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Case report

A rare case of bilateral primary renal Burkitt lymphoma presenting with acute renal failure ☆,☆☆

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ABSTRACT

Acute renal failure due to primary renal Burkitt lymphoma in children is extremely rare. We report a case with acute secondary renal failure in a 4-year-old boy who presented with abdominal pain, anorexia, and vomiting. Abdominal computed tomography scans showed bilateral nephromegaly with multiple hypoenhancing regions. Renal biopsy confirmed Burkitt lymphoma. There was no lymphadenopathy or evidence of other solid organ involvement. The patient was responsive to treatment using the EPOCH-R protocol (etoposide, prednisone, vincristine, cyclophosphamide , doxorubicin, and rituximab). Here, we describe the clinical and imaging features associated with this rare entity.

Introduction

Renal involvement in lymphoma is typically a secondary outcome, and the incidence ranges from 30%-60% [1]. Primary renal lymphoma (PRL) is defined as lymphoma involving the kidney without evidence of disease elsewhere and is extremely rare, presenting in <1% of all extranodal lymphomas [2]. Primary renal Burkitt lymphoma is even rarer, with only several cases reported in the literature [3]. Renal lymphoma is

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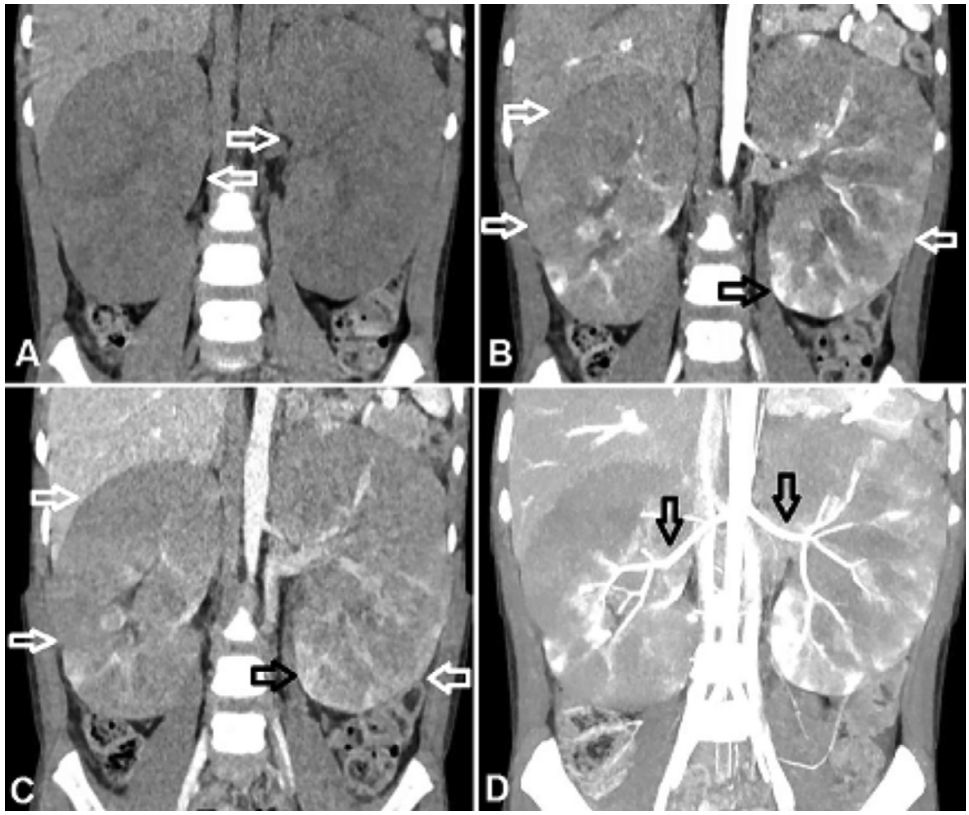

Fig. 1 – Abdominal computed tomography (CT) scans before treatment. (A) Noncontrast CT scans showed bilateral, symmetrical enlargement of both kidneys (arrows). The arterial phase (B) and venous phase (C) showed multiple hypoenhancing areas throughout both kidneys (white arrows), although some areas of the renal parenchyma still enhanced normally (black arrows). (D) Maximum intensity projection (MIP) showed that neither renal artery was occluded (black arrows).

typically asymptomatic and commonly detected only when it reaches the late stage [4]. Acute kidney injury is a life-threatening complication of renal lymphoma and occurs in 0.9% to 23% of patients diagnosed with renal lymphoma [4]. In this report, we aimed to describe a case of acute kidney injury induced by diffuse bilateral infiltration of Burkitt lymphoma in a child.

Case report

A 4-year-old patient was referred to the hospital presenting with oliguria, weakness, anorexia, vomiting, and abdominal pain for 1 week. No significant background medical history was noted. A physical examination revealed mild edema and hypertension (125/80 mm Hg). The results of laboratory studies showed normal complete blood count; serum creatinine concentration: 130 μmol/L (normal range 27-60 μmol/L); blood urea: 11 mmol/L (normal range 2.5-6.4 mmol/L); serum uric acid: 6.5 mg/dL (normal range 1.7-5.9 mg/dL); serum sodium: 140 mmol/L (normal range 134-143 mmol/L); serum potassium: 4.5 mmol/L (normal range 3.5-5 mmol/L); lactate dehydrogenase: 540 IU/L (normal range 150-500 IU/L); and normal serum aspartate aminotransferase and alanine aminotransferase levels. The urinalysis test showed no leukocytes, blood cells, or proteins. An abdominal ultrasound manifested diffuse enlargement of both kidneys, which were hypoechoic associated with loss of corticomedullary differentiation. An abdominal computed tomography (CT) scan showed bilateral nephromegaly, featuring multiple hypoenhancing areas with poorly defined boundaries relative to the normal renal parenchyma and no hydronephrosis (Fig. 1). No lesions were identified in any other organs, and no enlarged abdominal lymph nodes were detected. The differential diagnoses included acute kidney injury (AKI) due to tubular necrosis, pyelonephritis, or lymphoma. The combination of clinical symptoms, laboratory tests, and imaging features, however, were most suggestive of renal lymphoma. A renal biopsy was performed, and histopathology confirmed Burkitt lymphoma. A physical examination showed no large peripheral lymph nodes, and chest and brain CT scans were normal. Therefore, the patient was diagnosed primary renal Burkitt lymphoma with stage II according to the Ann Arbor staging system of American Joint Committee on Cancer (7th edition) [5]. The patient was treated with 3 cycles of the EPOCH-R (etoposide, prednisone, vincristine, cyclophosphamide , doxorubicin, and rituximab) protocol. After treatment, both kid-

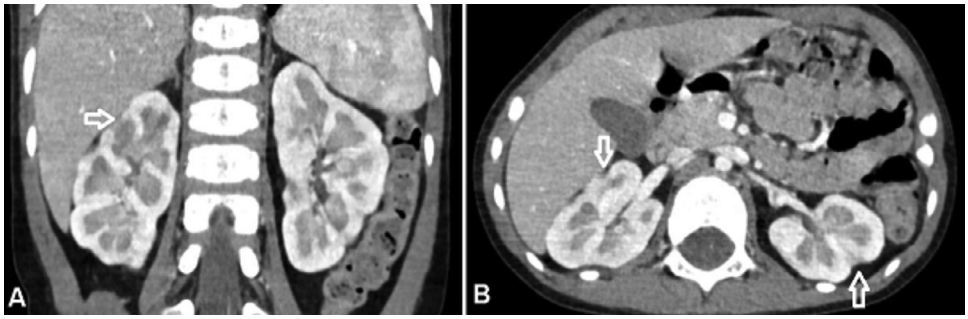

Fig. 2 – Abdominal computed tomography scans after 3 cycles of chemotherapy treatment. (A) Coronal and (B) axial images showed that the kidneys returned to normal size, the normal renal parenchyma enhanced strongly again, and the lymphoma-infiltrated areas became scars (arrows).

neys returned to normal size, and the lesions decreased in size and became renal cortical scars (Fig. 2).

Discussion

We present a case of AKI secondary to primary renal Burkitt's lymphoma. Literature searches reported by Castellano et al. [4], Haar et al. [6], and Agarwal et al. [3] revealed only a few case reports of this entity. Burkitt lymphoma appears primarily in children, with a male predominance [7].

AKI in PRL can occur due to various causes, such as tumor lysis syndrome, increased intrarenal pressure due to lymphoma cell proliferation, and urinary obstruction [4,6]. Renal obstruction may occur due to the tumor invasion of the ureters or renal pelvis or due to glomeruli obstruction [4,6]. Haar et al. [6] reported a pediatric patient diagnosed with AKI due to PRL without hematuria and proteinuria and suggested that AKI may be caused by tubular compression secondary to the interstitial proliferation of tumor cells. Similar to the patient described in the present study, the patient described by Haar et al. had symptoms of AKI with no evidence of hematuria or proteinuria, and the lesions in both kidneys appeared as diffuse infiltrations without a mass that invaded the urinary excretory system. Therefore, we hypothesize that the cause of AKI in the present patient might be increased intrarenal pressure.

Few studies have evaluated the imaging features of PRL due to the rarity of this disease. PRL may present unilaterally or bilaterally, as solitary mass or multiple masses, or as nephromegaly with ill-defined infiltrative lesions. Invasion of the inferior vena cava is uncommon. [2,8]. On ultrasound, PRL with masses appear hypoechoic and homogeneous, but PRL with diffuse infiltration PRL often presents as heterogeneous [9]. On CT scans, the lesions present as low density and hypoenhancing [9]. On magnetic resonance imaging, PRL appears isointense on T1-weighted images, slightly hypointense on T2-weighted images, and restricted diffusion [10]. However, the final diagnosis is always confirmed by histopathology. Some differential diagnoses include acute renal tubular necrosis, acute papillary necrosis, and pyelonephritis. Based only on imaging, PRL can be difficult to differentiate from other diagnoses, especially diffuse infiltration PRL. Clinical symptoms of AKI and laboratory test results such as creatinine blood test, urinalysis can be useful for the orientation of the underlying cause. The patient in the present case did not present with sepsis symptoms such as fever or low temperature, increased heart rate, breathless, confusion; or evidence of renal tubular epithelial cells in the urine microscopy test. Therefore, a diagnosis of renal lymphoma was suspected, which was confirmed by renal biopsy.

Some of the laboratory results that have been associated with poor prognosis in Burkitt lymphoma include elevated lactate dehydrogenase levels, creatinine $>$ upper limit of normal, and uric acid $\geq$ 7 mg/dL [11,12]. Intensive multiagent chemotherapy applied to Burkitt lymphoma is able to improve renal function and prognosis. Chen et al. [13] reported that the 1-year and 5-year relative survival rates of PRL patients with the stages ranges from I to IV were 78% and 64%, respectively. However, there is a lack of treatment information on chemotherapy in the study of Chen et al. [13]. The present patient did not present with any factors associated with poor prognosis and was responsive to treatment, which successfully reduced the lesions significantly.

Conclusion

PRL presenting with AKI is rare, and the imaging features are difficult to differentiate from other kidney diseases, resulting in PRL being easily misdiagnosed. PRL should be included in the differential diagnosis when patients present with bilateral nephromegaly on imaging. Early diagnosis and treatment result in a good prognosis and improved survival rates.

Author contributions

Tran PN, Truong QD, and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the contents.

Patient consent

Informed consent for patient information to be published in this article was obtained.

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