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Lymphoblastic Lymphoma: Epidemio-Clinical, Therapeutic and Prognostic Aspects

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Abstract **Original Research Article**

Lymphoblastic lymphoma (LL) is a multifactorial, frequent and highly progressive but curable cancer, and one of the haemopathies frequently managed at the Mohammed VI University Hospital, Marrakech. Seventeen patients with lymphoblastic lymphoma (LL) were included in this study. The annual hospital incidence of LL in Marrakech was 1.8 cases/year. The 6-10 age group was in the majority (n=11; 70%). The sex ratio was 3.25 in favor of boys, and 82% had RAMED, which corresponds to a low socioeconomic level. Dyspnea was found in 10 patients (60%). Stage 3 was in the majority (n=7; 70%). Diagnosis was made by cytology in 94% of patients, histology in 23% and immunohistochemistry in 82%. Four patients died of chemotherapy toxicity after blood product rupture, and 3 of disease progression on chemotherapy. The 10 patients in remission survived event-free for an average of 21 months.

Keywords: Lymphoblastic Lymphoma, Child, Marrakech, Epidemiology, Evolution.

INTRODUCTION

Lymphomas are malignant tumors that develop from the lymphoid system. They can occur in any organ containing lymphoid tissue, particularly in lymph nodes, the digestive tract and tonsils digestive wall and tonsils. Recently, further progress has been made on the new classification of lymphomas. These advances take into account morphological, histological, immunohistochemical and molecular criteria, as well as the nature of the cell of origin.

Non-Hodgkin's lymphomas represent a heterogeneous group of hemopathies characterized by malignant monoclonal proliferation of the lymphoid system (B or T cells), which tend to invade the whole organism. This heterogeneity translates into a variety of clinical, anatomopathological, immunological and cytogenetic presentations, and consequently, a very different prognosis from one form to another.

Only lymphoblastic lymphomas will be studied here. They arise from immature precursors (lymphoblasts) of the (B-LBL) and (T-LBL) cell lines in marrow or bone tissue. They are very rare, accounting for approximately 2% of all lymphomas, and occur in children between the ages of 6 and 11[3].

Diagnosis is based essentially on anatomopathological examinations (cytology, histology, immunohistochemistry). The treatment of lymphoblastic lymphoma was the first success of chemotherapy [3].

In Morocco, there have been few large-scale studies on the management of lymphomas, which motivated us to carry out an epidemioclinical study of lymphomas in children in the pediatric hematology and oncology department (SHOP) of the Mohammed VI university hospital in Marrakech.

MATERIALS AND METHODS

This is a retrospective study of 17 cases of children with lymphoblastic lymphoma from January 2012 and December 2021, i.e. over a period of 09 years. The files selected were those of patients who had spent at least ten days in hospital and had received at least one dose of chemotherapy. In addition, these files must contain anatomopathological and/or cytological confirmation of the diagnosis.

RESULTS

I. Epidemiological Study

A total of 17 cases of lymphoblastic lymphoma over a period of 09 years, an average of 1.8 cases per

year. The children were from consanguineous marriages in 65% of our patients from non-consanguineous marriages.

The distribution was as follows: 13 patients (76%) were male and 4 (24%) females. There was a predominance of males, with a sex ratio of 3.25. Patient age ranged from 13 months to 14 years, with an average of 7.5 years and a maximum frequency between 6 and 10 years.

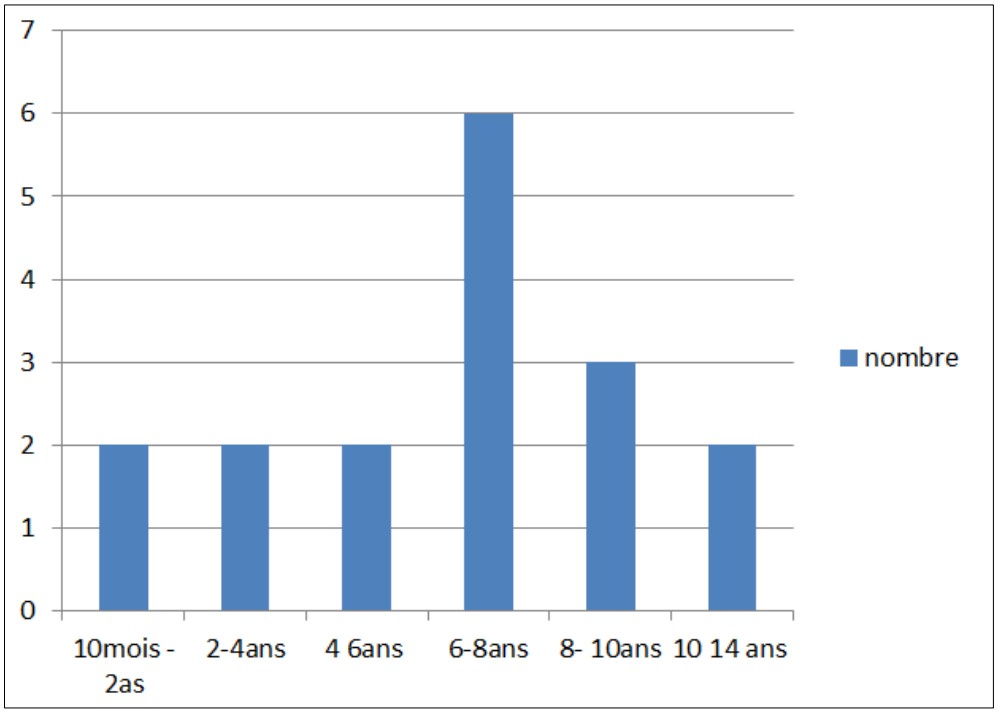

Figure 1: Distribution of patients by age.

The origin of our patients varies. 6 patients come from rural areas, i.e. 36% of cases, and the remainder from urban areas, i.e. 64% of cases. 80% of our patients attend school. In our series, 81% of patients are of low socio-economic status, and the percentage of mutualist patients does not exceed 8% (3 families).

II-CLINIC

A. Duration of Evolution before Hospitalization:

The time between the appearance of the first clinical manifestation likely to be related to the disease and admission ranged from 8 days to 5 months, with an average of 45 days. Fifty-two % of patients consulted the doctor less than a month after onset, 22% after between 1 and 2 months, and less than 5% after more than 4 months.

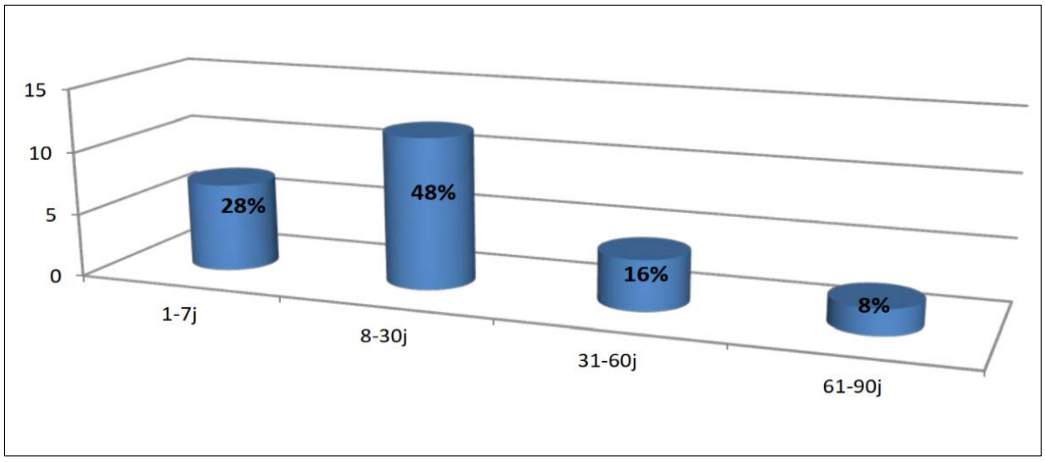

Figure 2: Patient consultation times from onset of symptoms to admission

B. Clinical Presentations:

1. Functional Signs

Dyspnoea was the main reason for consultation, accounting for 60.4% of patients. Next came in descending order, were superior vena cava syndrome and/or abdominal distension (17%), while 3 patients (11%) consulted for peripheral adenopathy. Only one patient had paraplegia.

2. Clinical Examination

On clinical examination, 60% of cases had a pleural fluid effusion syndrome;35% had peripheral adenopathy; 11% had peritoneal effusion and/or abdominal mass; 2 patients had pericardial effusion.

III- PARACLINICAL

A. Diagnostic work-up:

1- Chest X-ray

Carried out in 76% of patients, showed: Pleural effusion in 11 patients (78%); A mediastinal mass in the anterior mediastinum in 3 patients (17%);

2- Ultrasound

Abdominal ultrasonography was performed in 10 patients, and revealed: single or multiple abdominal masses in 2 patients; digestive thickening in 2 cases; abundant ascites and deep adenopathies in 2 cases; normal in the rest of patients

3- Computed tomography (CT)

Performed on 15 patients, showed: Mediastinal involvement in the form of a mediastinal mass in 10 patients (71%), 28% of patients with an abdominal mass, femoral vein thrombosis in a single patient, 7% with other localizations (cerebral and renal localizations).

4- MRI: Performed on a single patient with paraplegia, showed a medullary tumour process.

5-BIOLOGY: A biological workup is systematically performed on all patients, It showed a lysis syndrome in 4 cases.

B. Diagnostic Confirmation

Diagnosis was made by cytology following cytopuncture of adenopathies or masses in 94% of cases, and by histology in 23% by biopsy of the mass in cases where the mass was biopsiable or cytology was inconclusive. Immunohistochemical studies were requested in 82% of cases, in search of tumour cells using the following tumour markers: anti CD79a, anti CD 10, CD 20, CD 19, CD 22, CD2, CD 3, CD 16 and Ki 67 antibodies.

C. Histological Type:

Type T was the most frequent with a rate of 76% (13 patients) with a thoracic localization in 100% of cases, 4 patients had a thoracic and abdominal localization. Five % were B type (2 patients) and the remainder died before histological confirmation (10%).

D. Extension Work-Up

1) Myelogram

This was carried out in 76% of patients at the level of both crests, with only 4 patients (23%) showed bone marrow invasion at the time of diagnosis.

2) CSF Cytological Study:

Routinely performed in all patients was positive in 5 patients (29%) with neuromeningeal involvement on admission.

E. Patients' Initial Stage and Prognostic Groups:

The majority of patients (64%) presented with stage III disease and were classified in prognostic group B. prognostic group B stage IV accounted for 20% of cases.

IV-TREATMENT

The time from admission to the start of chemotherapy varied, from 1 day for some patients to 20 days for others. The average was 10 days, with 71% of patients starting chemotherapy within two weeks of admission, and 28% starting between 15 and 20 days after admission. Analysis of patient records showed that in 90% of cases, this delay was due to a lack of histological evidence. In 2% of cases, severe LTS was the cause, in 2% of cases, severe LTS was the cause, and in 2% of cases, the cause was a severe infectious condition requiring initial management prior to chemotherapy.

These patients were placed on alkaline hyperhydration combined with the administration of Zyloric®. They were monitored clinically and biologically, with clinical monitoring to detect signs of hyperkalemia or hypocalcemia, and daily ionograms, in addition to diuresis monitoring to prevent fluid overload.

Patients received different treatments according to histological type and stage of NHL:LT patients were treated according to the LMT 2004 protocol, and LLB patients were treated according to the MAT-GFAOP-11 protocol.

V. Evolution:

1. Remission

Complete remission was noted in (41%) of our patients were in complete remission (7cas/17): Two of our patients showed partial remission under treatment, i.e. 11% of cases: They retained a mass of 1.5 cm and 2.5 cm respectively at a follow-up of 2 years.

2. Complications:

2.1 Metabolic Toxicity:

We noted 5 cases (20%) of tumor lysis syndrome (TLS), 1 of which was a spontaneous TLS before the start of treatment, and 4 after the start of treatment.

2.2. Hematological Toxicity:

Ten patients (40%) experienced one or more episodes of cytopenias during treatment, necessitating transfusion. There were 10 cases of thrombocytopenia (58%), with 9 patients presenting a hemorrhagic syndrome hemorrhagic syndrome: purpura, digestive hemorrhage, retinal hemorrhage, cerebral hemorrhage. The platelet count ranged from 2,000 to 23,000 elements/mm3.

2.3 Mechanical Complications

One patient presented a thrombosis of the right femoral vein on a central catheter, he was put on anticoagulant with a good evolution.

3. Deaths:

In our series, we recorded 47% deaths (8 cases out of 17): 2 patients (11%) died before the start of treatment, 1 during the start of treatment, 1 after cure Ib, 2 after cure II b (11.7%) and 2 patients after the end of treatment. 75% of these patients died in hospital.

Causes of death were dominated by chemotherapy toxicity and disease progression under treatment.

DISSCUSION

NHL can occur at any age, but is rare before the age of 2. In Europe, LL is considered a disease of children and young adults, with peak incidence in the second decade of life. In this study, we found that the age group most affected was between 2 and 10 years, with a rate of 70.0%. The extreme ages were 13 months and 12 years. These results are similar to those of B.amina in Fez [2]. In the Bourahima a Mali series, patient age ranged from 3 to 11 years, with a maximum between 6 and 11 years [3].

There is a male predominance, particularly for lymphoma, where the sex ratio is 3:1 in both endemic and non-endemic zones [2]. The sex ratio for lymphoblastic lymphoma is 2:1 [2]. In our study, there was a clear male predominance, with an M/F sex ratio of 3.25.

Our results are similar to those of Irène [5], and B.amina, who respectively found in their study a predominance of 70.8% and 72% in favor of the male sex.

Most of the patients in our series come from underprivileged backgrounds, only 9% are of average socio-economic status. The origin of our patients varies. 6 patients came from rural areas (36% of cases), the remainder from urban areas (64% of cases).

NHLs are common in low- and middle-income countries. They are particularly in rural areas [2].

In Harif's series from sub-Saharan countries, 38% of cases lived in towns, 27% in villages, while 16.6% were of rural origin. Socio-economic conditions were precarious for most patients: parental income was estimated to be very low in 152/343, and low in 49/343, and only 12% of patients had social security coverage [7].

In our series, 81% of patients were of low socioeconomic status, and the percentage of mutualist patients did not exceed 8%.

In B.amina's series, 75% of patients with LL presented with an anterior mediastinal mass. They may present with chest pain, weezing, stridor, dysphagia, the main manifestation was dyspnea, superior vena cava syndrome is present in 3% of cases, fluid effusion syndrome in liquid effusion syndrome in 8% and severe respiratory distress in 3% of cases [9, 10].

As in our series, dyspnea and respiratory distress were the most frequent reason for consultation in 60% of cases, followed by superior sd cava in 11%.

The two most effective and frequently used protocols, with comparable survival rates of around 75-85%, are LSA2-L2 and BFM, initially described by Wollner in the USA and Riehm in Germany. These are intensive, semi-continuous and prolonged treatments lasting an average of two years. Asparaginase is an essential drug. CNS prophylaxis, also essential, is ensured by effective systemic treatment, IR and MTXHD. Brain irradiation is limited to CNS+ patients. A randomized European study (EuroLB02) is currently investigating dexamethasone compared with prednisone and the possibility of reducing the duration of treatment to to 18 months. Above all, she is looking for clinical and biological prognostic factors, which that would enable better stratification of treatment [8].

In our series, LTs were treated according to the LMT 96 protocol, based on the same principles as the LSA2L2 and BFM protocols. Improving mortality rates is the general rule in all management of lymphoma. Harif *et al*., observed 20% of deaths at the start of treatment, 45% in the pre-induction phase, 38% at induction and at induction and 17% at consolidation. He reported that treatment-related mortality decreased over the 3-year inclusion period (first year: 25.7%, second year: 19.1%, third year: 11.6%). Improved supportive care resulted in a increase in overall survival rates from 54% in year one to 73% in year three [7].

This high death rate is explained, on the one hand, by the delay in consultation due to lack of financial resources, ignorance and misunderstanding of the disease.

Most of our patients only consulted us after the initial symptomatology had worsened and some had consulted traditional healers before coming to us.

To us. On the other hand, this death rate is explained by the delay in diagnosis after admission, with an average delay in starting treatment of 15 days.

Extension work-up and obtaining anatomopathological results. Five of our patients died as a result of therapeutic toxicity. These deaths could have been avoided simply by the availability of blood products. Once again, we need to continue raising awareness of the need for donors. This context justifies the choice of less toxic protocols for African countries south of the Sahara, where the technical platform is even less efficient.

CONCLUSION

NHL in children has become a curable disease, whose prognosis has been radically transformed thanks to a sound diagnostic and therapeutic strategy based on prevention of SLT, polychemotherapy and hematological resuscitation measures. However, whatever the efforts made by specialized centers caring for these patients, results can only be improved if the diagnosis is rapidly established, before the onset of nutritional and metabolic complications responsible for early death during the initial phase of management.

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