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Case Report

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Primary pediatric stage III renal diffuse large B-cell lymphoma

Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

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Summary

Background: Although secondary renal involvement of non-Hodgkin lymphoma is frequently encountered, primary renal lymphoma is quite rare. We present a pediatric case of primary renal diffuse large B-cell lymphoma.

Case Report: A 12-year-old girl presenting with gross hematuria was referred to our hospital. Abdominal ultrasonography and imaging revealed a mass lesion in the superior pole of the right kidney. Serum creatinine and blood urea nitrogen levels were within normal ranges. Preoperative assessment of the mass indicated unspecified renal tumor. Right nephrectomy was performed and pathological examination showed diffuse large B-cell lymphoma. Postoperative fluorodeoxyglucose-positron emission tomography/computed tomography showed a small high-uptake lesion in the thyroid gland and aspiration cytology of the thyroid tumor demonstrated involvement of lymphoma, so stage III tumor diagnosed. After one course of chemotherapy, the patient achieved complete remission. She remains alive without disease, 3 years after completing a total of six courses of chemotherapy.

Conclusions: Primary renal lymphoma is a very rare entity and preoperative diagnosis may be difficult. However, this entity is often reported to show clinically aggressive characteristics and therefore should be considered among the differential diagnoses for unusual renal tumors in pediatric patients.

key words: **renal tumor • DLBCL-lymphoma • gross hematuria**

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Background

Although secondary renal involvement of non-Hodgkin lymphoma (NHL) is not uncommon among children, primary renal lymphoma is quite rare [1].

We present a case of primary renal lymphoma diagnosed as diffuse large B-cell lymphoma (DLBCL) following nephrectomy after discovery of a renal mass during examination for gross hematuria.

Case Report

A 12-year-old girl visited a local doctor complaining of gross hematuria. Her general health was stable with no fevers or symptoms of bladder irritation, such as painful or frequent urination. No masses could be felt in the abdomen on palpitation, but ultrasonography revealed a mass in the right kidney (Figure 1A), and the patient was referred to our hospital for further evaluation. Aside from uric blood, urinalysis did not reveal any abnormalities. Blood tests showed serum creatinine and blood urea nitrogen levels within normal ranges, with no sign of renal dysfunction. Soluble interleukin 2

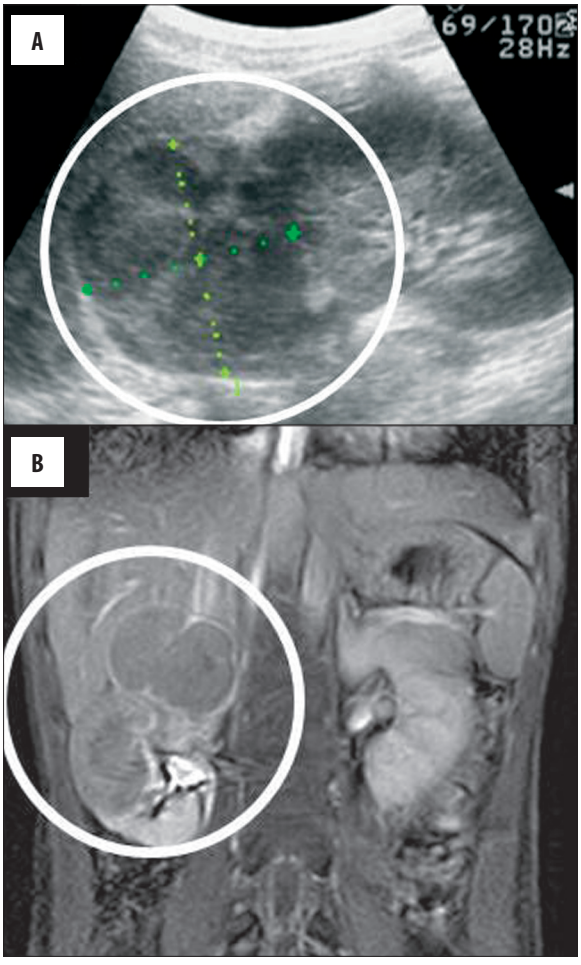

Figure 1. Imaging studies on admission. (**A**) Abdominal ultrasonography shows a mass lesion in the superior pole of the right kidney (white circle). (**B**) Abdominal magnetic resonance imaging shows a marginally enhanced tumor invading into the adrenal gland (white circle). The left kidney is normal.

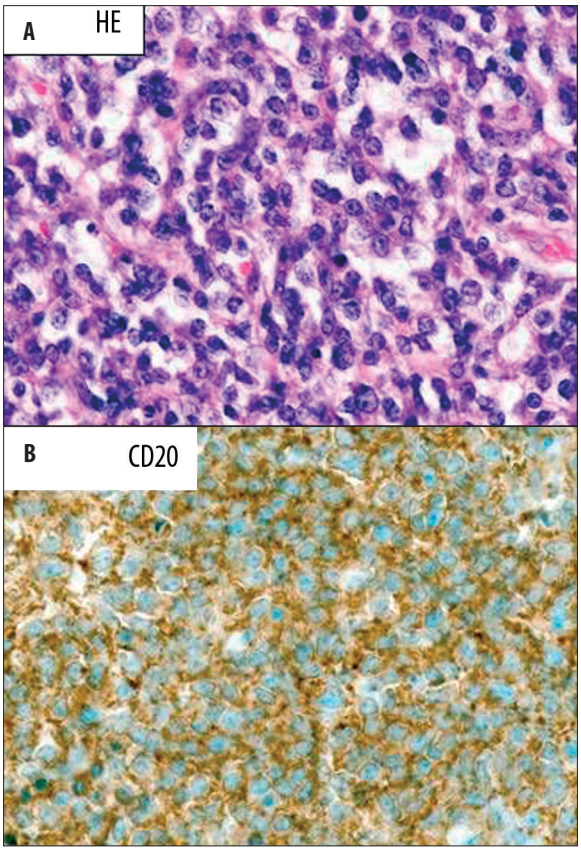

Figure 2. Histopathological study. (**A**) Tumor cells are round, with a high N/C ratio with HE staining. (**B**) Immunohistochemical staining shows diffuse positive results for CD20.

receptor, neuron-specific enolase, and other tumor markers were all negative. Abdominal magnetic resonance imaging (MRI) revealed a marginally enhanced 12-cm tumor-like lesion in the superior pole of the right kidney that appeared to be invading the adrenal gland (Figure 1B). No abnormalities were seen in the left kidney or any other parts of the abdomen, and chest computed tomography (CT) did not reveal any metastasis to the lung field or mediastinum. The mass was thought to be a localized tumor in the right kidney and right nephrectomy was therefore performed. Histopathological examination showed diffuse proliferation of large lymphoid cells, and immunostaining showed positive results for CD20, bcl-2 and bcl-6 and negative results for CD3, CD5 and CD10 (Figure 2A, 2B). We therefore diagnosed DLBCL. Positron emission tomography-computed tomography (PET/CT) was performed to examine the entire body, revealing an abnormal accumulation in the right lobe of the thyroid gland (Figure 3A, 3B). CT of the neck revealed a 9-mm low-density area in the same region (Figure 3C). Aspiration cytology under ultrasonographic guidance revealed an aggregation of immature atypical lymphocytes (Figure 3D) that was thought to represent invasion of the DLBCL. The disease was therefore diagnosed as stage III, and the patient was administered intensive chemotherapy comprising vincristine, dexamethasone, cyclophosphamide, methotrexate, cytarabine, and other drugs. The thyroid lesion decreased in size immediately after starting treatment. The patient received a total of 6 courses of chemotherapy and completed treatment without any serious complications for the solitary remaining kidney. As of the time of writing,

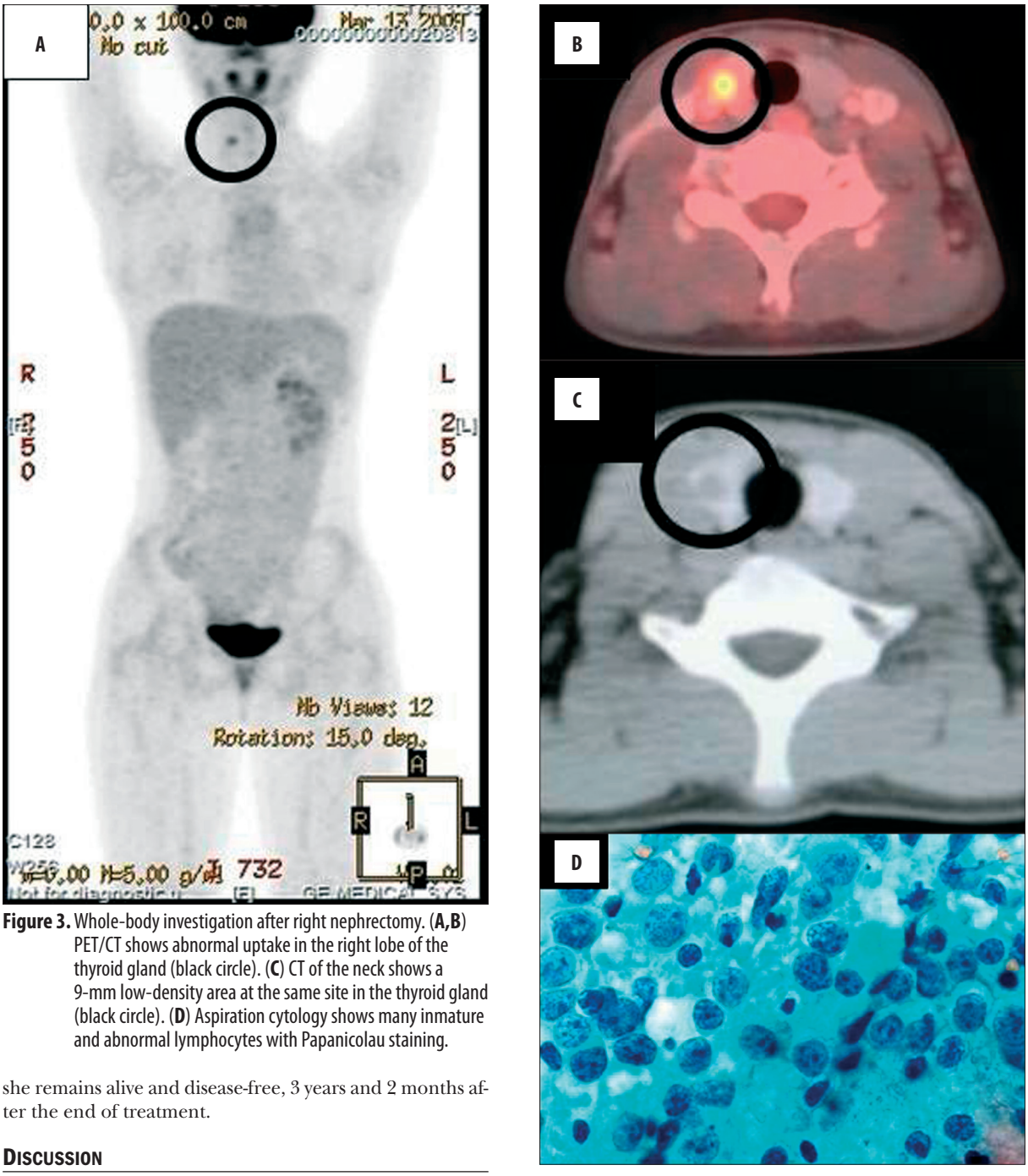

Figure 3. Whole-body investigation after right nephrectomy. (**A,B**) PET/CT shows abnormal uptake in the right lobe of the thyroid gland (black circle). (**C**) CT of the neck shows a 9-mm low-density area at the same site in the thyroid gland (black circle). (**D**) Aspiration cytology shows many inmature and abnormal lymphocytes with Papanicolau staining.

she remains alive and disease-free, 3 years and 2 months after the end of treatment.

Discussion

Tumors are rarely the cause of gross hematuria in children. According to a 10-year review of 342 patients ≤20 years old examining causes of gross hematuria, the cause was frequently unknown (34%), a urinary infection (14%) or trauma (14%). Only 4 patients (1%) had a tumor, involving bladder cancer in 3 and Wilms' tumor in 1 [2]. Regardless, abdominal ultrasonography should be actively pursued in children presenting with gross hematuria, as blood in the urine is a common first symptom of renal tumor [3,4].

Results for lymphoma on abdominal CT, MRI and other imaging modalities are often similar to those of localized renal cell cancer and Wilms' tumors, making differentiation difficult [5]. In the present case as well, as we did not identify the thyroid lesion prior to surgery and because the right kidney tumor was encapsulated, we suspected renal cell cancer or Wilms' tumor and proceeded with right nephrectomy, without suspecting lymphoma. We confirmed a diagnosis of DLBCL from pathological examinations after the surgery, and then identified a second lesion in the right lobe of the thyroid gland by PET/CT. The disease was thus diagnosed as stage III.

Primary renal NHL is extremely rare [1]. To judge a tumor as originating in the kidney, the lesion must be localized to the kidney or initial symptoms must be urinary tract symptoms [5]. In the present case, a lymphoma lesion was discovered in the thyroid gland in addition to the right kidney,

and sizes of these lesions varied substantially, with the right kidney tumor being 12 cm in diameter and the thyroid gland tumor only 9 mm. Moreover, the initial symptom was gross hematuria. We therefore judged the tumor in the right kidney as the primary tumor.

Most cases of primary renal NHL reportedly progress rapidly and display a poor prognosis [6]. In the present case, we performed intensive chemotherapy as treatment for stage III DLBCL in the solitary remaining kidney after nephrectomy of the right kidney. Although the patient completed treatment without any severe complications and remains alive and disease-free as of 3 years following the end of treatment, careful long-term follow-up will be required to confirm the absence of recurrence or renal dysfunction.

Conclusions

Primary renal lymphoma is very rarely observed as a pediatric NHL, and proper diagnosis upon discovery of a renal tumor may therefore be difficult. Nevertheless, in many cases of primary renal lymphoma, the disease progresses rapidly, renal function declines quickly, and general health deteriorates. Consideration of the possibility of malignant lymphoma as a differential diagnosis for renal tumor is therefore warranted.

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