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Case Reports

PRIMARY RENAL BURKITT'S LYMPHOMA

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ABSTRACT. Primary renal Burkitt's lymphoma is a rare disease. A two-and-a-half year old boy presented with poor appetite, severe malaise and constipation. Examination revealed bilateral large renal masses and also right facial palsy and right sided convergent squint. The rarity of this condition resulted in a delay of 13 days in arriving to a final diagnosis. The delay had most likely contributed to the lethal outcome despite an aggressive chemotherapy.

INTRODUCTION

Burkitt's lymphoma is a specific tumor of B-lymphocytes with endemic prevalence in tropical Africa and sporadic prevalence worldwide (1). The non-endemic variety shows a higher incidence of abdominal and peripheral nodal involvement (2,3), so that the most frequent clinical presentations are intestinal obstruction or abdominal masses (1). Although renal involvement is commonly found at autopsy as part of the disseminated disease, prominent renal involvement is very unusual at clinical presentation (4,5). The purpose of this paper is to report a case of primary bilateral Burkitt's lymphoma of the kidneys with a review of pertinent literature.

CASE REPORT

A previously healthy two-and-a-half year old boy was admitted to the pediatric ward on 10 April 1989 for poor appetite, severe malaise and constipation for one week prior to presentation. On examination the boy was found to be afebrile and alert. Blood pressure was 118/80 mmHg and Pulse was 120/min. He had bilateral large kidney masses. There was right facial palsy and right-sided convergent squint.

Results of blood counts, sedimentation rate, blood urea, creatinine, creatinine clearance, serum uric and vanillylmandelic acid levels as well as urine analysis and culture were normal. Chest x-ray showed no evidence of infiltration or secondaries. Cardiac shadow was normal. Ultrasonography showed bilaterally enlarged kidneys with some cystic changes in the renal parenchyma. In addition, a cystic lesion in the right lobe of the liver measuring 1.8 x 1.7 cm was identified. Intravenous pyelogram revealed distortion and displacement of the calyces of both kidneys. Abdominal CT-scan disclosed an enlarged liver with a small cystic lesion, normal spleen and markedly enlarged kidneys, the right being larger than the left. The inferior vena cava and aorta seemed to be involved (Figure 1). Chest and brain CT-scans were normal. In view of the facial nerve palsy cerebrospinal fluid (CSF) was examined but it did not reveal any abnormality. A provisional diagnosis of bilateral Wilms' tumor was made at this stage.

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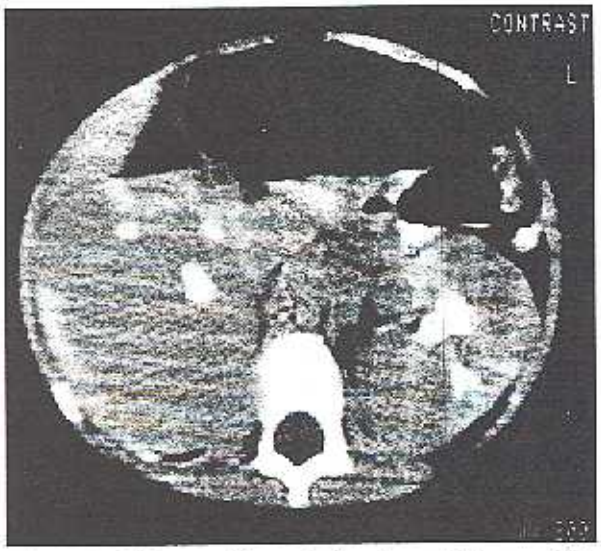

Figure 1. CT-scan with markedly enlarged kidneys; right more than the left. Prior to chemotherapy.

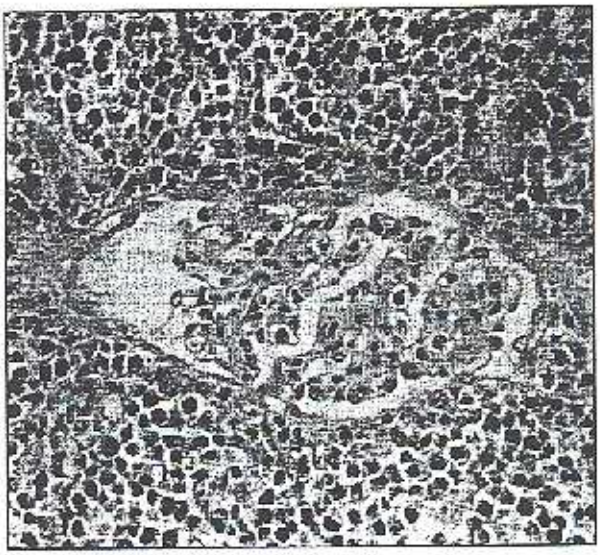

Figure 3. Renal parenchyma is diffusely infiltrated by lymphomatous tissue. A starry sky appearance is evident.

Exploratory laparotomy was performed; kidneys were diffusely enlarged displacing liver, stomach and intestines (Figure 2). Multiple needle biopsies were taken from both kidneys and from the suspected liver lesion. A right perirenal lymph node and three pre-aortic lymph nodes were excised. Frozen section biopsy report was inconclusive. Postoperative chemotherapy was given with daily intravenous (IV) actinomycin D for 5 days and I.V. vincristine given as a single bolus dose. Histopathology finally established the diagnosis of Burkitt's lymphoma infiltrating the renal parenchyma with starry sky appearance, decreased glomeruli and atrophic tubules (Figure 3). Liver biopsy was normal and the lymph nodes showed only reactive hyperplasia. Following this report the child received cyclophosphamide, vincristine and prednisone (CVP) intravenously and methotrexate, Ara-c and hydrocortisone (MAH) intrathecally. Repeated CT-scan showed marked reduction in the size of both kidney masses (Figure 4). One month later, a second cycle of CVP and MAH was administered and the patient was discharged in a satisfactory condition. Subsequent chemotherapy courses were given at monthly intervals. Six months later, the patient was still found to have the right facial palsy although the CSF and brain CT scan were normal. However, the EEG was suggestive of a right hemispheric lesion.

Chemotherapy was continued at monthly intervals. At the end of the seventh month since the start of chemotherapy, the patient was readmitted in a marasmic condition. He had developed a painful swelling of the right

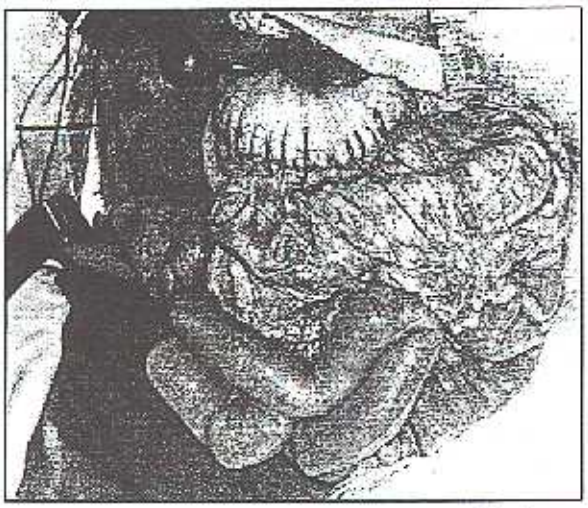

Figure 2. Intra-operative photograph showing the diffusely enlarged kidneys in the subhepatic space (arrow) displacing liver, stomach and intestine.

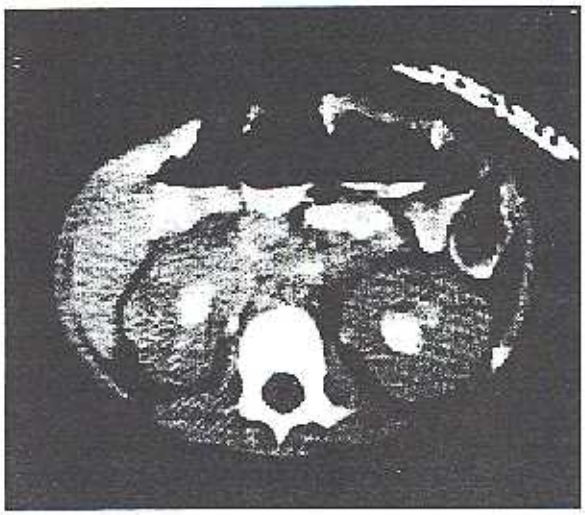

Figure 4. CT-scan following the first cycle of chemotherapy with dramatic reduction in renal size.

jaw, tender cystic swelling of the upper gum and proptosis of the right eye. Lungs, heart and vital signs were normal. The total leukocyte count was 4.2 x 10^9/L with polymorphs 22%, band forms 3%, lymphocytes 36%, monocytes 6%, eosinophils 1%, basophils 4% and atypical lymphocytes 9%, metamyelocytes 3%, myelocytes 6%, promyelocytes 5%, and lymphoblast 5%. CT-scan of brain done at this time was suggestive of probable infiltration. Despite intensive chemotherapy the patient developed cranial dissemination as evidenced by convulsions, facial twitching and syndrome of inappropriate ADH secretion (serum Na^+ 118 mmol/l, serum osmolarity 240 mOsmol/L; urine Na^+ 108 mmol/l, urine osmolarity 305 mOsmol/L). He died one month after the admission to the hospital.

DISCUSSION

Non-Hodgkin's lymphoma presenting as kidney masses is extremely rare. In a series of 423 patients with Non-Hodgkin's lymphoma, only two cases presented as renal masses (6). A higher prevalence has been suggested in two recent series (7,8). In most cases, renal involvement occurs late as part of a disseminated disease process (9). The occurence of primary renal lymphoma without detectable disease in other organs is exceptional (10,11). Emmet and Witten believe that renal lymphoma represents always a secondary involvement (9). However, clinical and intraoperative findings in our case, point to the kidneys as the most probable primary site. Commonly, renal lymphoma is of the nodular type. Other patterns include single or multiple large masses and diffuse infiltration of the kidneys (12). The latter pattern was seen in cases of sporadic Burkitt's lymphoma, while discrete renal masses were encountered in the African variety (13). The diffuse renal infiltration is found to be associated with decreased renal function, that improves on chemotherapy or irradiation (13,14). In our case, despite markedly enlarged kidneys with diffuse infiltration, decreased glomeruli and atrophic tubules, renal functions were found to be normal. Even the acute tumor-lysis syndrome, anticipated in such cases (5), was not observed. The recommended surgical reduction of tumor bulk prior to chemotherapy (1,16,17) was not done because of the nature of renal involvement.

Since Burkitt's lymphoma is the fastest growing human tumor, with a doubling time of 24 hours, the diagnosis must be established and definitive treatment started as quickly as possible, to avert potentially lethal complications (1, 15, 18). A delay beyond 48 hours may have a significantly negative effect on the outcome (15). The magnitude of this effect cannot be determined in this case. Prognosis is also dependent on tumor burden on initial presentation, clinical stage, age and attainment of complete regression of all tumors (1,15,16). Involvement of the central nervous system is an uncommon presenting feature but it is a frequent manifestation of relapse after regression has been achieved with chemotherapy (19,20). Bone infiltration is seen in 10% of cases (7). In this case the facial nerve weakness may possibly be due to CNS involvement undetected by CT-scan at the time of presentation. The squint may be explained as due to intracranial involvement or due to the retrobulbar involvement which became florid during the last admission.

Most important tools to establish the diagnosis of renal lymphoma are ultrasound, CT-scan and gallium 67-scintigram. The latter is the most sensitive test of all. Serum lactate dehydrogenase, uric acid and phosphate are sensitive indicators of tumor regression and relapse. The examination of CSF for malignant cells and bilateral iliac-crest marrow biopsies are mandatory (8,15,16). The therapy of choice is an aggressive combination chemotherapy consisting of cyclophospahmide, vincristine, doxorubicin and prednisone with intermittent intrathecal cycles of high-dose methotrexate and Ara-C (1,21,23). This regimen is considered most promising with a reported overall survival of 70% (24). However, tumors that relapse during chemotherapy or soon after cessation of chemotherapy behave as though a drug-resistant tumor cell clone has emerged (12,5). Drugs then have little effect on a rapid downhill course as illustrated in this case.

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