"A RARE CASE OF PRIMARY PLASMA CELL LEUKEMIA WITH RESPIRATORY INVOLVEMENT"

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INTRODUCTION

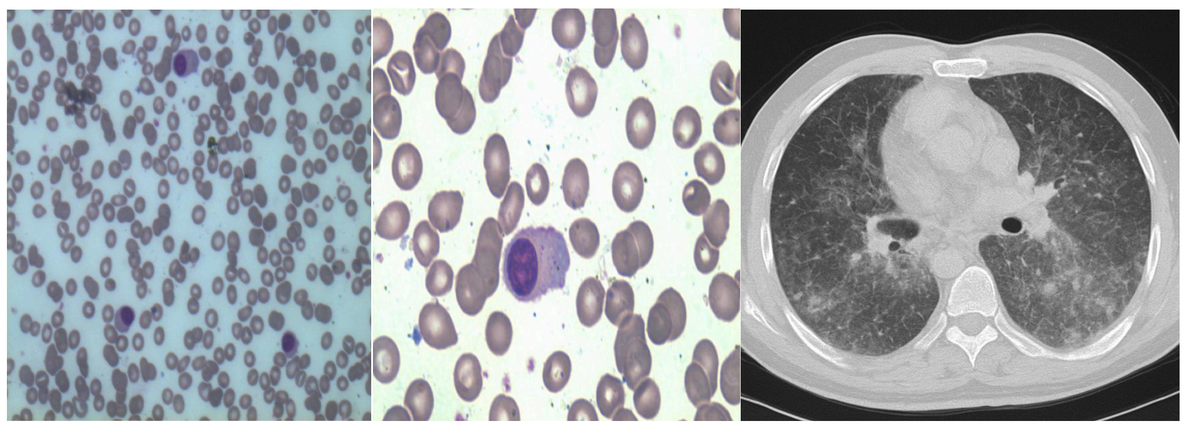
- Plasma cell leukemia (PCL) is a rare aggressive variant of multiple myeloma.
- PCL is diagnosed when
– clonal plasma cells constitute more than 20 % of the total circulating leukocytes, or
– when the absolute plasma cell count exceeds 2 × 10^9/L
- Soft tissue involvement is rare.

CASE HISTORY

- A 64-year-old male
- a chronic hukka smoker
- non-diabetic and non hypertensive
- Presented with 3-month history of:
– upper abdominal pain,
– weight loss,
– productive cough and
– Exertional breathlessness
- O/E: afebrile, PS-III, Pulse 82/min, BP 122/78, RR 18, Liver 13 cm,spleen – tip palpable, BS- B/L wheeze

INVESTIGATIONS

- Hb 3.5%, total leukocyte count 41,360/mm³,platelet count 71,000/mm³,ESR 95 mm/hr
- AFB- sputum negative, LDH – 1366
- Peripheral smear- 20% plasma cells, rouleux formation
- bone marrow aspiration revealed 70% plasma cells
- bone marrow biopsy confirmed 60% plasma cell infiltration.
- Increased β2-microglobulin (11.15 µg/mL)
- Monoclonal IgG kappa M-band was seen on
– Serum and urine electrophoresis
– Pleural fluid analysis also showed M- band
- Free kappa/lambda ratio of 111.11
- Imaging (CECT) revealed
– bilateral pleural effusion (more on the left),
– ground-glass opacities,
– ill defined nodular opacity with mediastinal LN largest – 16x12 mm,
– mild paraseptal emphysematous changes.
- The overall findings were consistent with plasma cell leukemia.
- The patient received six cycles of PAD chemotherapy
– Bortezomib, Doxorubicin, and Dexamethasone
- Significant clinical and hematologic improvement was seen
- M-band disappeared
- Maintenance therapy with Bortezomib and Dexamethasone was initiated
- Autologous stem cell transplant was advised



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- **Figure1&2: PBF showing abnormal plasma cells, Giemsa stain (20x &100x respectively)**
  Figure 3: CECT Chest showing ground glass opacities with bilateral pleural effusion

DISCUSSION

- PCL is a rare, aggressive form of plasma cell disorders, accounts for 0.5-3% of plasma cell disorders. [1]
- Clinically, PCL patients are diagnosed at a median age younger by a decade than MM patients [2,3]
- PCL tends to present more with extramedullary involvement than MM such as the liver, spleen, body cavities (pleura, pericardium, and peritoneum), and spinal cord. [3,4,5]
- PCL is characterized by decreased hemoglobin levels, cytopenia, hypercalcemia, renal insufficiency, and an increase in Lactate dehydrogenase and β2-microglobulin[2,4,5]
- Serum and urine protein electrophoresis with immunofixation should also be obtained to identify a monoclonal immunoglobulin, with the IgG subtype being the most common in PCL.
- This parallels our patient's investigations.
- Chemotherapy regimens commonly used in the treatment of PCL include bortezomib-based combinations, such as VAD (vincristine, doxorubicin,and dexamethasone) or CyBorD(cyclophosphamide, bortezomib, and dexamethasone)[6]
- Autologous stem cell transplantation (ASCT), may be considered for eligible patients with PCL. ASCT has shown promise in extending survival in PCL patients [7]

CONCLUSION

- This case report highlights the rare occurrence of soft tissue involvement in plasma cell leukemia (PCL), as well as the aggressive nature of the disease.
- The patient presented with extramedullary manifestations, pleural effusion, ground-glass opacities, nodular infiltrates, and mediastinal lymphadenopathy demonstrating the diverse clinical manifestations of PCL.
- This case underscores the importance of considering PCL as a differential diagnosis in patients presenting with unusual soft tissue involvement.

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