

## Acute Plasma Cell Leukemia Associated with Bence-Jones Proteinuria. A case Report

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### Introduction

Acute plasma cell leukemia is an extremely rare reticuloendothelial disease of unknown cause. It is manifested by the presence of a large number of immature plasma cells in the peripheral blood and bone marrow which extensively infiltrate other organs of the body such as the spleen, liver and lymph nodes. Average survival from the time of diagnosis of patients suffering from plasma cell leukemia is 8 months; where as patients with multiple myeloma survive 2 to 3 years. The purpose of this communication is to report on a patient with plasma cell leukemia associated with the uncommonly associated light-chain paraproteinuria.

Case report: A 60 year old Iranian male truck driver was admitted to the Tehran University Department of Medicine, Pahlavi Medical center, with a 25 days history of fever, generalized weakness, weight loss and crampy abdominal pain associated with diarrhea and melaena. The past history was unremarkable except for low back pain of 8 years duration attributed to osteo-arthritis. There was also a history of chronic gastritis of several years duration. The family history was unremarkable. The patient was married and was the father of 4 healthy children.

Physical examination showed a well developed, though weak, wasted and pale man. The patient was afebrile. The skin was pale and showed purpuric and petechial lesions over most of the body. There were generalized superficial lymphadenopathy. The spleen was palpable at 5 cm below the left costal margin. The eye grounds showed microhemorrhages and melaena was noted on rectal examination. Laboratory findings included a hemoglobin of 7 gm %, red blood cell count of 2 millions cumm, total peripheral white blood cell count of 53,000 cumm. The differential WBC count showed that 80 % of the cell were very immature looking plasma cells, the platelet count was 10,000 cumm, and the sedimentation rate was 100 mm/1 hour. Blood urea nitrogen was 128 mg %, serum uric acid 14.8 mg %, calcium 13 mg %, phosphorus 4.3 mg %, alkaline phosphatase was 6.6 K-A units. Serum protein electrophoresis showed 53 % albumin, 5 % alpha 1, 9 % alpha 2, 13 % beta 1, 15 % beta 2 and 5 % gamma globulin. Urine immunoelectrophoresis showed Bence-Jones (lightchain) protein.

The total 24 hour urinary output was only 250cc and microscopic urinalysis showed many casts and increased white blood cells. Examination of the urine for Bence-Jones protein was strongly positive. Stool examination for occult blood was positive. A radiological skeletal survey showed no osteolytic lesions and was unremarkable except for a mild generalized osteoporosis. Bone marrow examination revealed a grossly hypercellular marrow consisting entirely of immature and abnormal plasma cells.

The hospital course was brief and stormy and the patient expired nine days after admission of acute hemorrhage and renal failure.

At autopsy, acute hemorrhage was noted in the gastro-intestinal tract. There was generalized enlargement of the lymph nodes and spleen. Histology studies showed massive plasma cell infiltration of the liver, spleen, lymph nodes, bone marrow, gastro-intestinal mucosa and lungs.

### DISCUSSION

Acute plasma cell leukemia is one of the rarest forms of leukemia (1). Up to 1969 there were only 56 cases reported in the literature (2). The rarity of this leukemia was recently referred to at the International Congress of Hematology held in August 1960 in Munich (2).

Among the cases of acute plasma cell leukemia reported, there have been very few cases with light chain paraproteinuria (4). In contrast to multiple myeloma which occurs more commonly in males, plasma cell leukemia occurs equally in both sexes. Classically, plasma cell leukemia presents as a systemic disease with widespread leukemic infiltration of all organs. The peripheral blood and bone marrow picture is similar to that of any other acute leukemia, and there are no osteolytic lesions of the skeleton on radiological examination (5).

Several of the cases reported were described as having had intra- or extra — osseous plasmacytomata as well (6). If these cases are excluded it becomes apparent that true plasma cell leukemia occurs very rarely. The association of light- chain paraproteinuria is even more uncommon. The present case is believed to represent a classical case of plasma cell leukemia with Bence-Jones proteinuria. The patients reported in the literature have ranged in age from 1/5 to 78 years, but most of them have been between 45 and 60. The mean duration of life following recognition of this disorder has been 8 months, while that of multiple myeloma is 2-3 years (6).

In comparison to most leukemia, this particular type has a poorer prognosis and is less responsive to chemotherapy (2).

In the literature a few cases have been reported of plasma cell infiltration in many organs of the body with light-chain proteinuria and no abnormal radiological findings in the bones. Myeloma cells were not seen in peripheral blood, however, these cases are probably preleukemic and represent a stage intermediate between multiple myeloma and plasma cell leukemia (3).

### SUMMARY

Acute plasma cell leukemia with Bence-Jones proteinuria is reported in a 60 year old Iranian male with a 25 day history of acute onset of fever, weakness, weight loss, diarrhea and bloody stools. The patient was noted to be cachectic and anemic. He had purpuric and petechial skin lesions, generalized lymphadenopathy and splenomegaly. Up to 80% immature plasma cells were present in the peripheral blood and the platelet count was 10,000. Bone marrow was hypercellular and that most of it was composed of immature plasma cells. Serum electrophoresis showed increased beta globulins and Bence-Jones protein was strongly positive in the urine. The patient died after nine days in uremic coma with haemorrhagic diathesis. Autopsy showed wide spread infiltration of plasmocytes and plasmocytoblasts in all organs.

## RESUME

Le cas d'une leucémie aigue plasmocytaire avec la proteine Bence-Jones dans les urines, chez un homme iranien de 60 ans, est presenté avec une histoire a debut brusque et d'une évolution de 25 jours, avec fièvre, asthenie, amaigrissement, diarrhée et meloena. Le malade était anémique et cachectique; il avait des lesions cutanées de type purpurique et pétéchiial; des adénopathies generalisées et une splenomegalie.

Dans la formule sanguine on a trouvé 80 % des plasmocytes immatures et 10,000. plaquettes; la moële osseuse etait hyperactive et entierement infiltrée par des plasmocytes immatures.

L'electrophorese du serum a montré l'augmentation des béta globulines; dans les urines la proteine de Bence-Jones était fortement positive.

Lê patient après 9 jour de coma urémique, avec une diathèse hemorrhagique, est decédé.

A l'autopsie: on a trouvé dans toutes les organes des larges etendues d'infiltrations plasmocytaires et plasmoblastiques.

## References

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