Academic Journal of Cancer Research 6 (2): 65-68, 2013
ISSN 1995-8943

DOI: 10.5829/idosi.ajcr.2013.6.2.76105

A Case of CLL/SLL with Rapid Courses

B. Pushpa and J. Jeya Shambavi

Department of Pathology, Government Villupuram Medical College,
Villupuram, Tamil Nadu State, India

Abstract: A 55 year old woman presented with loss of weight and loss appetite for 6 month duration. Examination revealed generalized lymphadenopathy. Peripheral smear demonstrated hyper leukocytosis with predominant cells is lymphocytes. Prolymphocytes constituted 9%. Aspiration cytology of lymph node revealed increase in cellularity with monotonous population of small round lymphocytes with scattered atypical lymphocytes in background of red blood cells. A case of CLL/SLL is reported. This patient had group B symptoms suggesting a worst prognosis.

Key words: CLL/SLL · Prolymphocytes · FNAC · Richter's Syndrome

INTRODUCTION

Chronic lymphocytic leukemia/small lymphocytic lymphoma CLL/SLL is a Non Hodgkin's Lymphoma (NHL) characterized by neoplastic proliferation of B cells and both CLL and SLL share many morphologic and immunophenotypic features [1]. Most cases have a leukemic presentation at diagnosis. CLL and SLL are considered by WHO as one disease at different stages and not two separate entities [2-4].

Case Report: A 55 year old female patient presented to the OPD in Tertiary Care Centre in Government Villupuram Medical college and Hospital, TamilNadu with history of loss of weight, excessive sweat and loss of appetite for the past 6 months. There was also a history of fever with chills of one month duration. Examination revealed generalized lymphadenopathy involving cervical, axillary and inguinal nodes. The nodes were rubbery and non tender. Routine haematological investigations revealed Hb of 7g %, total WBC count of 1,25,000/cu.mm with predominance of lymphocytes constituting 87%. Platelet count was 1.5 lakhs /cu.mm.

Morphology: Peripheral smear revealed an increase in total WBC count with predominance of lymphocytes more than 85% (Fig.1). Among lymphocytes, atypical

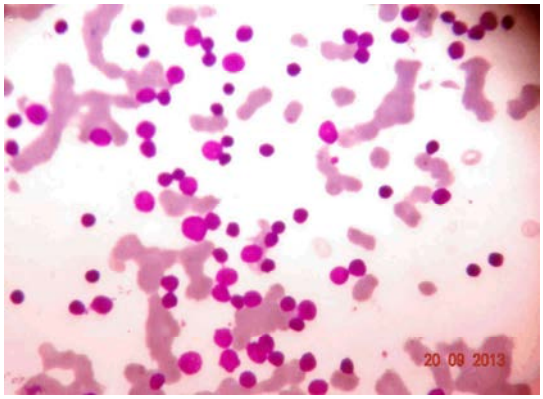

Fig. 1: Peripheral smear picture of CLL

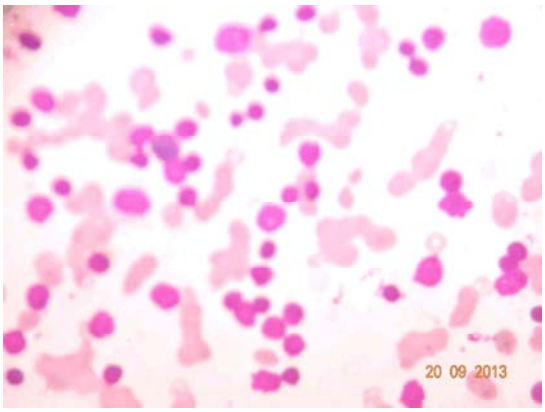

Fig. 2: Atypical lymphocytes and prolymphocytes under oil immersion

Corresponding Author: B. Pushpa, Department of Pathology, Government Villupuram Medical College, Villupuram, Tamil Nadu State, India.

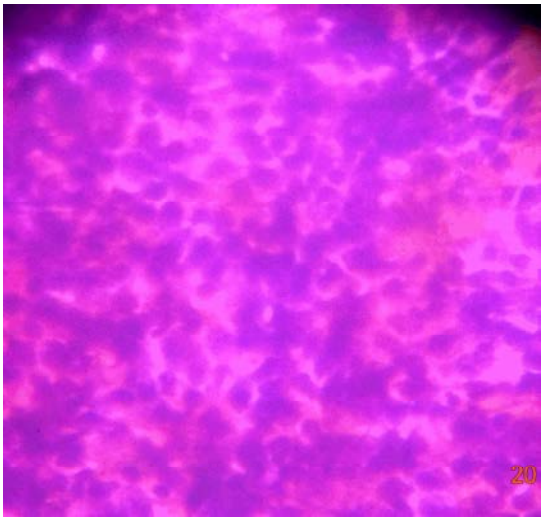

Fig. 3: FNAC Cervical node with increased cellularity

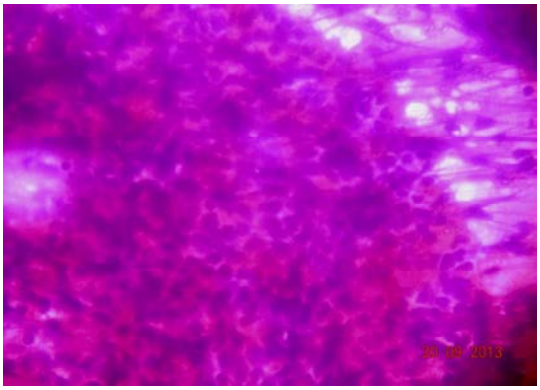

Fig. 4: FNAC Axillary node with sheets of neoplastic lymphocytes

*DR.B.PUSHPA, PROFESSOR OF PATHOLOGY
E Mail: ID:raja_pushpa@yahoo.co.in
Mobile: 9444295055
MS WORDS:1514
REF NO: 76105-AJCR

lymphocytes are seen and prolymphocytes constitute around 9% (Fig. 2). Occasional smudge cells are also seen. Red blood cells are microcytic and hypochromic. Platelets are adequate.

Ultrasound examination of the abdomen revealed a mild splenomegaly.

FNAC of the nodes revealed a lymphoid aspirate. Microscopy shows an increased cellularity(Fig.3) with monotonous population of small round lymphocytes with scattered atypical lymphocytes in background of red blood cells. The lymphocytes reveal scanty cytoplasm and nuclei with clumped chromatin.FNAC from axillary node reveals same picture (Fig.4).

Hence a diagnosis of CLL/SLL was made. This patient had group B Symptoms suggesting a worst prognosis.

DISCUSSION

CLL/SLL is a mature B cell neoplasm seen in middle and old age. It is a heterogenous disease and there is proliferation of CD5+ mature lymphocytes in blood, bone marrow and lymphoid tissue. Most cases are incidentally found in routine hemogram analysis and some present as either like leukemia or lymphoma with atypical lymphocytes in peripheral blood. However clinical presentation depends on the stage at presentation. Around 40% cases present with group B symptoms with decreased survival [5]. The median survival is around 10 year. However in the presence of group B symptoms the survival is poor.

The incidence of CLL/SLL is 6% in western world. According to Daniel Carreon *et al* [13] the incidence of CLL/SLL is higher in Indians than in western world. Males have higher incidence than females and the ratio is 17:10. CLL/SLL is an indolent neoplasm which usually well to chemotherapy [6].

CLL/ SLL is a neoplasm of mature B cells. In typical case, patient presents with fatigue, symmetric, bilateral lymphadenopathy and with mild hepatospelenomegaly. In the early stage, organomegaly is not present and incidentally found when examining peripheral smear for some other conditions. In late stage, bonemarrow the hematopoietic precursors are replaced by the proliferating lymphocytes and the patient presents with anemia. The lymph nodes involved are mainly cervical and supraclavicular lymphnodes. The lymphnodes reveal total effacement of nodal architecture by monotonous population of proliferating B lymphocytes with majority of cases showing spillage of tumor cells into the perinodal tissue. The tumor cells are small with round nuclei, condensed chromatin, inconspicuous nucleoli and scanty cytoplasm. A mild to moderate degree of pleomorphism is noted among the tumor cells [7]. This can cause diagnostic confusion with the most common differential diagnosis of mantle cell lymphoma. Mitosis is less.

In peripheral smear, presence of more than 5×10^9 B lymphocytes /L is essential for the diagnosis of B-CLL.In SLL, there should be presence of lymphadenopathy and /or splenomegaly with count of B lymphocytes in peripheral blood not exceeding $5x10^9$/L [8]. In diagnosis of early CLL is made count of

more than 4. $5x10^9$ /L is present. The lymphocytes are uniform, regular with nuclei and the chromatin is coarsely clumped. Atypical lymphocytes with nuclear indentation, folding and large sized lymphocytes may be seen. Smudge cells which are fragile cells formed while making smear are also seen.

Admixed with the lymphocytes are prolymphocytes which are larger cells with vesicular nuclei with prominent nucleoli and with moderate amount of cytoplasm. Based on number of prolymphocytes,the FAB subclassification is CLL,when prolymphocytes are less than10%. When prolymphocytes are 11-55%, it is called CLL with prolymphocytic transformation. When prolymphocytes are more than 55%, it is called prolymphocytic leukemia [9].The prolymphocytes when more than 10%indicates poor prognosis 80% of patients present with classical CLL and 20% have CLL/PLL or atypical CLL.

The stage of the disease is also assessed by bone marrow trephine biopsy histology in which the bone marrow infiltrate may be interstitial, nodular, mixed or diffuse.

CLL/SLL in which transformation occurs in aggressive lymphoma in which lymphnodes which demonstrate an immunoblastic large cell NHL but blood and bonemarrow reveal CLL picture. This is called as Richters syndrome and is seen in 3 to 8% of patients with CLL. The tumor mass spreads from lymphnodes to other lymphoid organs and in all cases bone marrow is involved.

CLL/SLL is reactive for CD5 and CD23 positive [10] ant this is especially useful in differentiating CLL/SLL from Mantle cell lymphoma. These cells are positive for pan B cell marker such as CD19, CD20 and CD21 [11]. Immunophenotyping is useful in differentiating CLL/SLL from other B Cell lymphoid proliferative disorders.

Here we have reported a case of CLL/SLL in female patient with group B symptoms with a short duration of history which is very rare for this disease. However recent epidemiological data suggests that the disease incidence is increasing in India.

To conclude, chronic lymphocytic leukemia (CLL) is defined as a malignant lympho proliferative disorder of small, mature-appearing monoclonal B-lymphocytes in the blood cells, bone marrow and lymph nodes. Small lymphocytic lymphoma (SLL) is a lymph node counterpart of CLL. The cell morphology, course and prognosis of the disease are almost similar [12]. CLL/SLL is a case of distinct entity with characteristic cytological and hematological features which has to be diagnosed in elderly. Most patients live up to 5-10 years after diagnosis but some die 2-3 years after diagnosis. Accurate diagnosis is essential as it has a favorable prognosis and amenable to treatment.

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