Case Report

Primary plasma cell leukemia in 32 years old male – A case report

Disha Singla[1], Gunvanti B. Rathod[2*]

[1]PG Student, [2]Assistant Professor
Pathology Department, SBKS MI & RC, Sumandeep Vidyapeeth, Vadodara, Gujarat, India

[*]Corresponding author email: **neempath@gmail.com**

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Abstract

Plasma cell leukemia (PCL) is a rare disease and is the least common variant of multiple myeloma accounting for 2-3% of all plasma cell dyscrasias. Histogenetically, plasma cell leukemia is derived from terminally differentiated B cells. It is diagnosed by presence of absolute plasma cell count >2000 per cm or >20% circulating plasma cells. Here we report a case of plasma cell leukemia, who presented with easy fatigability, weakness and high grade fever since 1 month. Hematological investigation revealed leukocytosis with plasmacytosis (7420/ mm^3). On bone marrow examination, >45% plasma blasts were seen. Biochemical analysis showed high LDH level (4236 U/L) and serum calcium level was also raised (12.3 mg/dl). Final diagnosis of plasma cell leukemia was made. As PCL is rare disease and it is even rarer to find them in a 32 years old. Here we are able to find and document the typical features of PCL.

Key words

Primary plasma cell leukemia, Leukocytosis, Plasmacytosis.

Introduction

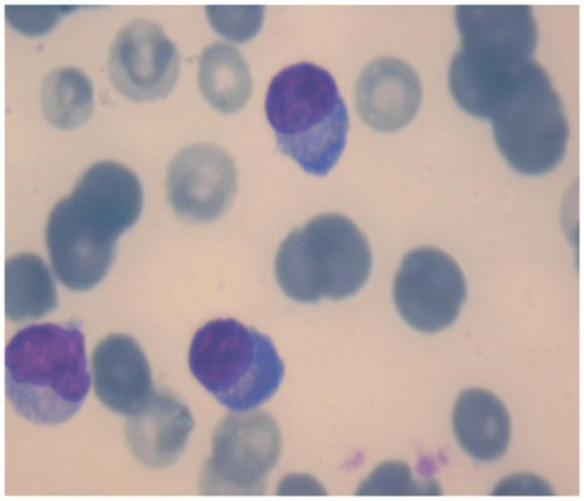
Plasma cell leukemia (PCL) can be considered the leukemic variant of multiple myeloma. The first cases of PCL were described in the early part of the twentieth century [1, 2]. The diagnosis is based on hematological features, including a plasmacytosis exceeding 2 x 10^9/ L and any evidence of a clonal plasma cell proliferation. There are two forms of PCL: the primary form occurring in individuals without preceding multiple myeloma, and the secondary form arising as a late manifestation in patients with multiple myeloma. Overall, incidence of PCL is less than 1 case per million populations [3]. The prognosis for primary and secondary PCL is reported as being poor with median survival of

about 6–8 months. The poor prognosis appears due to the biologically aggressive nature of the disease and the reduced performance status. More recently, autologous and allogeneic stem cell transplantation (SCT) has been utilized for the treatment of PCL [4]. In our case, despite of all the treatment patient expired which showed aggressiveness of the disease.

Case Report

47 years old, male presented with easy fatigability, weakness and high grade fever since 1 month. Abdominal ultrasound showed mild hepatomegaly with moderate splenomegaly. Hematological investigation revealed leukocytosis (35180/ mm^3) with plasmacytosis (7420/ mm^3). **(Photo – 1)** Platelet count of patient was low (63000/ mm^3). Urine examination showed Bence Jones proteinuria. Bone marrow aspiration revealed >45% plasma blasts. The cells were large in size with round to oval eccentric nucleus with pale blue cytoplasm. Few cells were multi nucleated and few showed peripheral cytoplasmic flare known as flame cells. **(Photo – 2, Photo – 3, Photo - 4)** There was reduction of megakaryocytes.

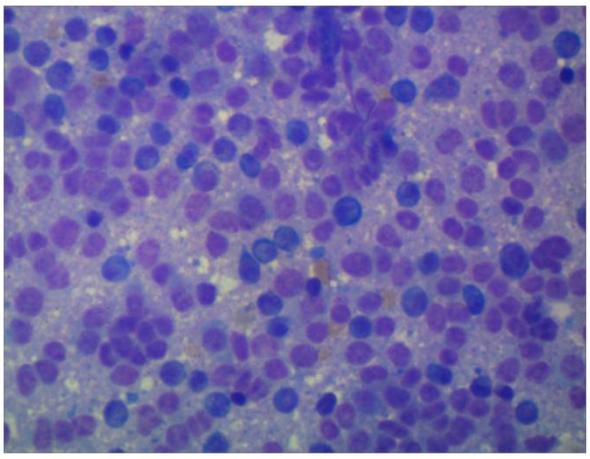
Photo - 1: Peripheral smear shows plasmacytosis. (Leishman stain, 40X)



Total serum calcium was elevated (12.3 mg/dl) with raised LDH (4236 U/L). Plasma electrophoresis revealed elevated β2 microglobulin level (4.3 g/dl) (normal value – 1.3-1.5 g/dl) **(Photo – 5)** and also serum globulin level was raised (6.1 g/dl) (normal – 1.8-3.6 g/dl). After clinopathological correlation, patient was diagnosed as plasma cell leukemia despite of all treatment the patient expired on twenty first day after diagnosis.

Photo – 2: Plasma electrophoresis shows a discrete band at gamma globulin level.

Photo – 3: Smear showed >45% of plasma blasts in bone marrow aspiration. (Leishman stain, 10X)



Discussion

Plasma cell leukemia (PCL) is a rare variant of multiple myeloma accounting for 2-3% of myeloma and other plasma cell dyscrasias. PCL is defined as >2,000/ mm^3 circulating plasma

cells with a plasmacytosis of >20% of the differential white count [5, 6, 7, 8]. Han, et al. (2008) reported on epidemiological and survival characteristics of 254,702 cases of lymphoid neoplasm from 1973 to 2003 and they found 221 cases of PCL among 42,065 cases of plasma cell neoplasm. Median age of PCL patient at diagnosis was 64 years and having worst prognosis of all lymphoid malignancies [9].

Photo – 4: Multi nucleated plasma blasts in bone marrow aspiration. (Leishman stain, 40X)

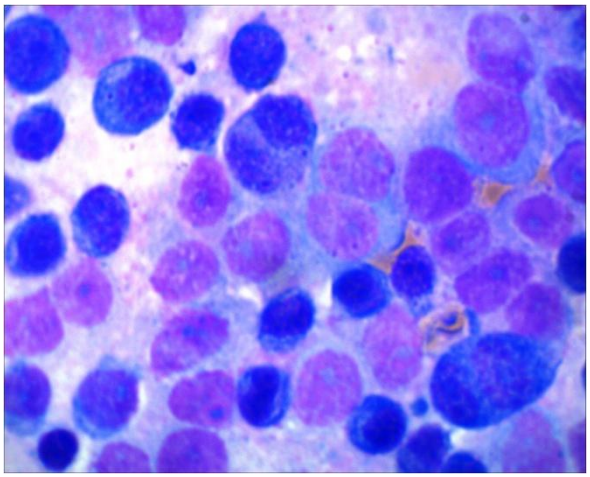
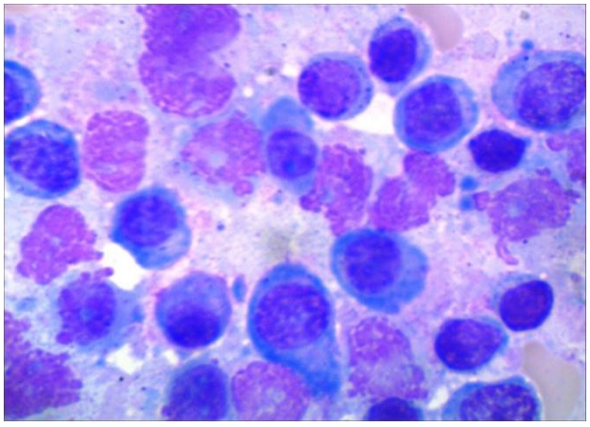


Photo – 5: Plasma blasts with flaring of cytoplasm known as flame cells in bone marrow aspiration. (Leishman stain, 40X)



Patient with primary plasma cell leukeumia are younger with extra osseous organ involvement and increased frequency of renal failure, anemia, thrombocytopenia, hypercalcemia, increased LDH, and β2 microglobulin. Patient's liver and spleen commonly involved, as compared to secondary disease [5, 6, 7, 10, 11]. They will rapidly progress to terminal stage of the disease. Our patient presented with hypercalcemia and increased LDH which are the known bad prognostic signs in the already aggressive disease [6, 12]. In contrast, secondary plasma cell leukaemia (SPCL) is advanced bone disease which often develops extra medullary manifestation later in disease course and nodal as well as splenic involvement is rare event.

Primary plasmacytomas has been associated with prior exposure to chemotherapy/ or radiotherapy. However this association remains difficult to confirm due to low incidence of this disease. Secondary PCL evolves from pre-existing multiple myeloma. Chang et al. found similar genetic abnormalities in both primary and secondary PCL. Del(17p), del(13q), del(1p21), t(4;14) and 1q21 were more frequent with PCL which are known predictors of poor outcome of multiple myeloma [13]. Rearrangement involving MYC genes are strongly associated with disease progression in multiple myeloma are seen in 33% of PPCL and SPCL which are the poor prognostic factors [14].

Diagnosis of PCL requires history, physical examination, laboratory and radiological investigation. Imaging studies include skeletal survey, bone mineral density and CT scan of abdomen, chest or pelvis. In primary PCL, IgG is the most common monoclonal protein and Bence – Jones proteinuria has also been reported. Our patient also presented with positive Bence – Jones proteinuria. CD56 (NCAM) characteristically expressed on myeloma cells is absent from patients with PCL [15]. Further, differentially expressed surface markers are CD9, CD20, CD117, and HLA-DR [5, 15]. Decreased expression of surface molecules of HLA-1, β -2M and CD40 on clonal plasma cells from patient with multiple myeloma and PCL compared to MGUS patient.

Plasma cell leukemia is highly resistant disease and prognosis is extremely poor. Response of PCL to treatment is not good. Median survival of 2-8 months is reported with M+P (Melphalan and

Prednisolone) regime or VBAP (Vincristine, Carmustine, Adriamycin and Prednisolone) regime [10]. Due to the dismal prognosis on conventional treatment, all patients who are medically suitable should be investigated for a stem cell transplant.

Conclusion

We can conclude from the case presented here that PPCL is a rare plasma cell malignancy which has a dismal prognosis unless treated aggressively with therapy akin to that used for acute leukemias. Given the rarity of the disease, we suggest that a registry of PPCL cases be commenced and there should be a universal treatment strategy to be adopted to rapidly investigate new treatment modalities.

References

1. Foa P. Sulla produzione cellulare nell infiammazione ed in altri processi analoghi specialmente in cio che si riferisce alle plasmacellule. Folia Haematol (Leipz), 1904; 1: 166–167.
2. Gluzinski A, Reichenstein M. Myeloma und Leucaemia lymphatica plasmocellularis. Wien Klin Wochensch, 1906; 19: 336–339.
3. Kosmo MA, Gale RP. Plasma cell leukemia. Semin Hematol, 1987; 24: 202-8.
4. Hayman SR, Fonseca R. Plasma cell leukemia. Curr Treat Options Oncol, 2001; 2: 202–216.
5. Garcia-Sanz R, Orfao A, Gonzalez M, et al. Primary plasma cell leukemia: Clinical, immunophenotypic, DNA ploidy, and cytogenetic characteristics. Blood, 1999; 93: 1032–1037.
6. Dimopoulos MA, Palumbo A, Delasalle KB, Alexanian R. Primary plasma cell leukaemia. Br J Haematol, 1994; 88: 754–759.
7. Bernasconi C, Castelli G, Pagnucco G, et al. Plasma cell leukemia: A report on 15 patients. Eur J Haematol, 1989; 51(Suppl): 76–83.
8. International Myeloma Working Group: Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: A report of the International Myeloma Working Group. Br J Haematol, 2003; 121: 749–757.
9. Han X., Kilfoy B., Zheng T., Holford T.R., Zhu C., Zhu Y., et al. Lymphoma survival patterns by WHO subtype in the United States, 1973-2003. *Cancer Causes and Control*, 2008; 19, 841–858.
10. Noel P, Kyle RA. Plasma cell leukemia: An evaluation of response to therapy. Am J Med, 1987; 83: 1062–1068.
11. Vela-Ojeda J, Garcia-Ruiz Esparza MA, Rosas-Cabral A, et al. Intermediate doses of melphalan and dexamethasone are better than vincristine, adriamycin, and dexamethasone (VAD) and polychemotherapy for the treatment of primary plasma cell leukemia. Ann Hematol, 2002; 81: 362–367.
12. Voutsadakis IA. Plasma cell leukemia. Haema, 2000; 3: 82-9.
13. Chang H, Qi XY, Samiee S, et al. Genetic risk identifies multiple myeloma patients who do not benefit from autologous stem cell transplantation. Bone Marrow Transplant, 2005; 36: 793–6.
14. Tiedemann et al. Primary plasma cell leukaemia. Leukemia, 2008, 22: 1044.
15. Pellat-Deceunynck C, Barille S, Jego G, et al. The absence of CD56 (NCAM) on malignant plasma cells is a hallmark of plasma cell leukemia and of a special subset of multiple myeloma. Leukemia, 1998; 12: 1977–1982.