

Review Article

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Renal Burkitt Lymphoma: An Unusual Extra-Nodal Condition Diagnosed in a Child in Niger

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ABSTRACT

Renal Burkitt lymphoma is an aggressive malignant extra-ganglionic blood disease. It is an unusual event in children. A case is reported below. The patient was 8 years old; she was presenting with acute renal failure and edematous syndrome; proteinuria was mild. There was no organ enlargement nor adenopathy. After a while the diagnosis of Burkitt lymphoma was made after an anatomic-pathological examination of a kidney biopsy taken as part of an etiological assessment of this acute renal failure (urea=35 mmol/L, serum creatinine= 880 µmol/L) with an impaired general condition (WHO 3). The histological characteristics strongly suggested a lymphomatous process. An immunohistochemical examination confirmed renal Burkitt lymphoma. Unfortunately the patient died before chemotherapy was attempted.

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Introduction

Burkitt lymphoma is an aggressive malignant blood disease. It is a non-Hodgkin malignant lymphoma (NHML) and has very rapid monoclonal proliferation of lymphoid cells (Burkitt cells) from the B line. Described in 1958 by an English surgeon Denis Burkitt in Kampala (Uganda, Africa), it represents the first cancer identified to be induced by the Epstein Barr virus (EBV) [1, 2]. Three forms have been described [3].

- An endemic (African) form, where the EBV genome is found in the clonal state in the tumor cell in 98% of cases, often with maxillofacial involvement.
- A sporadic (non-African) form with abdominal predominance.
- A form related to immune deficiency.

Extra-nodal localizations, such as cerebrospinal, ovarian, testicular, have been described [4]. However, renal Burkitt lymphoma is an unusual event in children [5]. Its diagnosis is based on a kidney biopsy and pathophysiological examination. It is confirmed by

immunohistochemistry. This extremely rare tumor is difficult to diagnose. Chemotherapy may improve survival of these patients. This case is reported because of its peculiarity.

Observation

A girl aged 8 years, without a specific medical history was seen in the Nephrology Department of the Zinder National Hospital (Niger) for severely impaired renal function (urea=29.8 mmol/L, serum creatinine = 567 µmol/L) discovered in the context of edematous syndrome. The patient reported edema of the lower limbs and puffiness of the face that had begun 3 weeks previously these symptoms increased to edematous syndrome, followed by anterior cervical swelling and fever. From an initial consultation in a village health center, the patient was referred to the National Hospital of Zinder with nephrotic syndrome.

At the Pediatric Department of the hospital, explorations showed impaired renal function and necessitated transfer to the nephrology department. A clinical examination ascertained a WHO Performance status of 3.

The patient had edematous syndrome, dyspnea, and anemic and tumor syndromes; the ganglion areas were not enlarged. Para-clinically, a urine strip showed proteinuria + and negative hematuria. A blood count showed a hemoglobin level of 8.1 g/L and a white blood-cell count of 13,400/mm³. At that time urea was at 35 mmol/L, serum creatinine at 880 μmol/L; proteinuria was at 210 mg/d. After 4 weeks of hospitalization, a kidney biopsy was performed under general anesthesia because there was no other organ to be apparently dysfunctioning. The samples were sent out in a pathology laboratory in Kano (Nigeria), i.e. there is no reliable histopathology laboratory in Niger. Regarding histological findings in microscopy glomeruli were unremarkable and there were scanty preserved tubules. The rest of the tissue was overrun by sheets of fairly uniform-size lymphoid cells with hyperchromatic nuclei and scanty cytoplasm. No others specific features were seen (Figures 1). The immunohistochemistry revealed malignant lymphoid cells diffusely CD20-positive (Figure 2) malignant lymphoid cells diffusely CD3-negative (Figure 3) and nearly 100% of the tumour cells are Ki67-positive that indicating brisk proliferative activity (see Figure 4). It took a week to get the histopathological report.

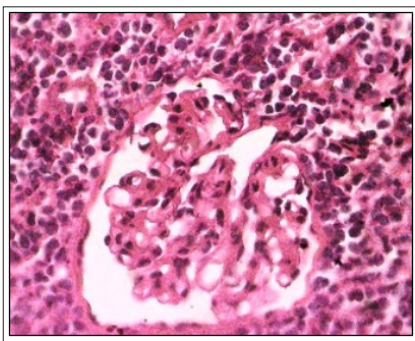


Figure 1: Malignant Lymphoid Cells with Fairly Uniformly Sized Nuclei and Scanty Cytoplasm Effacing the Renal Architecture, but Sparing the Glomeruli and Tubules. H and E Stain, x400

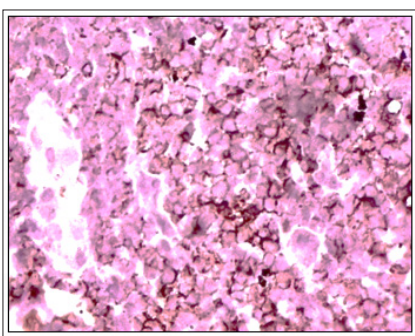


Figure 2: Malignant Lymphoid Cells Diffusely CD20-Positive. x400

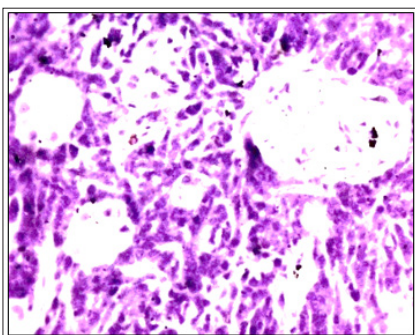


Figure 3: Malignant Lymphoid Cells Diffusely CD3-Negative. x400

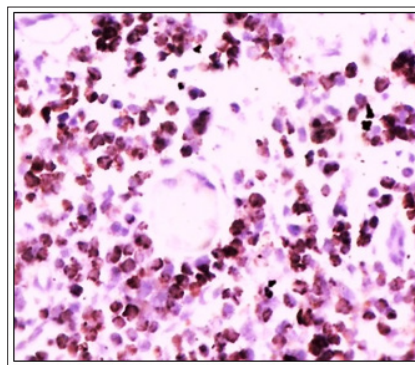


Figure 4: Nearly 100% of the Tumor Cells are Ki67-positive; Indicating Brisk Proliferative Activity. x400

In the meantime, therapeutically, our patient received a diuretic-based treatment: five blood transfusions and 11 hemodialysis sessions. Unfortunately, 24 hours after the last hemodialysis session, the patient suffered a cardiovascular arrest before implementing specific chemotherapy.

Discussion

Cases of African Burkitt lymphoma in children usually occur between ages 2 and 14 years, with a slight male predominance. In children it accounts for 20% of all types of cancer and 35--50% of cases of NHML [6]. However, renal Burkitt lymphoma is an extremely rare condition, accounting for less than 1% of kidney tumors. Its cause remains unknown but there are risk factors (Epstein--Barr virus, endemic malaria, immunodeficiency, and cytogenetic abnormalities) [2]. If NHML is suspected from the nature of the tumor syndrome and presence of kidney damage, its diagnosis is anatomo-pathological from a kidney biopsy and is then confirmed by immunohistochemistry. An early diagnosis permits better care.

In our 8-year-old patient, the histology showed tissue that was invaded by fairly uniformly sized lymphoid-cell clusters with hyper-chromatic nuclei and abundant cytoplasm. These characteristics strongly suggested a renal lymphomatous process. Immunohistochemistry of the kidney biopsy revealed malignant lymphoid cells erasing architecture but sparing the glomeruli and tubules. These diffuse lymphoid cells expressed CD20 but not CD3. Almost 100% of these tumor cells were positive for Ki67, indicating rapid proliferation.

Prognosis depends on the Murphy classification

Stage I: maxillary or orbit or isolated ganglion involvement;
Stage II: more than one maxillary, unilateral or bilateral with or without orbital involvement;
Stage III: any abdominal or intra-thoracic involvement, with or without facial involvement;
Stage IV: more than 10% cerebrospinal fluid and/or marrow invasion and clinical and biological scalability [7, 8, 9].

Suspecting renal Burkitt lymphoma, the presence of fever >38°C, hyperleucocytosis, and inflammatory anemia, we predicted the patient was stage III, and group B and b according to Murphy's prognostic classification. Our patient should have had a myelogram, an osteomedullary biopsy, a lumbar puncture, and a cerebral and medullary MRI. However, we did not have time to organize these exams. Because renal Burkitt lymphoma remains difficult to identify, Murphy's classification serves as a basis for treatment [5-11]. Regardless of the disease's stage, the reference treatment is poly-chemotherapy based on cyclophosphamide 300 mg/m²,

vincristine 1 mg/m², prednisolone 60 mg/m², cerebrospinal injection of both methotrexate 15 mg at D1 and Depomedrol 40 mg [12].

Conclusion

Renal Burkitt lymphoma is an extremely rare extra-nodal malignant blood disease. It should be suspected based on suggestive clinical signs. Histology and immunohistochemistry should be performed to confirm a diagnosis.

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