Journal of Hematopathology (2019) 12:169–170
https://doi.org/10.1007/s12308-019-00349-9

LETTER TO THE EDITOR

Primary plasma cell leukaemia in a 20-year young adult male: a rare presentation

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Received: 16 November 2018 / Accepted: 25 February 2019 / Published online: 20 May 2019

Primary plasma cell leukaemia (pPCL) is a rare and aggressive variant of plasma cell neoplasm and its diagnosis is based upon the percentage (≥ 20%) and absolute number (≥ 2×10^9/L) of circulating plasma cells in the peripheral blood [1]. It accounts for about 2–4% of all plasma cell dyscrasias and the median age of presentation is 55 years, a decade younger than the average age of presentation of the other plasma cell neoplasms. In young adults, it is even rarer and only a few isolated case reports have been published to date [2–5]. We report an exceptionally rare and an unusual presentation of pPCL in a 20-year-old male from India. He had typical symptoms of the disease like peripheral blood plasmacytosis, thrombocytopenia and renal failure with no evidence of any osteolytic lesions. To the best of our knowledge, this is the first case of pPCL in a 20-year-old male to be reported from India.

A 20-year-old male was referred to All India Institute of Medical Sciences, New Delhi, with more than 2-month history of high-grade fever, abdominal discomfort, vomiting and decreased appetite. On examination, his vitals were stable and laboratory parameters showed haemoglobin (Hb) of 5.5 g/dl (normal range, 13.5–17.5 g/dl), white blood cell count of 14.2×10^9/L (normal range, 4.0–11.0×10^9/L) and platelets count of 18×10^9/L (normal range, 150–450×10^9/L). Renal function tests were found to be deranged with blood urea levels of 119 mg/dl (normal range, 7–20 mg/dl) and serum creatinine of 5.2 mg/dl (normal range, 0.6–1.2 mg/dl). Serum proteins were deranged with A/G ratio reversal—0.38 (normal range, 0.8–2.0). Urine routine microscopy suggested evidence of haematuria with 10–12 RBCs/hpf and 4–5 pus cell/hpf. He also had increased calcium levels of 11.8 mg/dl (normal range, 8.5–10.2 mg/dl) while all other routine biochemical parameters were normal. Peripheral blood smear examination demonstrated marked rouleaux formation with mild leucocytosis and numerous atypical plasma cells (30% of the differential count; Fig. 1). Bone marrow aspirate was diluted, however, showed infiltration by up to 40% plasma cells of all nucleated cells. Urine and serum protein electrophoresis revealed a sharp and dense M band 2.2 g/dl (normal range, 0.037–0.286 g/dl) in the beta region which on immunofixation was IgA lambda type. Serum-free light chain assay ratio was also deranged with $\kappa/\gamma = 0.05$ (normal range, 0.26–1.65). Beta 2 microglobulin was elevated with a value of > 10.08 mg/L (normal range, 0.7–1.8 mg/L). A diagnosis of pPCL was made based on clinical findings and laboratory parameters. He was then started on standard VCD chemotherapy regimen which includes bortezomib (2 mg, weekly), cyclophosphamide (300 mg/m^2 on days 1, 8, and 15) and dexamethasone (40 mg, weekly). After the first cycle of chemotherapy, his peripheral blood smear examinations revealed a significant reduction in plasma cells (< 10%), improvement of various haematological and biochemical parameters from baseline. His haemoglobin level increased to 8.5 mg/dl, there was reversal of renal complications with normalisation of urea (40 mg/dl) and creatinine (0.8 mg/dl) levels and improvement of serum protein levels (6.9) including A/G reversal (2.1 and 4.8). Calcium levels (7.3 mg/dl) also came down to normal range and M protein levels reduced to 0.7 g/dl. After the 3rd cycle of chemotherapy, there was significant improvement in the CBC counts and biochemical parameters and his peripheral blood smear demonstrated complete absence of plasma cells. Patient received a total of 4 cycles of VCD with further improvement of symptoms but later was lost to follow up so his response assessment and overall survival could not be commented upon.

Due to its rarity, pPCL has not been explored in detail and there are few formal studies available to date which describe

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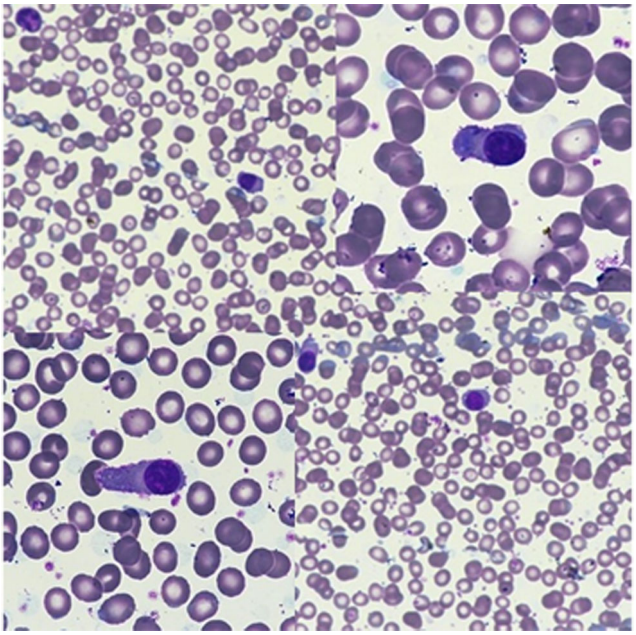

Fig. 1 Marked rouleaux formation with mild leukocytosis and numerous atypical plasma cells

its incidence and epidemiology in the general population [6]. There is paucity of literature describing the clinical presentations, morphologic variations, immunophenotype and treatment modalities of PCL both from Western countries [7, 8] and India [6, 9]. pPCL presents de novo in the leukemic phase without any prior history of multiple myeloma (MM) while secondary PCL (sPCL) shows leukemic transformation in a patient previously diagnosed with MM. Clinical signs and symptoms of PCL are similar to MM with high incidence of anaemia, lymphadenopathy, organomegaly, extramedullary involvement and renal failure and less incidence of osteolytic lesions [6].

In this case, the patient demonstrated an aggressive clinical course with typical features of the disease, i.e. severe anaemia, renal failure and lack of bone involvement. Diagnosis was based primarily on findings of peripheral blood, bone marrow aspiration, electrophoresis and immunofixation. There is no defined standard chemotherapy regimen available due to lack of prospective randomised trials on treatment of PCL. Most of the earlier cases of PCL had a very short survival time, but, with advent of novel agents like bortezomib and various other immunomodulatory agents the median survival time has significantly increased these days [7]. Our patient was treated with bortezomib-based regimen to which he responded adequately and after the first cycle, his subsequent peripheral blood smear examinations revealed a significant reduction in plasma cells with improvement of haematological parameters and normalisation of various biochemical parameters. Response assessment could not be done and a final comment on overall survival cannot be made as patient was lost to follow up after 4 cycles of VCD.

To conclude, we can say that this case highlights the clinical and haematological profile of pPCL in young adults but a definite opinion on the disease outcome cannot be made due to a relatively short follow-up. The significance of our findings in this case needs to be tested in a larger patient sample including immunophenotypic, molecular and cytogenetic investigations to unravel any relevant mechanism of pathogenesis and treatment.

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