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A case report of plasma cell leukemia presenting as cast nephropathy

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Abstract

A 50 yr old female patient presented in nephrology department with high grade fever, hypertension, abdominal pain, hematuria, loss of appetite, weakness, palpitation, for 20 days. Then during the assessment, she was found to have a high level of BUN and creatinine along with raised serum calcium. Subsequently raised TLC with 50 percent plasmacytoid cells are seen in peripheral blood smear. Serum protein electrophoresis revealed two paraprotein bands in the gamma region. Our reported case in this paper is a primary plasma cell leukemia with unusual renal manifestation.

Keywords: Plasma cell leukemia, nephropathy

Introduction

Plasma cell leukemia is a rare form of plasma cell dyscrasia constituting 2 - 4% of malignant plasma cell dyscrasia and is diagnosed when >20% plasma cells are present in the peripheral blood, with an absolute plasma cell count of >2 × 10^9/L(1,2).

PCL presentation may be primary- de novo or secondary-evolving from an existing case of myeloma as part of the terminal phase of the disease. Primary case constitute 60 to 70 percent of all cases(3).

Because its clinical features, response to chemotherapy, and prognosis are different from those of typical multiple myeloma, primary PCL is a distinct clinicopathological entity(4). It is very important to recognize this entity sufficiently early, so that one can offer combination chemotherapy at the earliest stage followed by stem cell transplant, which can prolong patient survival.

Case report

A 50 yr old female patient presented with high grade fever, loss of appetite, weakness, palpitation, abdominal pain and hematuria for 20 days. On examination BP 160/90 mmHg, Temperature 102 F and PULSE 114/min.

High level of BUN and creatinine along with raised serum calcium (12.2mg/dl) was observed.Total protein was raised and albumin /globulin ratio was reversed. On subsequent haematological examination patient showed high WBC count (29500 cells/µl), decreased RBC count (1.7 million cells/µL), anemia (HGB 5.7g/dl), adequate PLT (143000 cells/µL), high blood calcium (11.9 mg/dl). EDTA sample was sent for CBC and peripheral blood examination.

Peripheral blood smear showed rouleaux formation and late normoblasts and in differential count revealed 50 percent mature and immature plasma cells. Bone marrow aspiration showed marked infiltration of plasma cell and precursor (52% of nucleated cells), with fine chromatin, eccentric nuclei, prominent nucleoli and perinuclear clearing. Routine urine examination showed protein (++) RBC (15-20/HPF) and tubular cast. Protein electrophoresis was advised for patient and it revealed gamma peak.

Our patient was diagnosed to have plasma cell leukemia based on the findings of peripheral smear, bone marrow aspiration and protein electrophoresis.

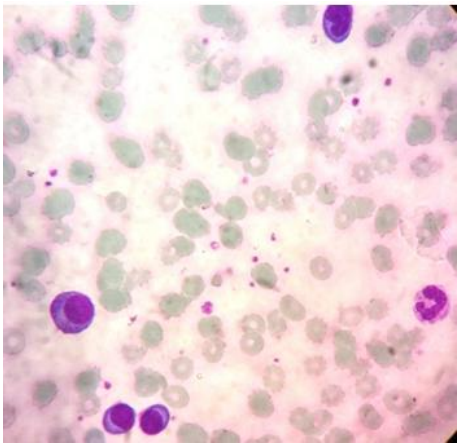

Fig. 1: Peripheral blood smear showing plasma cells and rouleaux formation, Leishman's stain, 1000x.

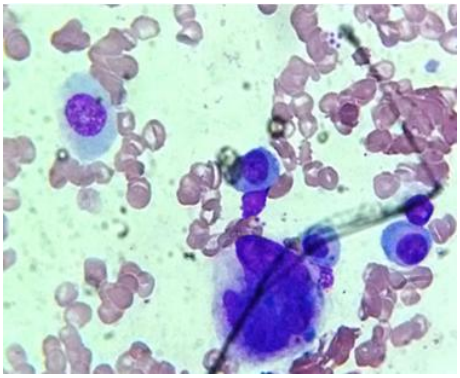

Fig. 2: Bone marrow aspiration smear showing plasma cell and one Megakaryocyte , Leishman's stain, 1000x.

Discussion

Plasma cell leukemia is a rare neoplastic disease characterized by plasma cell proliferation in the bone marrow with invasion of peripheral blood and internal organs. By definition, our patient had Plasma cell leukemia. Primary PCL is more aggressive than that of multiple myeloma. There is higher presenting tumor burden and higher frequencies of extramedullary involvement(5). Anemia, thrombocytopenia, hypercalcemia, renal impairment is common in primary PCL. Our case also presented with anemia, hypercalcemia and renal dysfunction. Rouleaux formation is usually evident in the peripheral blood smear which was also seen in our case. But in our case there was no thrombocytopenia which usually occurs in more than 50% cases of primary PCL. Leukocytosis ranges from 20 to more than 100×109/L with 20% to 100%of plasma cells. In our case, there was leukocytosis. An elevated BUN and/or creatinine occur in 75% of cases of PCL versus only 40% of cases of multiple myeloma

The incidence of lytic bone lesions is slightly lower than that usually observed in multiple myeloma(6). Most patients with PCL have a monoclonal IgG heavy chain or light chain in the serum and Bence Jones proteinuria occurs in about 80% of cases(7). Our case showed elevated M component of Gamma fraction in protein electrophoresis. Also, our case showed proteinuria.

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