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Plasma Cell Leukemia–A Case Report and Review of Literature

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Case Report
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Abstract

Plasma cell leukaemia (PCL) is one of the most aggressive and rarest forms of plasma cell dyscrasia. As prognosis is very poor, it is very important to recognize this entity sufficiently early so that one can offer combination chemotherapy at the earliest which can prolong survival.

Keywords: Plasma cell leukaemia; Hepatosplenomegaly; Total Leukocyte Count

Abbreviations: PCL: Plasma Cell Leukaemia; MM: Multiple Myeloma; TLC: Total Leukocyte Count; IG: Immunoglobulin.

Introduction

Plasma cell leukaemia (PCL) is an unusual and aggressive form of plasma cell dyscrasia [1]. Primary PCL occurs de novo, whereas secondary PCL is the leukaemic transformation of relapsed or refractory multiple myeloma (MM). The incidence of PCL is 2–4%, of which 60–70% of cases are primary and 30–40% are secondary, although the incidence of the latter type is rising [2,3]. By definition, there are more than 20% plasma cells in the peripheral blood and an absolute plasma cell count of more than 2x109/L [4,5]. The median age of patients with primary PCL is younger than those with secondary PCL (52–65 years old versus 65–70 years old) [2]. Clinical presentation of PCL includes anemia, thrombocytopenia, renal dysfunction, hypercalcemia, bone pain, lytic lesions, infections, and hepatosplenomegaly etc.

Case Report

A 61-year-old male presented to the emergency with complaints of weakness, fatique, backache, dyspnea and high grade fever since 1 month. He did not have an significant past and family history. During a general physical examination, there was evidence of pallor without lymphadenopathy, his vitals were stable. A systemic examination was unremarkable with no evidence of hepatosplenomegaly.

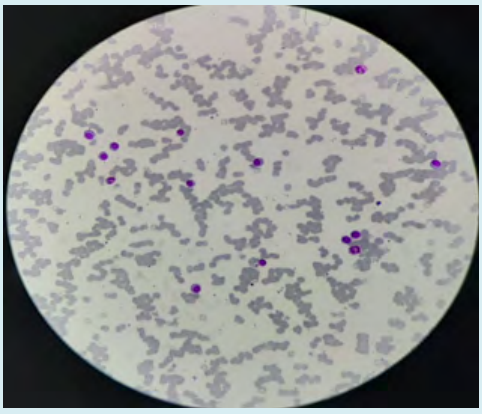

Figure1: Low power view PBF showing rouleax formation (Giemsa- 40X).

A complete hemogram showed anaemia with a haemoglobin (Hb) level of 7.5 g/dL, leukocytosis with a total leukocyte count (TLC) of 26 × 109/L and platelet count of 2Lac/mm^3. A peripheral blood smear showed moderate

degree of anisopoikilocytosis with microcytic/hypochromic red blood cells, macrocytes and elliptical cells along with extensive rouleaux formation (Figure 1). A differential leukocyte count revealed 38% plasma cells with an absolute plasma cell count of 9880/mm^3 (Figure 2).

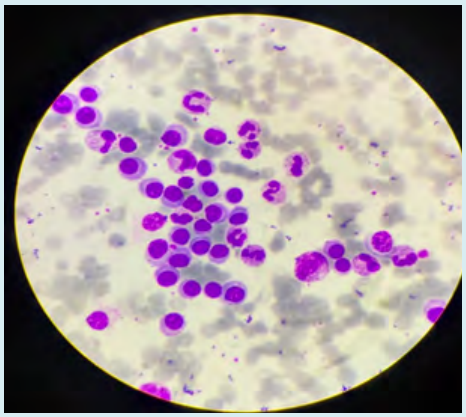

Figure 2: High power view: PBF shows many Plasma cells (Giemsa- 100X].

The erythrocyte sedimentation rate was 70 mm/hour. Biochemical investigations: Renal function tests were deranged showed high levels of serum creatinine (3.3 mg/dL), blood urea (147 mg/dL) and serum uric acid (13.0 mg/dL). Radiological investigations, including a chest X-ray, abdominal ultrasound and skeletal survey, did not show any lesions. Serum protein electrophoresis was done. High resolution serum protein electrophoresis revealed densely staining monoclonal gammopathy a Monoclonal spike (M spike) in gamma globulin region. The raised globulins were found to be immunoglobulin (Ig) D on serum immunofixation electrophoresis. Bence Jones proteins were absent in the urine.

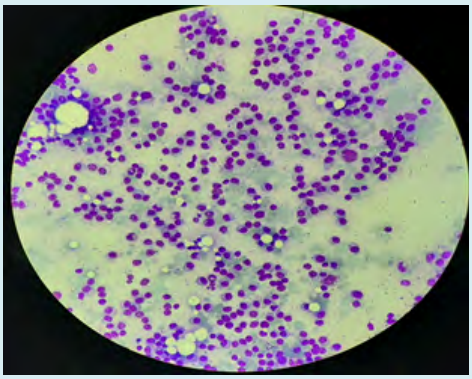

Figure 3: Bone marrow aspirate showing diffuse involvement by plasma cells (Giemsa- 40X).

Bone marrow aspiration and biopsy were done in the department of pathology GGSMC Faridkot. BM aspirate smears showed diffuse involvement of bone marrow by plasma cells constituting 62% of all hematopoietic cells (Figure 3). Binucleated forms also noted. Bone marrow biopsy shows sheets of Plasma cells (Figure 4). Based on the findings of peripheral smear and bone marrow the final diagnosis of Plasma Cell Leukemia was made (pPCL).

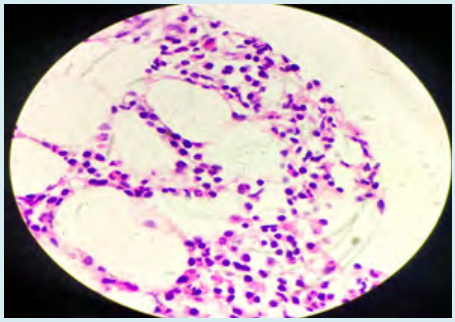

Figure 4: Bone marrow biopsy showing sheets of Plasma cells (Giemsa- 100X).

Discussion

Plasma cell leukemia is a rare and aggressive variant of plasma cell dyscrasia, accounting for only 2-4% of all plasma cell dyscrasias .The first case of plasma cell leukemia (PCL) was recognized by Gluzinski and Reichentein [6]. The presentation may be primary, de novo, or secondary, evolving from an existing case of myeloma as part of the terminal phase of the disease. About 60% to 70% of cases are primary. The incidence of primary PCL is very rare and reported to occur in less than one in a million [7]. PCL patients usually have accompanying anemia, hypercalcemia, renal insufficiency and organomegaly. Two types of PCL are seen: secretory and non-secretory. No M-protein is detected in the non-secretory type of PCL. PCL is more frequent in the light chain only, IgE and IgD myeloma and is less frequently seen in IgG or IgA myeloma.

By definition, there are more than 20% plasma cells in the peripheral blood and an absolute plasma cell count of more than 2x109/L [4,5]. Clinical presentation of Plasma cell leukemia is more aggressive than that of multiple myeloma with a higher presenting tumour burden and higher frequencies of extramedullary involvement, anemia, thrombocytopenia, hypercalcemia, renal impairment, increased levels of serum lactate dehydrogenase, beta-2 microglobulin and plasma cell proliferative activity [8]. In a case series by Rajeswari, et al. four out of 16 patients with PCL were under the age of 40 years, with the youngest being 25 years old [9]. Although primary and secondary PCL are considered different clinical entities, both have a poor

prognosis. The median survival rate ranges from 6.8–12.6 months without novel therapy, although this increases to over three years after autologous stem cell transplantation [1,10]. Though the clinical and laboratory features of primary and secondary plasma cell leukemia are similar, the response to therapy and overall survival in primary plasma cell leukemia goes from poor to worse. Response to treatment of PCL is poor. Median survival is less than 1 year. Since the prognosis is so poor, intensification of high dose chemotherapy followed by allogenic/autologous stem cell rescue should be tried [11]. The most important prognostic factor in pPCL remains response to treatment as patients presenting with the disease that is resistant to initial therapy have the poorest outcome [12].

The current treatment plan follows the steps involved in the management of multiple myeloma. Induction therapy includes various bortezomib-based regimens such as VDT-PACE (bortezomib, dexamethasone, thalidomide, cisplatin, adriamycin, cyclophosphamide, and etoposide), VDT (bortezomib, thalidomide, and dexamethasone), VAD (bortezomib, doxorubicin, and dexamethasone), VRD (bortezomib, lenalidomide, and dexamethasone) or VMP (bortezomib, melphalan, and prednisone). Although the best induction regimen for PCL is not known. Recently, lenalidomide and bortezomib-based regimens have demonstrated activity and are more widely used. In a multicenter retrospective study involving 42 patients with pPCL, bortezomib-based therapies were associated with 69% response rates, 1–3-month median survival time [13]. Although there is great variability in treatment plans, typically individuals <65 years in good performance status are treated with aggressive induction therapies such as VDT-PACE. PCL has a relatively poor prognosis, due to its very aggressive nature involving extramedullary organs, lytic bone lesions and bone marrow failure. Treatment includes immunomodulators, proteasome inhibitors, and autologous stem cell transplantation. Outcomes are not promising, however, even after treatment; median survival after chemotherapy and transplant is not more than three years [14]. As prognosis is very poor, it is very important to recognize this entity sufficiently early so that one can offer combination chemotherapy at the earliest which can prolong survival.

Conclusion

Plasma cell leukemia is a rare and aggressive form of plasma cell dyscrasia with poor prognosis. It is very important to recognize this entity early so that appropriate treatment regimens initiated.

References

1. Jelinek T, Kryukov F, Rihova L, Hajek R (2015) Plasma cell leukemia: From biology to treatment. Eur J Haematol 95(1): 16-26.

2. Tiedemann RE, Gonzalez Paz N, Kyle RA, Santana Davila R, Price Troska T, et al. (2008) Genetic aberrations and survival in plasma cell leukemia. Leukemia 22(5): 1044-1052.

3. Bladé J, Kyle RA (1999) Nonsecretory myeloma, immunoglobulin D myeloma, and plasma cell leukemia. Hematol Oncol Clin North Am 13(6): 1259-1272.

4. McKenna RW, Kyle RA, Kuehi WM, Harris NL, Coupland RW, et al. (2017) Plasma cell neoplasms. In: Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, Thiele J, editors. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. 4th (Edn.), France. IARC.

5. Noel P, Kyle RA (1987) Plasma cell leukemia: An evaluation of response to therapy. Am J Med 83(6):1062-1068.

6. Gluzinski A, Reichentein M (1906) Myeloma und leucaemia lymphatica plasmocellularis. Wien KlinWochenschr 19: 336.

7. Naseem S, Kaur S, Gupta R, Kashyap R, Nitayand S (2012) Plasma cell leukaemia; Case series from a tertiary centre with review of literature. Indian J Hematol Blood Transfus 28(1): 10-14.

8. Kar R, Priyadarshini SG, Niraimathi M, Basu D, Badhe BA (2012) Clinico-pathological spectrum of primary plasma cell leukemia diagnosed at a tertiary care centre in South India over 5 year period. Indian J Hematol Blood Transfus 28(3): 170-174.

9. Rajeswari G, Paul TR, Uppin MS, Uppin SG, Rao DR, et al. (2014) Plasma cell leukemia: A case series from south India with emphasis on rarer variants. Indian J Med Paediatr Oncol 35(3): 211-214.

10. Fernández de Larrea C, Kyle RA, Durie BG, Ludwig H, Usmani S, et al. (2013) Plasma cell leukemia: Consensus statement on diagnostic requirements, response criteria and treatment recommendations by the International Myeloma Working Group. Leukemia 27(4): 780-791.

11. Mak YK, Chan CH, Chen YT, Lau SM, So CC, et al. (2003) Consolidation therapy with autologous blood stem cell transplantation in a patient with primary plasma cell leukaemia. Clin Lab Haematol 25(1): 55-58.

12. Musto P, Simeon V, Todoerti K, Neri A (2016) Primary plasma cell leukemia: Identity card 2016. Curr Treat Options Oncol 17(4): 19.

13. Katodritou E, Terpos E, Kelaidi C, Kotsopoulou M, Delimpasi S, et al. (2014) Treatment with bortezomib-based regimens improves overall response and predicts for survival in patients with primary or secondary plasma cell leukemia: Analysis of the Greek myeloma study group. Am J Hematol 89(2): 145-150.

14. International Myeloma Working Group (2003) Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Working Group. Br J Haematol 121(5): 749-757.