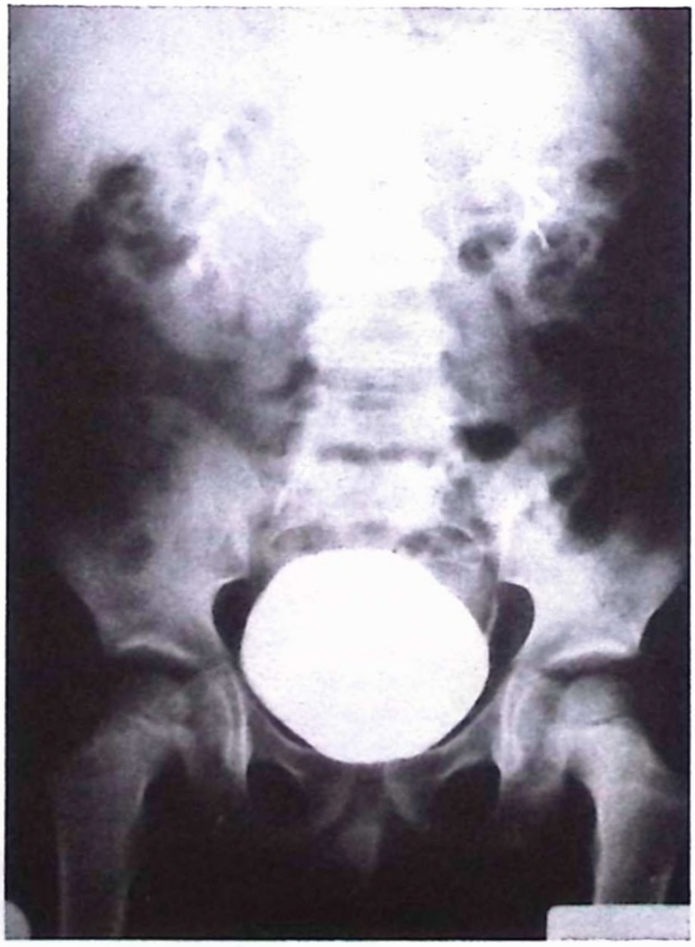

Fig. 5: Normal appearance of the kidneys seen on the intravenous pyelogram after therapy.

Discussion

Lymphomas rank third after lung and breast cancer in frequency of metastasis to the kidney[2]. Richmond et al[3] have reported that kidneys are infiltrated more frequently than any other organ or tissue. In his autopsy series of 696 patients with lymphoma, the incidence of renal parenchymal involvement was 34 percent. In the same study only 14 percent of 142 patients whose antemortem clinical data was available had renal involvement recognized prior to death. In only 0.5 percent of these 696 patients could the cause of death be attributed to lymphomatous infiltration of the kidneys.

Ehrlich and Kuhn[11] reported 41 percent of renal involvement in 32 non-Hodgkin's lymphoma cases and recommended that abdominal CT be done in all children with non-Hodgkin's lymphoma. They also emphasized that renal involvement was more common than previously reported.

Although secondary renal involvement in non-Hodgkin's lymphomas is not uncommon, primary renal lymphoma is rarely reported in children[1-7]. The clinical manifestations are not overt when the disease is limited to the kidneys in the early period of the illness. Most of the reported cases have shown secondary involvement of the kidney as a part of the disease[3,4,7,8].

Our patient was an unusual case in that he exhibited bilateral renal involvement without any other findings of lymphomatous involvement. Jaffe and Tefft[2] reported a case which was first diagnosed radiologically as bilateral Wilms' tumor but whose tonsillectomy material and biochemistry showed lymphosarcoma and renal failure.

Glicklich et al[7] reported three cases and found 14 similar cases after a review of the literature. Four of them were pediatric patients. Only one had no primary involvement other than kidney. This patient also displayed renal failure. Recently Falconieri and Melato[9] reported a case of a 47-year-old woman with malignant lymphoma that presented as an isolated renal mass.

Occult lymphomas can be encountered clinically by manifestation of renal failure at the inital presentation. Diagnosis must be established by renal biopsy[8]. The combination of sonographic and computer tomographic findings will support the diagnosis of lymphoma which can be proven by renal biopsy. These noninvasive techniques should be considered prior to renal biopsy[11-14].

In spite of the high percentage of renal involvement in non-Hodgkin's lymphoma, urinary findings are uncommon and death attributed only to renal involvement is rare. Randolph et al[8] reported a case with lymphomatous infiltration of the kidneys presenting as uremia of unknown cause. Interstitial lymphomatous infiltration of the kidneys by tumor cells can cause atrophy in nephron units. In uremic patients the other causes of uremia i.e., hypercalcemia, hyperuricemia, ureteral obstruc-

tion, amyloidosis, and immunologically mediated necrosis should be considered and then ruled out[7,8]. Renal functions were normal in our case. Many patients with normal renal function have been reported[8]; this is true especially for early disease, but most of the advanced cases show a disturbance in renal function.

Systemic chemotherapy or limited radiotherapy is effective in controlling renal involvement. Treatment sensitivity of the disease confirms the diagnosis[7,8].

In our case there were no findings of renal failure, and systemic chemotherapy was administered promptly. The patient responded to the treatment and the enlarged kidneys regressed in size within two months. The findings of an exploratory laparotomy and an extensive staging procedure showed no other primary site. Although the renal involvement was bilateral, we believe that our patient was an interesting case of primary renal lymphoma.

Summary

An unusual case of non-Hodgkin's lymphoma with primary renal involvement in a four-year-old boy is presented. The diagnosis was established by renal biopsy. The findings of an exploratory laparotomy and an extensive staging procedure showed no other primary site. The patient has been followed up for 14 months and no evidence of recurrence has been observed.

REFERENCES

1. Davis FM, Olivetti RG. Primary lymphosarcomatosis of kidneys, adrenal glands and perirenal adipose tissue. *J Urol* 66:106, 1951.
2. Jaffe N, Tefft M. Unsuspected lymphosarcoma of the kidneys diagnosed as bilateral Wilms' tumor. *J Urol* 110:593, 1973.
3. Richmond J, Sherman RS, Diamond HD, Craver LF. Renal lesions associated with malignant lymphomas. *Am J Med* 34:182, 1962.
4. Iskandar SS, Jennette JC, Weiss LS. Primary cerebral lymphoma with glomerular renal involvement. *Arch Pathol Lab Med* 109:524, 1985.
5. Shah I, Parekh N, Nayak PK, Taher S. Renal oncocytoma associated with diffuse lymphoma. *Urology* 22:314, 1983.
6. Hahn FJ, Peterson N. Renal lymphoma simulating adult polycystic disease. *Radiology* 122 (2 Suppl):655, 1977.
7. Glicklich D, Sung MW, Frey M. Renal failure due to lymphomatous infiltration of the kidneys. Report of three new cases and a review of the literature. *Cancer* 58:748, 1986.
8. Randolph VL, Hall W, Bramson W. Renal failure due to lymphomatous infiltration of the kidneys. *Cancer* 52:1120, 1983.
9. Falconieri G, Melato M. Malignant lymphoma presenting as an isolated renal mass. *Histopathology* 12:217, 1988.
10. Büyükpamukçu M, Sarialioğlu F, Akyüz C, Cevik N. Combined chemotherapy in 76 children with non-Hodgkin's lymphoma excluding Burkitt's lymphoma. *Br J Cancer* 56:625, 1987.

11. Ehrlich AD, Kuhn JP. Renal involvement in childhood lymphoma: the role of CT. *AJR* 143:692, 1984.
12. Bosniak MA, Horii SC, Megibow AJ, et al. Correlations of CT and ultrasound in the evaluation of lymphoma of the kidneys. *AJR* 139:207, 1982.
13. Garvie NW, Chu JM, Nuttall J, McCready VR. Renal involvement with non-Hodgkin's lymphoma: investigation of a recent case. *Br J Radiol* 51:547, 1978.
14. Siegel SW, Risius B, Tubbs R, et al. Bilateral solid renal masses in a young man [clinical conference]. *J Urol* 135:327, 1986.