Plasma Cell Leukemia Presenting with Hyperleukocytosis and Anaplasia

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Received: 5 September 2016 / Accepted: 13 October 2016 / Published online: 26 October 2016
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A 65-year-old female presented with NYHA grade IV dyspnoea, abdominal pain, and high-grade fever. She had anemia (Hb—103 gm/L), hyperleukocytosis (TLC— $138.5 \times 10^9/L$) and a normal platelet count ($311 \times 10^9/L$). The peripheral smear showed 42% atypical, blastoid cells, many with clefted, bilobed nuclei and conspicuous nucleoli. A few had plasmacytoid morphology and multiple coarse azurophilic cytoplasmic inclusions (panel-A and inset). The bone marrow aspirate showed 65% similar cells with more anaplasia (panel-B and inset). Staining for myeloperoxidase was negative. Flowcytometric immunophenotyping revealed a neoplastic plasma cell phenotype with a bright CD38, CD138, and monotypic surface kappa light chain expression and negativity for CD19, CD56, other B- and T-cell antigens. Immunohistochemistry on trephine biopsy (Fig. 1c, d) was positive for

CD38 and CD138 (Inset, Fig. 1d). Serum protein electrophoresis showed a prominent M-spike of IgG-kappa type and a diagnosis of Plasma Cell Leukemia (PCL) was made. Her renal functions and skeletal survey was normal. Treatment was started with bortezomib, lenalidomide and dexamethasone and she responded well with normalization of cell counts after the first cycle and is currently under treatment.

Plasma cell leukemia is a rare and aggressive neoplasm with a poor prognosis. The diagnosis can be suspected by identifying the typical plasma cells on morphology in peripheral smear. Very high leucocyte counts ($>100 \times 10^9/L$) have been occasionally reported [1]. Anaplastic plasma cells have been reported in cases of plasma cell myeloma [2, 3] but are a rarity in PCL. The present case highlights the rarity of both hyperleukocytosis

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Fig. 1 **a** Peripheral smear with atypical/blastoid cells (May-Grunwald Giemsa Stain, $\times 1000$). **b** Bone marrow aspirate showed similar cells with anaplasia (May-Grunwald Giemsa Stain, $\times 1000$). Trephine biopsy section showing hypercellular marrow spaces with diffuse

replacement of normal hematopoiesis (**c**, H&E stain, $\times 100$) by immature looking cells (**d**, H&E stain, $\times 1000$). *Inset in d* showing positive immunohistochemistry on trephine biopsy for CD38 (*left*) and CD138 (*right*) (hematoxylin counterstain, $\times 400$)

along with anaplastic morphology of plasma cells in a case of PCL mimicking acute leukemia. Flow cytometry and serum electrophoresis are invaluable for establishing the true nature of the anaplastic cells and appropriate management thereafter.

Compliance with Ethical standards

Conflict of interest The authors have no conflicts of interest to declare.

Ethical Approval All procedures performed in studies involving the human participant were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical

standards. This article does not contain any studies with animals performed by any of the authors.

Informed Consent Informed consent was obtained from the subject included in the study.

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