Medical Laboratory Journal
Online ISSN: 2538-4449
Golestan University of Medical Sciences

Primary plasma cell leukemia in a 19-Year-Old female: An unusual presentation of a rare entity

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Abstract

Plasma cell leukemia (PCL) is a rare form of plasma cell dyscrasia with 2 variants: the primary form, which occurs de novo in patients with no previous history of multiple myeloma (MM), and the secondary form, which represents a leukemic transformation in patients with a previously recognized MM. Unlike myeloma, PCL typically follows an aggressive course, and the median age at presentation is usually above 50 years. In this report, we present a case of primary PCL that manifested at 19 years of age, an exceptionally rare occurrence.

Article History

Received: 20 December 2022
Received in revised form: 16 October 2023
Accepted: 24 January 2024
Published online: 25 February 2024
DOI: 10.29252/mlj.18.1.32

Keywords

Leukemia, Plasma Cell
Multiple Myeloma
Neoplasms
Female

Article Type: Case Report

Introduction

Plasma cell leukemia (PCL) is a rare yet aggressive malignancy, comprising 1-2% of all plasma cell dyscrasias (1). It can manifest either as a de novo condition (primary) or as a result of leukemic transformation in advanced multiple myeloma (MM). Plasma cell leukemia is clinically and biologically different from MM, typically diagnosed at a younger age, displaying a greater tendency for visceral and extramedullary involvement and presenting unique biologic, immunophenotypic, and cytogenetic characteristics. As a result, its accurate and timely diagnosis is crucial for appropriate management (2).

Case report

A 19-year-old female presented to the Hematology Department of the Indira Gandhi Institute of Medical Sciences in Patna, India, in May 2022, with a 1-month history of fever, headache, and weakness. Her medical and family history was unremarkable. During the clinical examination, the only notable finding was unexplained pallor that did not respond to hematinics. Her hemogram revealed the following: a hemoglobin level of 5.0 g/dL, a total white blood cell count of 14.9 x 109/L, and a platelet count of 149 x 109/L. The peripheral smear showed a microcytic hypochromic picture with Rouleaux formation, along with the presence of 36% plasma cells, including plasmacytoid lymphocytes (Figure 1a).

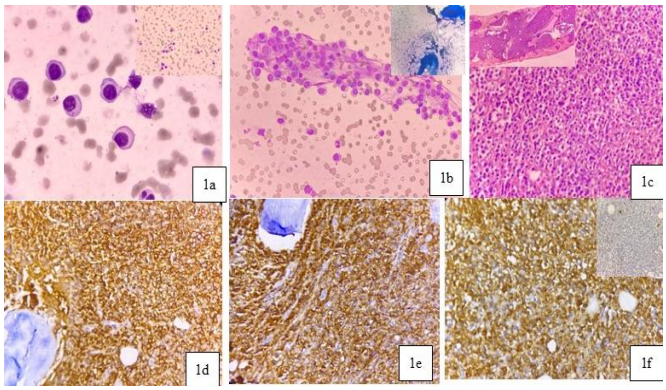

Figure 1. Microphotographs of the peripheral blood smear, bone marrow aspirate, bone marrow biopsy, along with immunohistochemistry
Figure 1a. Peripheral blood smear showing the presence of plasma cells (Leishman stain; 100X). The inset shows the Rouleaux formation (40X).
Figure 1b. Bone marrow aspirate showing diluted marrow with entangled plasma cells in fibrin meshwork (Leishman stain; 100X). The inset shows an occasional hypercellular bone marrow particle infiltrated by plasma cells (10X).
Figure 1c. Bone marrow biopsy showing hypercellular marrow with diffuse infiltration by plasma cells (H&E stain, 40X). The inset shows hypercellular marrow with plasma cell infiltration (10X).
Figure 1d. Immunohistochemistry showing CD 138 positivity (DAB; 10X)
Figure 1e. Immunohistochemistry showing CD 38 positivity (DAB; 10X)
Figure 1f. Immunohistochemistry showing kappa light chain restriction (DAB; 10X). The inset shows negativity for the lambda light chain.

Bone marrow aspiration smears were hemodiluted and showed infiltration by approximately 50% plasma cells, including abnormal forms. Many plasma cells were observed in clusters entangled in a fibrin meshwork (Figure 1b). The bone marrow biopsy was hypercellular, with findings consistent with plasma cell dyscrasia (Figure 1c). An ultrasound scan of the abdomen revealed no organomegaly or lymphadenopathy. Liver and renal function tests showed hypoalbuminemia, hyperphosphatemia, hyperuricemia, and elevated creatinine levels (5.2 mg/dL). Serum protein electrophoresis (SPE) and immunofixation studies indicated the presence of immunoglobulin G (IgG) kappa paraprotein at a concentration of 4.3 g/dL. A skeletal survey showed no osteolytic lesions.

Immunohistochemistry performed on the bone marrow biopsy showed positivity for CD38 and CD138 with kappa light chain restriction, while lambda light chain was negative (Figures 1b to 1f). Serum protein electrophoresis revealed a monoclonal M band (Figure 2).

Unfortunately, the patient succumbed to her illness before the initiation of treatment.

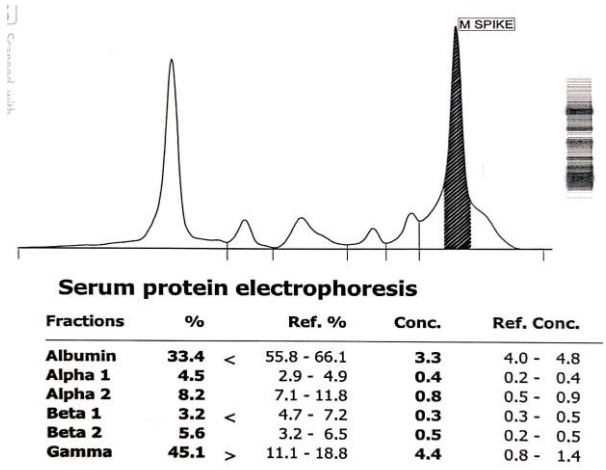

Figure 2. Serum protein electrophoresis showing a monoclonal M-band

Discussion

Plasma cell leukemia is a highly aggressive neoplasm with no consensus on a standard chemotherapy regimen due to its rarity. It comprises fewer than 1 patient per 1 000 000 population (3). It is characterized by the presence of >20% plasma cells in peripheral blood, with its incidence ranging from 2% to 4% of all myelomas (4,5). It has 2 variants - the primary form, which arises de novo in patients with no previous history of multiple myeloma, constituting 60%, and the secondary form, which consists of a leukemic transformation in previously recognized multiple myeloma, constituting the remaining 40% of total cases (6).

Phenotypically, these cells originate from the proliferation of CD38-expressing plasma cells. A minority of cells also express CD10, HLA-DR, and CD20. Primary PCL shows higher expression of CD20 compared to multiple myeloma (7). Additionally, plasma cells from both primary and secondary PCL lack CD56, which is important for anchoring plasma cells to bone marrow stroma. More than 80% have a diploid/hypodiploid DNA content, (6,8) and cytogenetic studies show complex karyotypes with multiple numerical and structural abnormalities. Up to 90% may show chromosome 13 monosomy (8).

Due to the low frequency of PCL, most of the data has come from case reports or small series of cases. In almost all the series, the median age ranged between 53 and 57 years (about 10 years younger than the median age in the myeloma series). The youngest age reported was 30 years (6).

Primary PCL follows a more aggressive course with a higher frequency of extramedullary involvement in the liver, spleen, and lymph nodes. It is also clinically more severe, manifesting as thrombocytopenia, anemia, hypercalcemia, and impaired renal function. Garcia-Sanz et al. identified ten variables with unfavorable prognostic value on the survival of primary PCL cases, of which a serum beta 2 microglobulin level > 6mg/L and S-phase bone marrow plasma cells >4.5% retained independent value on multivariate analysis (9).

Response to treatment for PCL is poor, with a median survival of less than 1 year, and the longest survival reported was 28 months (5). Failing to achieve 50% clearance of blood plasma cells within 10 days after the initiation of treatment is a predictor of no response (3,8).

Using a single alkylating agent with prednisolone is not appropriate for patients with primary PCL. Survival is significantly better in PCL patients treated with polychemotherapy compared to melphalan and prednisolone (9). Drugs used include vincristine, adriamycin, dexamethasone, and/or cyclophosphamide and etoposide (8). Alternatively, VCMP/VBAP is also used.

Since the prognosis is so poor, intensification of high-dose chemotherapy followed by allogeneic/autologous stem cell rescue should be considered (8,9).

Conclusion

This case report emphasizes its rarity, young age at presentation, and the unusual clinical and laboratory findings. Persistent anemia unresponsive to standard treatment should raise the index of suspicion and prompt further investigations aimed at excluding uncommon malignancies.

Ethical statement

Ethical clearance was obtained from the Institutional Ethical Committee with reference number (318/IEC/IGIMS/2022).

Conflicts of interest

None

Patient consent

Written informed consent was duly obtained.

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How to Cite:
Shuchismita, Jamal I, Choudhary V. Primary plasma cell leukemia in a 19-Year-Old female: an unusual presentation of a rare entity. *Med Lab J.* 2024;18(1):32-3.