Plasma cell leukemia presenting as acute renal failure : a case report

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Abstract: *Plasma cell leukemia is a rare form of malignant plasma cell dyscrasia. It can occur as a primary form without prior evidence of multiple myeloma or as a secondary form which is a terminal event in multiple myeloma. A case of primary plasma cell leukemia presenting as acute renal failure is reported here.*

Key Words: primary plasma cell leukemia, acute renal failure

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Introduction

Plasma cell leukemia (leukemic myelomatosis) is the malignant clonal proliferation of B cells, the incidence of which ranges from 1.6 to 5% reported in different series.[1-3] The criteria for diagnosis of plasma cell leukemia is presence of more than 20% of plasma cells in peripheral blood smear or absolute plasma cell count greater than $2x10^9$/L. [1,3,4]

Plasma cell leukemia (PCL) can occur as primary plasma cell leukemia to denote de novo appearance of PCL without preceding multiple myeloma (MM), the exact incidence of which is not known. Howerer it is believed to be less than 1 case per million.[4] Secondary PCL is very rare and occurs as a terminal event in MM. It develops in 1-2% cases of MM.[1]

Case Report

A 50 year male, presented with fever, weakness, loss of appetite and weight for 15 days. Patient also had vomiting, diarrhoea and oliguria since 2 days. On examination patient had tachycardia, soft and tender hepatomegaly and neck stiffness. Kernig's sign was negative. Provisional diagnosis of acute renal failure with uremic encephalopathy was made.

Hematological investigation revealed hemoglobin of 5.8 g/dl, total leukocytic count $36.2x10^9$/L, platelet count $31x10^9$/L and ESR 100 mm 1st hour by Westergren's method. Peripheral blood smear showed bluish background with rouleux formation, normocytic hypochromic anaemia, thrombocytopenia and differential count showed neutrophils 55%, lymphocytes 06% and plasma cells 39% (Fig. 1A). Plasma cells showed variable maturity. Biochemically blood urea was 190 mg/dl, serum creatinine 8.3 mg/dl, total protein 11.1 g/dl, albumin 1.6 g/dl, globulin 9.5 g/dl, A:G ratio 0.16, blood calcium 7.5 mg/dl and urine calcium 10.5 mg/dl. Urine showed specific gravity of 1.020, protein ++ and Bence Jones protein was positive . CSF analysis was within normal limits. Bone marrow aspiration showed high cellularity with plasma cells of 77%. Plasma cells were of varying maturity consisting of mature / immature cells and binucleate / trinucleate forms (Fig. 1B). Megakaryocytes were decreased. Abdominopelvic ultrasound showed mild hepatomegaly and minimal ascites. X-ray skull showed multiple punched out osteolytic lesions in occipital

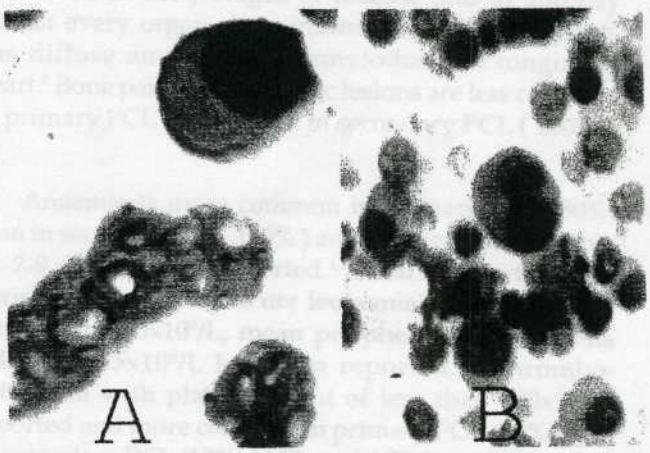

Fig. 1A. Photomicrograph of peripheral blood smear showing plasma cell and rouleux formation. 1B Marrow aspiration smear showing plasma cells.

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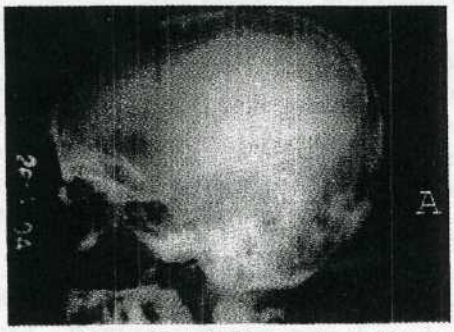

Serum Protein Electrophoresis

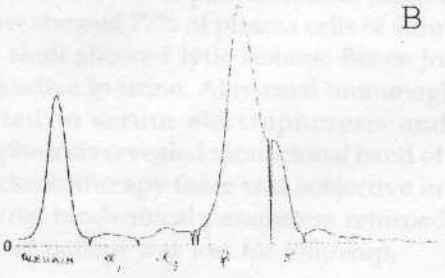

Fig. 2A. Photograph of X-ray skull showing lytic lesion in occipital region. **2B.** Serum electrophoresis showing 'M' spike in beta region.

region (Fig. 2A). X-ray spine and pelvis were within normal limits. Serum electrophresis showed 'M' spike in beta region, 'M' spike was 5.74 g/dl and other protein fractions were albumin 2.4 g/dl, alpha 1 - 0.3 g/dl, alpha 2 - 0.4 g/dl, beta 7.1 g/dl and gamma 0.4 g/dl (Fig. 2B). Serum immunoeletrophoresis revealed monoclonal band of IgG kappa. Hence final diagnosis of primary PCL with acute renal failure was made. Patient was put on cyclophosphamide, prednisolone and decadron. With adequate hydration and supportive treatment, renal biochemical parameters returned to normal over a period of 2 weeks. Patient was discharged and advised to come once in a month for pulse chemotherapy. Patient was lost for follow up.

Discussion

Plasma cell dyscrasia are a group of disorders having the proliferation of a single clone of immunoglobulin secreting cells as a common feature. The incidence of various plasma cell dyscrasia is as follows: Monoclonal gammopathy of undetermined significance – 60-70%; MM-15%; Amyloidosis – 9%; B cell lymphoproliferative disorders : NHL- 5%, WM- 2%, CLL- 2%; Solitary plasmacytoma- 1%; Plasma cell leukemia- Rare.[5]

The concept of PCL began with the first case report by Foa in 1904 in a woman.[1,6] Only three cases in Indian literature and 67 cases in western literature were reported between 1965 and 1986.[5] PCL is a rare neoplastic plasma cell disease and primary PCL is still rare, the incidence of which is not known and very few cases has been reported in literature.[1]

Primary PCL commonly occurs in individuals between 50 and 60 years of age groups with male prepondarance.[1,2] The duration from onset of symptoms to diagnosis varies from a few hours to 8 months in primary PCL in comparison with 16.5 months in secondary PCL.[1,2] The presenting symptoms are weight loss, weakness, hemorrhagic manifestation and easy fatiguability.[3,4] Hepatomegaly, spleenomegaly and lymphadenopathy are reported in 75%, 53% and 40% of cases of primary PCL respectively and are more common than in secondary PCL.[1,3,5] Leukemic infiltration by PCL has been reported in lung, pleura and testis in early stages of disease and at autopsy almost every organ show infiltration.[1,3] Some patients has diffuse amyloiosis or amylodosis of tongue or heart.[1] Bone pain and osteolytic lesions are less common in primary PCL (50%) than in secondary PCL (100%).[1,3]

Anaemia is more common in primary PCL (80%) than in secondary PCL (35%) and the mean hemoglobin of 7-8 g/dl has been reported.[1,3] Total leukocytic count varies similar to any acute leukemia from normal to more than $250x10^9$/L, mean peripheral blood plasma cells of $32.5x10^9$/L has been reported.[1-3] Thrombocytopenia with platelet count of less than $100x10^9$/L reported and more common in primary PCL (50%) than in secondary PCL (10%).[1,3] Elevated BUN and creatinine occurs more common in primary PCL (75%) than in secondary PCL (40%).[1] Hypercalcemia is also reported. Electrophoretic abnormality in serum is reported in 90% of PCL cases.[1] 'M' protein in serum is reported in 50% cases of primary PCL and 80% in secondary PCL.[1] Serum immunoeletrophoresis analysis have shown IgG monoclonal component as most common (50-80% cases) abnormality followed by IgM and least common being IgA, IgD and IgE.[1-3] However, some cases have hypogammaglobulinemia, and others normal serum proteins (non secretory type).[1,6] Bence Jones proteinuria reported in 75% cases of primary PCL and urine globulin spike reported in 85% cases of primary PCL compared to 100% in secondary PCL.[1,3] Urine eletrophoresis showing predominanace of lambda light chain has been reported.[3] Renal function is impaired as a result of hypercalcemia or Bence Jones proteins or both.[6] The renal failure is more common in primary PCL than in secondary PCL.[4] Bone marrow aspiration is necessary for diagnosis of PCL, even if clinical, biochemical and radiological evidence is convincing.

Primary PCL are treated with chemotherapy, irradiation, autologous bone marrow transplantation

and Interferon.[5] The prognosis and result of treatment in PCL are quite discouraging.[3] However, the patients with primary PCL respond initially to chemotherapy unlike secondary PCL which responds rarely.[4] Median survival in primary PCL is usually less than three months compared to 33 months in secondary PCL.[2] Infection and hemorrhage contribute significantly to mortality and morbidity.[1,6]

In our case, the patient is 50 years male with acute onset of symptoms, hepatomegaly and signs of renal failure having anaemia, thrombocytopenia, leukocytosis and 39% plasma cells in blood smear. Bone marrow showed 77% of plasma cells of varing maturity. X-ray skull showed lytic lesions. Bence Jones protein was positive in urine. Abnormal immunoglobulin was detected in serum electrophoresis and immuno-eletrophoresis revealed monoclonal band of IgG kappa. With chemotherapy there was subjective improvement and renal biochemical parameters returned to normal. However patient was lost for followup.

To summarise, plasma cell leukemia should always be considered in patients with leukemia in whom the circulating cells are suggestive of plasma cells. Early diagnosis and classification of disease is important to plan the treatment and assess the prognosis. However poor response to treatment and short survival in PCL stress the need to develop new therapeutic approaches.

References

1. Kamla Gupta, Chopra BK, Kanchan Saini, Bhardwaj BL. Primary plasma cell leukemia – a case report. *JAPI* 1996; 44 (1) :53–55 .
2. Neelam Varma, Alok Jain, Neelam Marwaha, Shobha Sehgal, Mehta SK. Primary plasma cell leukemia : a case report and review of literature. *Indian J Cancer*1988; 25:114–6.
3. Arora S, Nair CN, Gopal R, Saikia, Nadkaini KS, Das Gupta A, Advani SH. Primary plasma cell leukemia. *JAPI*1987; 35(10): 733–5.
4. Sunita, Gupta DK, Saluja S, Sharma M, Bharin S, Narang M. Secretory and non-secretory primary plasma cell leukemia – report of two cases. *Indian J Hemat & Blood Transf* 2003; 21(4): 144–5.
5. Desai SR, Angarkar NN, Kulkarni AG. Primary plasma cell leukemia. *JAPI*2004; 52:510–11.
6. Bhawana A Badhe, Debdatta Basu, Pampa ch Toi, Dutta TK, Ghotekar LH. Plasma cell leukemia – a case report. *Indian J Pathol Microbiol*2003; 46(3):484–7.