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CHRONIC LYMPHOCYTIC LEUKAEMIA IN YOUNG: A CASE REPORT

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ABSTRACT

Chronic lymphocytic leukemia is a neoplastic entity pertaining to lymphocytes when they get accumulated in either lymph nodes thereby call as Small lymphocytic lymphoma or spilled up in the circulation of the blood. These lymphocytes are mature looking, relatively immunologically competent usually expressing B cell phenotype markers. The median age of affection of patients at the time of diagnosis is 71 years and its incidence increases with age. Its incidence in an age group <50 years is quite uncommon accounting only to 10-15% of total diagnosed cases. Here we are presenting such a rare case report of chronic lymphocytic leukaemia where age of affection is less than 40 years.

KEYWORDS: B cell phenotype, Chronic lymphocytic leukaemia,Small lymphocytic lymphoma, Young adults.

INTRODUCTION

Chronic lymphocytic leukemia is a neoplastic entity pertaining to lymphocytes where either they get accumulated in lymph nodes there by causing a neoplasm of lymh node known as Small lymphocytic lymphoma or spilled up in the blood circulation to produce leukemic picture known as Chronic lymphocytic leukaemia. These lymphocytes are mature looking, relatively immunologically competent usually expressing B cell phenotype markers. Clinical course of the disease is quite variable (1-2).

Most of the time the diagnosis is incidental as patients have come for symptoms pertaining to some other disease entities and while performing routine investigations for that disease ; complete blood count reveals leukemic leukocytosis and very high absolute lymphocyte count (3). initial asymptomatic phase may finally lead to certain manifestations characterstic of the disease for e.g. pyrexia of unknown origin, loss of appetite, weight loss, lymphadenopathy, hepato-splenomegaly and shortness of breath on exertion (3).

The median age of affection of patients at the time of diagnosis is 71 years and its incidence increases with age. (4) Its incidence in an age group <50 years is quite uncommon accounting only to 10-15% of total diagnosed cases (5-6).

Though elderly population show B-cell immune-phenotype markers in CLL ; it has been found that Young age CLL patients show lymphocytes expressing T-cell immune-phenoype markers (3). Here we are reporting a case of CLL in young adults.

CASE REPORT

A 30 years old male in medicine ward presented with complains of bilateral lower limb swelling for 1 year, generalized weakness for 15 days and cough with sputum for 10 days. Patient was not giving any complain of pain in the lower limb, fever, breathlessness, night sweats, petechae or bleeding, malena or pain in abdomen. Patient was a bidi smoker (1 pack/day). General examination reveals pallor accompanied with generalized lymphadenopathy characteristically involving the submandibular, sub mental, post auricular and posterior triangle lymph nodes. On further evaluation we found that the lymph nodes were discrete, mobile, non tender, firm to hard without any signs of acute inflammation. On Respiratory system examination reveals bilateral rhonchi. Central nervous system examination and cardiovascular system examination were all within normal limit. On per abdomen examination, abdomen was soft and non-tender.

The haematological examination revealed; Hb—4.6 gm/ dl, Hct—35.8%, RBC—3.97 * 106 /microl, MCV—65 fl, MCH—23 pg, MCHC—31 g/dl, TLC—4100/cumm, DLC-N-45, L-40, E-2, M-3, Platelet count—2 lacs. Peripheral blood smear showed microcytic hypochromic red blood cells with anisopoikilocytosis.. WBC series is within normal limits. Occasional (1%) prolymphocytes were also seen. Large number of smudge cells were present.

On USG whole abdomen: Mild hepatomegaly with heterogenousechotexture, moderate splenomegaly was found.

A diagnosis of CLL was made and therapy started. The patient is doing well after 6 months of follow up.

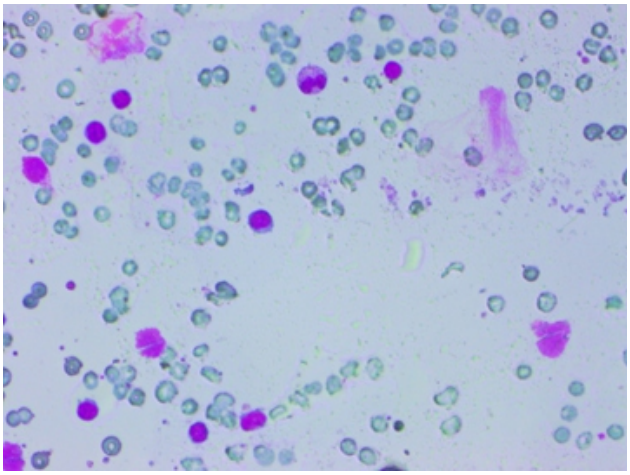

Fig. 1: Peripheral blood smear (Leishman stain 100x), showing smudge cells & lymphocytosis

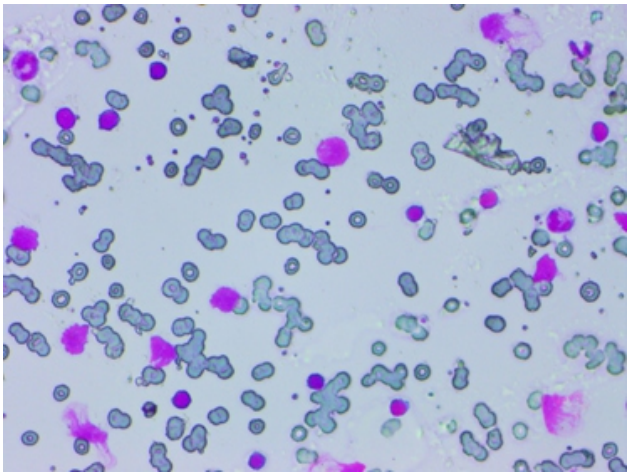

Fig. 2: Peripheral blood smear (Leishman stain 100x), showing smudge cells & lymphocytosis

DISCUSSION

CLL is a disease entity affecting mainly elderly population , with a median age of affection is 71 years with a slightly higher male preponderance shown by the M:F ratio of 1.7:1 (3)CLL is slowly progressive disease (3).In western population CLL is a commonest form of chronic leukemia with an incidence of 25-30% whereas least common in Asian continent with an incidence of 2-3%(3) Certain databases like National Cancer Institute's Surveillance and Epidemiology End Results(SEER) database study, showed that in 2009; 11% of all the CLL diagnosed patients were <55 years of age (4).Similarly , a population based study done in Europe indicates that 7-20% of patients with CLL are < 55 years of age at diagnosis (7-8). Survival in CLL patients is age independent and has the potential to alter the utility of prognostic testing given the higher mortality from competing health problems in older individuals (9-13).

In a study by Tait D, Shanafelt, MD et al, it has been found that CLL patients who were aged <75 years at the time of diagnosis had shorter survival than that of the aged matched general population, and moreover it was not affected by the stage of disease but survival did not differ from the age matched general population among CLL patients aged > 75 years at diagnosis.

In a study by Parikh S.A. et al, presentation of young CLL patients showed significant differences with that of the older patients. It showed that young patients were more likely to present with intermediate risk disease and young CLL patients were likely to suffer from adverse clinical and biological characterstics.

Montserrat et al found progressively increasing number of newly diagnosed CLL matched by a constant rate of about 20% young patients per year and that similar clinical feature among young patients and elderly patients. Moreover traditional clinical factors for example stage of the disease, lymphocyte count, lymphocyte doubling time and histology of bone marrow bear the same known prognostic effect on survival. The only siginificant difference between the two age groups is higher male female ratio in the younger group which can be due to the fact that young females having protective endocrine effect.

Due to better performance status, lower incidence of severe comorbities, and a different attitude towards the diagnosis of CLL and available treatment optionsit was found that younger patients take more interest in clinical trials. On the other hand, younger patients also experience significant psychological stress related to the diagnosis of CLL because of certain issues like repercussions for employment status and responsibility towards their children and family(14).

Importance of analyzing the CLL in young adults lies in several ways like clinical characterstic of CLL in young patients have not been known; it is not described weather the prognostic parameters identified for CLL can also be applied to younger patients. It also helps in identifying young patients with CLL as candidates for experimental treatment approaches like bone marrow transplantation, high dose chemotherapy and hematopoietic growth factor(15).

Newer prognostic markers for CLL are genomic abnormalities detection by FISH or by array based karyotyping, Ig heavy gene somatic mutation (IgVH)

status, expressing of ZAP 70,thymidine-kinase activity of neoplastic cells and beta 2 microglobulin level (14).

High expression of ZAP70 and CD38 is always associated with an aggressive clinical course and poor prognosis (16).

Dohner et al reported the effect on survival for patients carrying adeletion 11q was greater in patients <55 years of age(17).

It was found that in comparison to time to first treatment and overall survival in all age groups, stage was found to be a powerful predictor (14). In elderly patients a traditional conservative management is done; for younger patients it is unsatisfactory and curative therapeutic efforts should be done(18-20).

Consistent proportion of complete response can be induced by certain Purine analogues like fludarabine, however, even in apparent complete responders residual disease is still present suggesting that in most, if not all, cases the leukemic clone is not eradicated (21-23).

The limited data available appear promising, because both autologous and allogeneic transplants can effectively induce long-term disease-free survival and in some cases the molecular disappearance of the leukemic clone (24).

CONCLUSION

CLL being a leukemia of elderly population is extremely rare in young adults, but if it occurs, it is significant in many ways. Importance of analyzing the CLL in young adults lies in several ways like clinical characterstic of CLL in young patients have still not been known; it is not described weather the prognostic parameters identified for CLL can also be applied to younger patients. It can also help to identify the candidates for experimental treatment approaches like bone marrow transplantation, high dose chemotherapy and hematopoietic growth factor. Moreover in elderly patients a traditional conservative management is done; for younger patients it is unsatisfactory and curative therapeutic efforts should be done. So because of all these reasons awareness of occurrence of this entity in young population is extremely useful.

ABBREVIATIONS

CLL- Chronic Lymphocytic Leukeamia

MCV- Mean Cell Volume

MCH- Mean Cell Hemoglobin

MCHC- Mean Cell Hemoglobin Concentration

TLC- Total Leukocyte Count

DLC- Differential Leukocyte Count

WBC- White Blood Cell

CD- Cluster Differntaition

FISH- Flourescent In-Situ Hybridisation

SEER-Surveillance and Epidemiology End Results

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