

Splenic Marginal Zone Cell Lymphoma: Case Report

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DOI: [10.36347/sasjs.2020.v06i02.013](https://doi.org/10.36347/sasjs.2020.v06i02.013)

Received: 19.02.2020 | Accepted: 26.02.2020 | Published: 29.02.2020

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Abstract

Case Report

Splenic marginal zone cell lymphoma (LZMS) is a very rare B lymphoma; Representing 2% of all NHL, this lymphoma invades the spleen, perisplenic nodes and frequently the marrow, which can be a source of diagnostic traps. Patient aged 65, hospitalized for the exploration of a splenic tumor mass confirmed by a computed tomography (CT) scan. The biological assessment finds an inflammatory SD + high LDH. A splenectomy was performed. The microscopic study of the operating room showed a diffuse lymphomatous proliferation with small cells, with labeling by CD20. CD 5 and CD 43 are negative. The diagnosis of LZMS was accepted. LZMS affects the subject over the age of 50, usually characterized by the presence of a large splenomegaly without lymphadenopathy. The hemogram shows in three quarters of the cases the inconsistent presence of villous lymphocytes. The diagnosis is essentially anatomopathological, it shows a constant nodular or sometimes diffuse attack of the white pulp of the splenic parenchyma. The tumor cells are small, expressing the B lymphoid markers: CD19, CD20, CD22, CD79. They are negative for CD5, CD10, cyclin D1 and CD43. No specific cytogenetic abnormality of LZMS was identified. It is an indolent lymphoma, the treatment of which has not yet been codified, depends on prognostic factors. Death is linked to the risk of transformation to large cell lymphoma.

Keywords: Splenic marginal zone lymphoma, splenectomy, prognosis.

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INTRODUCTION

Splenic marginal zone cell lymphoma (LZMS) is a very rare, low-grade, low-grade non-Hodgkin's lymphoma (NHL) B, which accounts for 1 to 2% of all NHL [1]. It is a separate entity in the WHO 2016 classification of hematopoietic tumors, which sometimes poses a problem of differential diagnosis with other indolent lymphomas [1]. It is characterized by isolated splenomegaly without lymphadenopathy, sometimes associated with moderate blood lymphocytosis.

CASE REPORT

65-year-old patient, without significant pathological ANTCD, hospitalized for pain in the left hypochondrium evolving for 2 years, with postprandial vomiting evolving in a context of unencrypted weight loss.

The clinical examination finds a splenomegaly; the biological assessment finds an inflammatory SD + high LDH. The CT scan finds a tumor-like tissue process of the spleen, measuring 95

mm long axis, associated with some splenic lymphadenopathies and minimal ascites (Figure 1).

A diagnostic and therapeutic splenectomy was performed (figure 2), the postoperative follow-ups were simple. Histological and immunohistochemical aspect is compatible with diffuse large B-cell lymphoma, with tumor proliferation with small round cells, arranged in a diffuse layer (figure 3), the tumor cells are large, rounded and atypical (figure 4), diffuse expression of CD20 by tumor cells (figure 5), CD3 marks the reaction T lymphocytes (figure 6), and the proliferation index evaluated by Ki67 is high (figure 7).

The diagnosis of splenic lymphoma of the marginal zone was retained. An extension assessment was carried out including a BOM, which showed a suspicion of spinal infiltration. The patient benefited from adjuvant chemotherapy due to the presence of poor prognostic factors (abdominal lymphadenopathy, increased LDH, presence of large cells with splenic histology).



Fig-1: Spleen tissue mass measuring 95mm major axis



Fig-2: Macroscopic image of the splenectomy patch. The splenic parenchyma is occupied by a heterogeneous whitish lesion

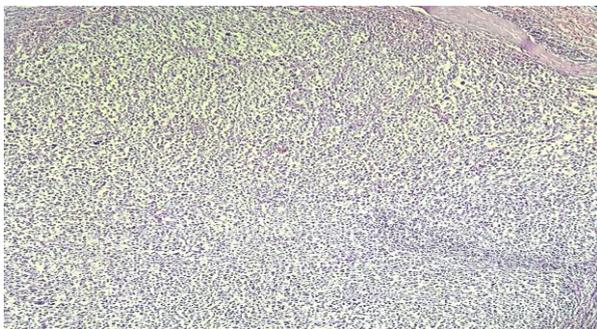


Fig-3: tumor proliferation with small round cells, arranged in a diffuse layer.

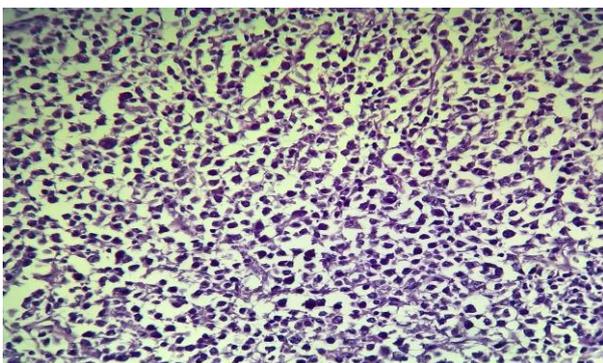


Fig-4: The tumor cells are large, rounded and atypical

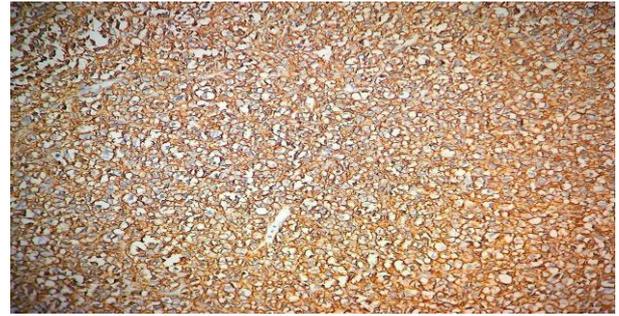


Fig-5: Diffuse expression of CD20 by tumor cells

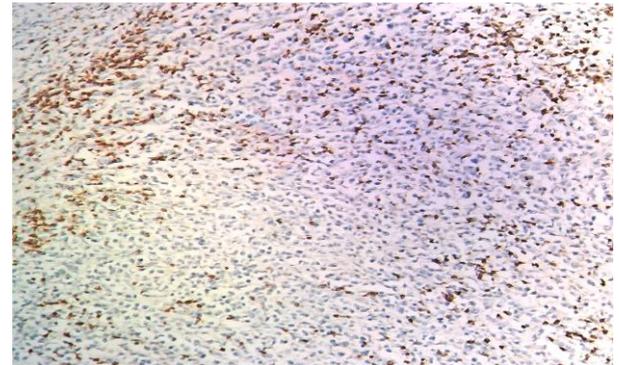


Fig-6: CD3 marks reactive T cells

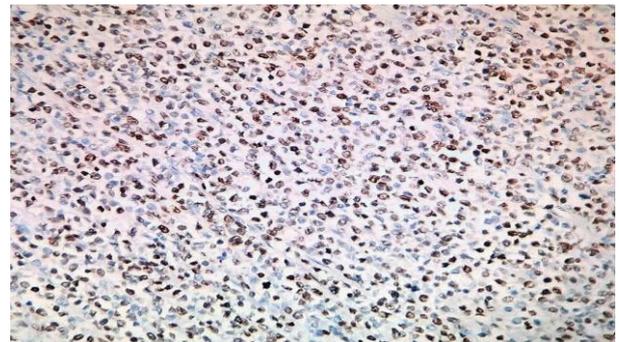


Fig-7: The proliferation index evaluated by the Ki67 is high.

DISCUSSION

First described in 1992 [1], LZMS is an indolent non-Hodgkin's lymphoma originally splenic [1], which represents 1 to 2% of all non-Hodgkin's lymphomas [1]. It preferentially affects subjects over 50 with an average age at diagnosis of 65 and a sex ratio of 1. In some series, LZMS seem frequently associated with chronic HCV infection. Other authors have shown that it is associated with an active chronic HCV infection that regresses with antiviral therapy [1, 2]. The usual presentation is that of a limited tumor syndrome with isolated splenomegaly, sometimes associated with lymphadenopathy of the splenic hilum [1]. More rarely, hepatomegaly and intra-abdominal lymphadenopathy are reported (10% of cases). Medullary infiltration is almost constant, 95% of LZMS being immediately stage IV at diagnosis [2]. The blood count found circulating lymphomatous cells in 64% of cases, sometimes taking on the appearance of villous lymphocytes [1-3]. One or more cytopenias are frequently found, in connection with hypersplenism, an

autoimmune mechanism or, a central mechanism by spinal cord invasion. Protein immunoelectrophoresis reveals a monoclonal protein in a third of cases, most often of the IgM type, and rarely exceeding 30 g / l [2]. The blood smear shows the inconsistent presence of villous lymphocytes. The most reliable diagnostic criterion is the achievement of a typical splenic histology, or in the absence of splenectomy, the presence of a circulating B lymphocyte clone with a villous morphology [1, 3]. Tumor cells express one of the B lymphoid markers: CD19, CD20, CD22, and CD79b. They are negative for CD5, CD10, Cyclin D1 and CD43. If there are indisputable cases of LZMS CD5 +, the diagnosis must nevertheless be retained with extreme caution and it is advisable to eliminate in these unusual cases the diagnoses of chronic lymphoid leukemia or of pre-leukemic phase of lymphoma with mantle cells. The expression of DBA44 is observed in approximately 80% of cases [1,4]. CD11c expression is often present, but inconsistently (47%) [1, 4]. Exclusive splenic involvement remains rare but not exceptional, as our observation shows. In some cases, both blood immunophenotyping and BOM are negative, and only a pathology examination of the splenectomy patch can make the diagnosis [1,5]. This shows a proliferation affecting the marginal perifollicular area which destroys the mantle, surrounding and replacing the germinal centers of the residual white pulp. Sinusoidal infiltration is very characteristic. These are small round cells with a fairly abundant cytoplasm, and with irregular nuclei, sometimes villous, which colonize the follicles, surrounded by larger cells with clear cytoplasm which resemble the cells of the area. Autoimmune manifestations [1, 2, 4]. This autoimmunity is often of isolated and multiple biological translation in the same patient: autoimmune thyroiditis and antinuclear antibodies, autoimmune pancytopenia, rheumatoid factor and antinuclear antibodies, antiphospholipid syndrome, adult Still's disease, circulating lupus anticoagulant antibody and Gougerot Sjögren syndrome (SGS) [1, 2, 4, 7]. No specific cytogenetic abnormalities LSZM has not been identified, however Deletions in 7q as well as Translocations involving the genes of the Ig heavy chains of immunoglobulins whose t (11; 14) (q3; q32), are the most frequent anomalies [1,8].

The development is indolent with a median survival of 8 to 13 years depending on the series, and a five-year survival of 76% [1, 9]. Death is linked to the progression of lymphoma or its transformation into diffuse large cell lymphoma, which occurs in around 10% of cases [10, 11]. The prognostic factors identified are not the same according to the series. Thieblemont *et al.* [7] show that the occurrence of autoimmune manifestations and the existence of a monoclonal protein are associated with a shorter time to lymphoma progression. In the series by Arcaini *et al.* [12] covering a larger number of patients (309 cases), the detrimental factors identified are the existence of anemia

(hemoglobin less than 12g / dl), an increased LDH level, and a decreased albumin (<3,5g / dl) [1, 12].

To date, the treatment is still poorly codified; it varies between abstention from therapy, splenectomy, chemotherapy (anthracyclines, fludarabine) and / or splenic irradiation. No treatment has significantly lengthened the survival of patients with LZMS. If the LZMS associated with the hepatitis C virus are primarily antiviral treatment, splenectomy remains the treatment of choice in case of cytopenia or major splenomegaly, chemotherapy being proposed in case of contraindication to surgery or clinical progression after splenectomy. Adjuvant chemotherapy after splenectomy in the event of poor prognostic factors (general signs of evolution, abdominal lymphadenopathy, increase in LDH, presence of large cells with splenic histology) increases the rate of complete remission but does not modify survival, nor the risk of transformation and relapse [1].

CONCLUSION

Splenic lymphoma of the marginal zone is an indolent lymphoma, the treatment of which until now has not been standardized, depends essentially on prognostic factors. Overall survival is 76% at 5 years. Death is linked to the risk of transformation to large cell hemopathy.

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