

## Primary Plasma Cell Leukemia Presenting with Chest Pain

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### Abstract

The term plasma cell leukemia (PCL) is the most aggressive form of the plasma cell dyscrasias and it's used when the number of circulating plasma cells is significant. It's defined by the presence of  $>2 \times 10^9/L$  peripheral blood plasma cells or accounting for  $>20\%$  of the differential white cell count, that does not arise from preexisting multiple myeloma.

**Keywords:** Plasma cell leukemia; Myeloma; Chest pain

### Letter to Editor

Plasma cell leukemia (PCL) is a rare and aggressive form of plasma cell dyscrasias and it's used when the number of circulating plasma cells is significant. It is defined by an absolute plasmacytosis of greater than  $2 \times 10^9$  cells/L or greater than 20% plasma cell in the peripheral blood [1]. PCL is classified as primary when it presents "de novo" in patients with no evidence of previous multiple myeloma and as secondary when it is observed as a leukemic transformation of relapsed or refractory diseases in patients with previously recognized multiple myeloma.

A 48-year-old Caucasian male with no significant past medical history was referred to our hospital because left side chest pain for four months. He had no exposure to environmental toxins. On physical exam, he was afebrile and he had no palpable adenopathy or hepatosplenomegaly. Chest X-ray revealed a lytic lesion in the 8th left rib. Initial laboratory investigations revealed the following: white blood cell count  $61.3 \times 10^9/L$  with 3% of plasma cells, hemoglobin 8.7 g/dl and normal platelet count. Beta-2 microglobulin and LDH were elevated (5.2  $\mu\text{g/mL}$  and 717 U/L respectively). Peripheral blood smear showed red blood cell rolls and atypical appearance of white blood cells. A serum protein electrophoresis was performed that demonstrated an IgG lambda monoclonal protein (2.630 mg/dL). A monoclonal peak of 1.57 g/dL was seen in the serum. Urinary analysis showed immunoglobulin free light chains Kappa (Bence-Jones protein). Bone marrow biopsy revealed a diffuse infiltration of atypical plasma cells (85%) lacking blastic features. Flow cytometry immunophenotypic characteristics were as follows: positive for CD38, CD138, CD27, and negative for CD19, CD56, CD117. Patient was referred to the department of hematology where he received chemotherapy according to the protocol Dexamethasone, Lenalidomide and Bortezomib.

Plasma cell leukemia is a rare and aggressive plasma cell disorder. First cases of PCL were described at the beginning of the twentieth

century [2]. It's a different clinic-pathologic entity from myeloma multiple: PCL is observed in younger patients (median age ranged between 52 and 65 years), more common in African Americans than in Caucasians [3], with a increased incidence of urinary free light-chain (Bence-Jones). PCL is characterized by an aggressive clinical presentation, rapid clinical course, leukocytosis, extramedullary involvement, marked bone marrow infiltration by immature plasma cells and high LDH serum levels [3]. Evaluation of the plasma cells in patients with PCL demonstrates differences in marker expression between multiple myeloma and secondary plasma cell leukemia: C38 and CD138 are expressed in both, but CD56; characteristically expressed on myeloma cells, is absent from patients with PCL [4]. Combination therapy of alkylating agents and glucocorticoids are unsatisfactory. Nowadays, effective induction therapy is based in intensive chemotherapy with alkylating agents including anthracyclines such as HyperCVAD (hyper-fractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone) or PACE (cisplatin, doxorubicin, cyclophosphamide and etoposide) or bortezomib-based combinations [3]. Otherwise, in patients younger than 50 years of age with a suitable donor, a myeloablative allogeneic transplantation can be considered.

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