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Case Report

Plasma cell leukemia

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Background

Monoclonal gammopathies comprise a wide range of entities characterized by the clonal proliferation of plasma cells (PC's) which are terminally differentiated B cells [1]. They are characterized by the secretion of electrophoretic ally and immunologically homogeneous (monoclonal) proteins [1]. When the number of circulating PCs is significant, the term plasma cell leukemia (PCL) is usually used. Whereas monoclonal gammopathy of undetermined significance (MGUS) is the most common form of PC's gammopathy, PCL is a rare entity, comprises only 2 % of patients with plasma cell neoplasm's [2].

Keywords: Plasma Cell Leukemia; Multiple Myeloma; Hypercalcemia

Case report

A 61 years old male was admitted with general weakness, abdominal pain and vomiting. On physical examination, his blood pressure was 163/89 with a regular rhythm of 86 beats per minute, 90% saturation, without dyspnea and no fever. He had normal heart sounds without murmurs, alveolar breathing, his abdomen was not tender nor stiff, without hepatosplenomegaly. No peripheral edema or a neurological deficit was found. Hemoglobin level was 13.8 gr% (normal indices), 19,370 WBCs/mm^3, 82,000 PLT/mm^3, mild basophilia 0.23x10^9/L, eosinophils level was normal, LUC was12%. Biochemistry blood work revealed mildly elevated liver enzymes- AST-141 U/L (normal range:17-59), ALT-125 U/L (normal range: 21-72) , high total protein level – 9gr/dl (normal range:6-8), globulin 6.6 gr/dl (normal range: 2.3-3.5), hyperuricemia – 9mg/dl (normal range: 3.5-8.5), hyperphosphatemia – 5.3mg/dl (normal range: 2.4-4.5), hypercalcemia – 16.5mg/dL (normal range: 8.5-10.5). Creatinine -1mg/dL (within normal range), sodium and potassium levels were normal. TSH and T4 levels were normal. Due to extremely high levels of calcium, this measurement was double checked and confirmed. Peripheral blood smear (Figure 1) showed multiple immature plasma cells - comprising greater than 20% of leukocytes. Bone marrow aspiration and biopsy (Figure 2) revealed 50% atypical young myeloma cells. Flow cytometry (FACS) showed 37% of plasma cells positive to CD138, CD38, CD27, CD20, CD31, CD28, CD29, CD34, and lambda. These findings were compatible with PCL. Protein electrophoresis revealed a pick of IgA kappa – 4 gr/dl and low levels of IgG and IgM. Serum free kappa light chain was 3864 mg/L, free Lambda light chain was 5.2 mg/L and the ratio free Kappa/Lambda was 738.76. Fluorescence in situ hybridization (FISH) examination revealed loss at p53 gene area in 10% of nuclei and loss of CDKN2C gene at 15% of nuclei. Karyotype analysis was normal. Chest X-Ray was normal. Computed tomography (CT) of the brain showed a retention cyst at the right maxillary sinus and a central occipital osseous protrusion. Spinal and pelvic CT revealed numerous skeletal lytic lesions, an extensive osteoporosis of the vertebras with L4 vertebral collapse, bilateral hip joint dislocation, with an avascular necrosis of left hip and kidney stones. The Electrocardiogram showed a sinus rhythm with a right bundle branch block, the PR interval and the QT inter-

vals were normal, without ischemic changes, without signs of left or right ventricular hypertrophy.

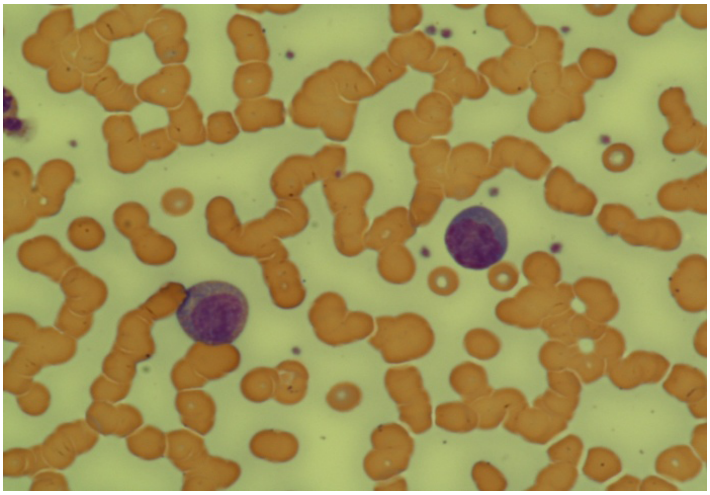

Figure 1. Peripheral blood's smear showing myeloma cells and RBC's with Rouleaux formation.

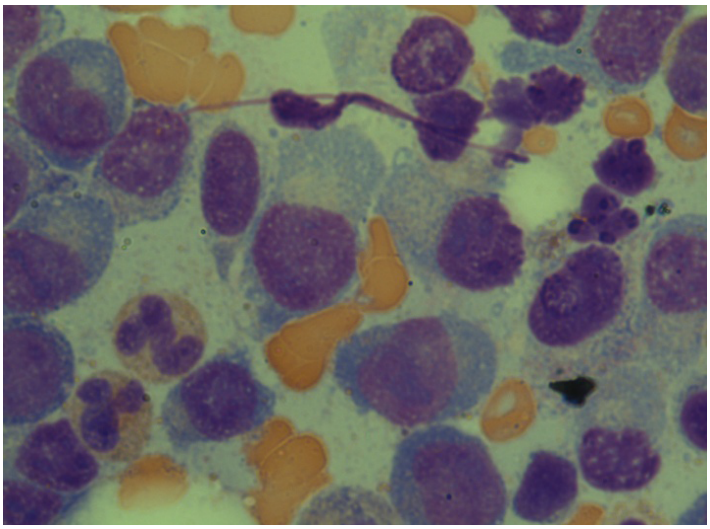

Figure 2. Bone marrow aspiration showing diffuse infiltration by plasma blast myeloma cells.

He was treated with massive hydration – 3 liters of normal saline, intravenous Dexamethasone 20 mg twice daily, Pamidronate 90 mg once daily and Bortezomibe and Cytoxan. The patient was confused at the day prior the treatment initiation, but became oriented again after one day of treatment. His calcium level decreased gradually and after one week was within the normal range. The renal function remained intact during the entire time. He is now a candidate for Hematopoietic stem cell transplantation.

Discussion

We present a patient who was admitted with general weakness and vomiting, had numerous skeletal lytic bone lesions and high calcium level. Peripheral blood smears showed 3874 plasma cells/μL and the bone marrow examination revealed a plasmatic infiltrate with 50% atypical young myeloma cells. In order to obtain uniform criteria for the diagnosis of plasma cell leukemia (PCL) Kyle et al [3] proposed an absolute plasma cells count greater than 2000 plasma cells/μL and more than 20% of the WBCs in peripheral blood.

Primary plasma cell leukemia (pPCL) is defined as a malignant plasma cell proliferation first diagnosed in the leukemic phase, while secondary plasma cell leukemia (sPCL) describes a leukemic transformation of a previously diagnosed multiple myeloma probably as a consequence of clonal transformation [1]. The median time to leukemic transformation for patients with multiple myeloma who transform into sPCL is 21 months [4].

In our patient, according to these diagnostic criteria and in the absence of pre-existing multiple myeloma - the diagnosis was determined to be the rare entity of primary PCL. Median age of diagnosis of pPCL is 55, a decade earlier than the average age of diagnosis of multiple myeloma. The hypercalcemia that was his presenting sign, and fatigue (the leading clinical symptom) - were probably secondary to the extensive metastatic bone lesions.

Patients with PCL are treated with aggressive induction therapy (including alkylating agents) followed by hematopoietic stem cells' transplantation. Chemotherapy alone is the principal option for patients who are ineligible for stem cells transplantation [5].

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