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G. ATIPO-TSIBA, L.O. NGOLET, I. KOCKO, A.D. ELIRA AND D.LUSINA

SUMMARY

We present the case of malignant Non-Hodgkin splenic lymphoma with villous lymphocytes regarded as atypical chronic lymphoid leukemia. This was a 62 years old male patient admitted in the Haematologic Department of Brazzaville Teaching Hospital for an enlarged spleen, anaemia and lymphocytosis. The initial abdominal CT noticed a homogenous splenomegaly and a large retroperitoneal tumour mass measuring 148 X 101mm. The initial count blood cell revealed a lymphocytosis with a lymphocyte count at 82 Giga/l, severe anaemia with a haemoglobin rate at 4.2g/dl, platelet count at 68 Giga/l. Peripheral smear examination showed irregular lymphoid cells with a homogenous distribution, condensed chromatin corresponding to villous lymphocytes. Immunophenotyping showed B lymphoid monotypic population cells positive for CD19, CD 20, and FMC7, moderately positive for CD23 and negative for CD5, CD43 and CD 79b. Therapeutically, combination chemotherapy RCVAD (Rituximab, Vincristin, Daunorubicin, Dexamethasone) give good clinical and haematological response. This report illustrates the need for excellent interdisciplinary collaboration and the interest of the cytology and immunophenotyping in the lymphoid proliferation management.

INTRODUCTION

The splenic lymphoma with villous lymphocytes is a rare chronic lymphoproliferative syndrome of B cells. It is considered as a leukemic stage of splenic lymphoma of the marginal zone. This disease is characterised by the presence of villous lymphoid cells in the blood. This pathology is a relatively benign entity. But some cases may require treatment because of symptomatic splenomegaly and / or severe cytopenia (1). Few cases have been reported in Africa. We repport one case of the aggressive forme of splenic lymphoma with villous lymphocytes, with better evolution after treatment and ten months of follow up.

CASE REPORT

A 62 years old Man admitted to the hematology department for exploration of a voluminous splenomegaly associated with leukocytosis.

The symptoms began 5 months before by an abdominal pain in the upper quadrant and left flank, causing dyspepsia, intermittent constipation, and heavy sweating. The evolution was marked by the installation of progressive physical weakness.

His initial survey noted: conscious patient, pallor of the skin and mucous membranes; conjunctival jaundice, temperature 38. 4°C, current weight 67 Kg (74 kg previous weight).

Abdomen taut, shiny, lack of collateral circulation, bulky mass of the left hypochondrium reducing lumbar touch extended up to 18 cm from the lower costal margin (splenomegaly) associated with moderate hepatomegaly (hepatic arrow 13 cm), no peripheral lymph nodes, rectal examination was normal. Tachycardia at 112 / min PA 130/70 mmHg, and the rest of the clinical exam was unremarkable.

Additional tests:

- Thoracic XR was Normal
- Upper gastrointestinal endoscopy showed an esophageal varices grade I and duodenal ulcer
- Abdominal CT showed a bulky kystiforme regular retro peritoneal tumor mass measuring 148 x 101 mm, located in the rear cavity of the omentum displacing the stomach above the pancreas, with an homogeneously increasing of spleen volume (figure 1).
- Initial blood count: leukocytes 129 giga / l (lymphoid cells 86 giga / l), Hb 4.2 g / l; platelets 68 giga / l. cytological appearance of blood smears (Figure 2) shows the presence of lymphoid cells having inhomogeneous cytoplasmic irregularities distributed over the contour, dense chromatin, corresponding to lymphocytes villous VS: 40 mm (first time); AST 40 U / L ALT: 24UI / l LDH: 542U / l, creatinine: 16 mg / l.
- Proteinogram, total protein 81 g / l (albumin 44.3 g / l; Gamma globulins 13.9 g / l; uric acid 44.5 mg / l Factor V 113.5% creative protein 217 mg / l.
- Serologies (HIV; Hepatitis B ; Hepatitis C) were negative. Immunophenotyping of the cell

population (Beckmann Coulter FC 500) showed a lymphoid population CD19 + B monotypic, CD5 _, CD23 + partial CD20 + high intensity, CD43 _ FMC 7+, CD79b _

The treatment, outside general measures, was a chemotherapy RCVAD type (Rituximab 500 mg DT Day1, cychophosphamide 800 mg DT Day1 and V8, vincristine 1 mg DT and DT Daunorubicin 10 mg continuous infusion of 24 hours Day1 to Day4 and Dexamethasone 40 mg DT Day1 to Day5) in 21-day cycle.

The evolution was marked by complete resolution of all clinical signs after three (3) cures.

A full assessment after three treatments allowed to continue to the 4 th complementary treatments (28-day cycles). Almost four (4) months after cessation of any treatment, the patient remains in complete clinical and hematologic remission.

Figure 1

Axial section of mesenteric region after injection of contrast product

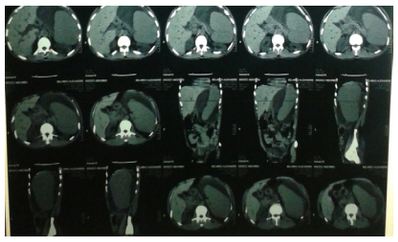
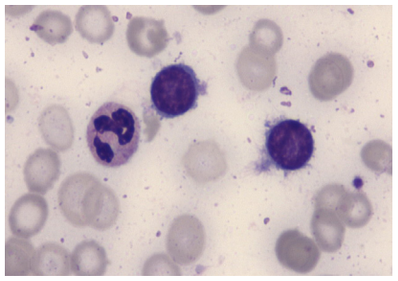


Figure 2

Presence of lymphoid cells on peripheral blood smear



DISCUSSION

The Splenic lymphoma with villous lymphocytes is an uncommon and rare chronic hémoprolifération lymphocytic B lymphoid cells which are similar in size and appearance to chromatin cell chronic lymphocytic leukemia B (1, 4, 7.8). This pathology seems to have a male predominance with an average age between 60 and 65 (3, 6, 8). Clinically the disease is characterized by an almost constant splenomegaly (1, 3, 9), frequent hepatomegaly, but the association with extra nodal tumor lesions is rare and exceptional (2, 7). Immunophenotyping confirms the nature of B lymphoid cells that are often negative CD5 (5,8). The positivity of the lymphoid cells with CD20 markers, CD19, IGMS is constant according to the literature (2, 4, 5). The cytogenetic analyses when performed, can help find certain abnormalities such; t (14; 14) (q32.13; q32.33) deletions of del (14) when using the technique of hybridisation in situ by florescence (10) not done in this case by lack of materials. It is reported in some cases (6, 8) involvement or association with a chronic infection with hepatitis virus C (HCV classical serology is negative in our case). The treatment described in the series often associated splenectomy (3), chemotherapy based on alkylating associated with splenic irradiation (1, 9) outside of septic complications leading to unsatisfactory long-term results.

Very few studies demontrated encouraging therapeutic responses with fludarabine and déoxyconformycine in immothérapie (6,8). We note, however in this case an excellent medium-term therapeutic response by combining rituximab to a polychemotherapy adapted to local resources as well as to the acute presentation as initial leukemia.

In conclusion the presentation of this exceptional clinical case by semiological signs justifies the importance of cytology combined the peripheral blood and the bone marrow as well as the immunophenotyping in the diagnosis of chronic lymphocytic proliferation. There is no consensus on the treatment of this type of clinical cases.

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