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CIRCULATING VILLOUS LYMPHOCYTES IN PERIPHERAL BLOOD - SMZL A CASE PRESENTATION.

Pathology

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ABSTRACT

SMZL with leukaemic manifestation and hairy cell leukaemia are uncommon haematological malignancies, originating from spleen and bone marrow respectively and both share overlapping clinical and morphological features.We present a rare case of SMZL, diagnosed with help of flowcytometry. Monoclonal B cells expressed CD19,CD20 and Kappa and lacking CD103,CD23 and Lambda expression. Single dose of Rituximab improved hematological parameters further confirm the diagnosis.

KEYWORDS

SMZL (Splenic Marginal Zone Lymphoma) , Circulating Villous Lymphocytes, Cytoplasmic Projections, Polar Distribution, Flowcytometry, Immunophenotype.

INTRODUCTION

Lymphocytes having villous morphology in peripheral blood film and Bone marrow aspiration are uncommon. Villous lymphocytes are cells having an oval nucleus with the "cobblestone" pattern of nuclear chromatin typical of more mature lymphoid cells. The cytoplasmic projections found on cells looks like villi which have different varieties of distribution in different disorders. Villous lymphocytes may be mistaken for hairy cells, however in hairy cells projections have more even distribution around circumference of cells whereas polar distribution in Splenic marginal zone lymphoma (SMZL). There is considerable overlapping of lymphomas which displays villous lymphocytes circulating in the peripheral blood with splenomegaly such as HCL, SMZL, HCL-V and PLL [6]. Marginal zone lymphomas represent a group of lymphomas originating from B lymphocytes, present in distinct compartment called Marginal zone of secondary lymphoid follicles [1]. Few SMZL cases have been reported in literature.

SPLENIC MARGINAL ZONE LYMPHOMA (SMZL)

SMZL is a B cell neoplasm consisting of small cells, involving the white pulp follicles of spleen, hilar lymph node, bone marrow and peripheral blood [1]. It is an incidental diagnosis made after short history of splenomegaly, peripheral lymphocytosis and cytopenias [5].

In adults MZL accounts for 5-17% of all Non-Hodgkin's lymphoma, from which very few cases of SMZL are reported [1]. To diagnose SMZL and distinguish it from indolent B cell lymphoproliferative disorder is challenging, without spleen histology, but is possible with help of immunophenotyping (flowcytometric) results [5].

We describe a case of a middle aged male patient having massive splenomegaly with characteristic immunoprofile. Diagnosis of SMZL was made on basis of flowcytometry report.

Case Report

A prospective study was done on a 58 year old male patient of Northwestern zone of India attended the Medicine department of Geetanjali Medical college and Hospital, Udaipur Rajasthan with chief complaint of abdomen pain in left iliac region since 15 days, followed by fever since 3 days, weakness, anorexia and weight loss. He had no specific past history of Diabetes Mellitus, HTN, TB. Patient had moderate build with BMI 0 14 Kg/m^2. On his physical examination we found massive splenomegaly. Blood investigations, Ultrasonography, CECT abdomen were advised.

1. His CBC report was Hemoglobin – 11.4 gm/dl, TLC – 46000/cumm, Neutrophils - 16%, Lymphocytes - 68%, monocytes – 15%, Eosinophils -1%.
2. Peripheral blood film and Bone marrow aspiration shows villous lymphocytes having hairy cytoplasmic projections.
3. NCCT Whole abdomen depicts gross splenomegaly and hepatomegaly. Spleen enlarged in size measures 26 cm, splenuculus is also noted.

At last for confirmation Flowcytometry was advised which reveals Non CLL Bcell Chronic Lymphoproliferative disorder with diagnosis of SMZL on bais of presence and absence of various CD markers.

DISCUSSION

In this report we illustrate a rare case of SMZL, its clinical features, blood investigations and treatment therapy. A prospective study was done on a middle aged North western male patient of India who came at Gmch, Udaipur with chief complaints of abdomen pain in left lumbar region since 15 days, fever since 5 days, weakness, and anorexia and weight loss since few days. He had non specific past history. His built was moderate with BMI – 14. USG, CT abdomen reports reveals splenomegaly, hepatomegaly and splenuculus. According to blood reports lymphocytic leukocytosis with mild reduced platelet count, clinical picture had D/D of HCL, HCL-V and SMZL. In this era of precision medicine confirm and early diagnosis is must for starting early treatment.

From recent studies it was found that confirm and early diagnosis of Leukemias and Lymphomas can be made by immunophenotyping /flowcytometry.Specific CD markers expression and absence helps in confirming various diagnoses. Flowcytometric report of our patient reveals it as a Non CLL B cell lymphoma, i.e, SMZL (6, 7). Markers in favour of SMZL were IgM, IgD, CD19, CD20 were expressed and CD79a, CD5, CD10, CD103 were not expressed (7, 8).Therefore this particular panel of monoclonal antibodies confirms diagnosis of SMZL and rules out HCL.

In a study on diagnosis of Splenic B cell lymphomas in Bone marrow, Amie Behdad et al, illustrates that presence of monoclonal B cells lacking expression of CD5 and CD10 markers can be excluded from other low grade B cell neoplasm, as follicular lymphomas are CD 10 +, CLL is CD5/CD23 + and MCL is CD5 +. Some 10% expresses CD5 while SMZL never expresses CD103, CD11c, and CD25 (HCL) (2).

Aliyah R et al in his study reveals immunoprofile of SMZL,i.e, IgM+,IgD+/-,CD19+,CD20+,CD5-,CD10-,CD23-,CD25-,CD103-. HCL have CD19+,CD20+, CD23-,CD103+,CD11c+,CD25+,CD123+ , CLL have CD19+,CD20+,CD23+,CD5+,CD10- and in Follicular Lymphoma CD20+ and CD10+ (3).

Tanus Vigetal discusses following markers, SMZL – CD25+, CD103+/-, CD123-. HCL-CD25+, CD103+, CD123+,HCL-V-CD25-,CD103+,CD123-. SDRPL – CD25-, CD103-,CD123-.(4)

Santos et al and Arcaini et all tells importance of rituximab monotherapy in treatment of SMZL. (5,6). Santos et al depicts SMZL immunoprofile as - CD19+,CD20+,CD22+,CD79a/b+,CD5-,CD10-,CD43- and CD23-/+ (6) whereas Arcaini depicts CD20+,CD79a+,CD5-/+,CD10-,CD103-.(5)

Finally, Baceggio et al in 2010 studied CD5 expression, a subset of

SMZL with higher lymphocytosis on 24 cases and concluded the existence of SMZL cases having CD5+ profile and tells about requirement of more case studies on this topic.(7)

Treatment – Patient was then treated symptomatically and by monoclonal therapy of Rituximab. Patients of SMZL improves fastly by Rituxiamb monotherapy (7, 8). His TLC falls down to 11000 by single dose of Rituximab within 3 days and he improved symptomatically. Complete follow up for 6 months was done of patient and no recurrence was noted.

Table 1: Comparision Of Various Immunophenotypic Findings In Various Lymphomas – (1)

VARIABLE	CD5	CD10	CD11C	CD25	CD103
SMZL	-/+	-	+/-	-/+	-/+
HCL	-	-/+	+	+	+
HCL-V	-	-	+	-	+
LPL	-/+	-	+	+/-	-

Table 2: Comparision Of Antiglobulins:

VARIABLE	IgM	IgD	IgG
SMZL	+	+/-	+/-
HCL	-/+	+	+
HCL-V	-/+	-/+	-/+
LPL	+	-	-

SMZL: Splenic marginal zone lymphoma, HCL: Hairy cell lymphoma, HCL-V: Hairy cell lymphoma – variant, LPL: lymphoplasmacytic lymphoma ,+ :positive,- :negative.

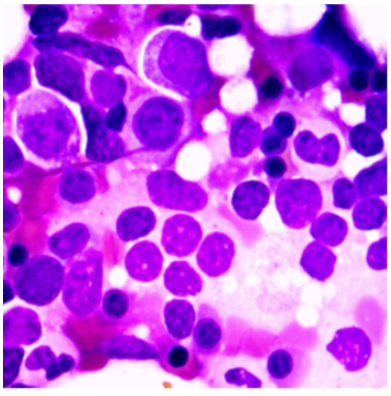

Fig A. Circulating Villous Lymphocytes In Bone Marrow Aspiration.

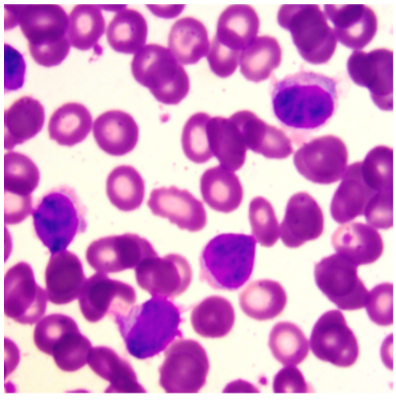

Fig.B : Circulating Villous Lymphocytes With Polar Distribution Of Cytoplasm In Pbf.

CONCLUSION

Despite the varinity of SMZL, there has been a major effort made to give confirm and early diagnosis of various lymphomas. The villous lymphocytes circulating in the peripheral blood can be mistaken as hairy cells of HCL, but in SMZL the projections are localized to one or both poles of cell. Bone marrow shows lymphocytic infilterates. The neoplastic cells of SMZL expresses IgM, IgD, CD19, CD20 and CD79a. They do not expresses CD5,CD10,CD103. CD11c is expressed by nearly ½ of patients with SMZL in flowcytometric reports. Very good prognosis of Rituximab monotherapy is reported in SMZL patients. Therefore specific molecular targets, immunophenotyping, studies are very much effective for early and confirm diagnosis of SMZL patients for starting their proper treatment. And Rituximab monotherapy had proven best treatment option for SMZL patients to recover.

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