Case Report

An Unusual Presentation of Plasma Cell Leukemia with Undiagnosed Multiple Myeloma

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ABSTRACT

A 54 year old man with history of cardiomyopathy and chronic fibrillation first presented with bloody diarrhea. Then during the assessment of his abnormal lab. data revealed cast nephropathy in renal biopsy, gamma peak in protein electrophoresis and fine lytic lesions in skull graphy.

His bone marrow aspiration showed multiple myeloma and blood smear revealed plasma cell leukemia. Our patient was diagnosed to have multiple myeloma and plasma cell leukemia

Plasma cell leukemia is a rare form of plasma cell dyscrasia.The second type evolves as a terminal event in some of the patients with multiple myeloma. PCL and multiple myeloma simultaneously recognizes in this case(very rare event).

Key words: Plasma cell leukemia, Multiple myeloma

Introduction

Plasma cell leukemia (PCL) is a rare form of plasma cell dyscrasia characterized by the presence of more than 20% plasma cells in peripheral blood and an absolute plasma cell count more than 2 $\times 10^9$/L (1). PCL can be classified into two types. The primary form presents de novo in patients with no previous history of multiple myeloma and usually features a rapid clinical progression and a short survival. The second type evolves as a terminal event in 12% of cases of multiple myeloma (2).

Because its clinical features, response to chemotherapy, and prognosis are different from those of typical multiple myeloma, primary PCL is a distinct clinicopathological entity. It is very important to recognize this entity sufficiently early, so that one can offer combination chemotherapy at the earliest stage followed by stem cell transplant, which can prolong patient survival (3). Son des et al reported 2 cases of plasma cell leukemia in July 2005(4). Desai et al also reported a 90-years male case with primary plasma cell leukemia in June 2004 in India (5).

Our reported case in this paper is a 54-years old man that had secondary PCL. PCL and multiple myeloma were simultaneously recognized in this case (a very rare event).

Case report

A 54-years old man with a history of cardiomyopathy and chronic fibrillation, first presented with bloody diarrhea, then converted to non-bloody, and after a few weeks got malaise and lightheadness and oliguria. During workup he was found to have a high level of BUN and creatinine (BUN = 140, Cr=4.5).

Patient admitted to Khorshid hospital in Isfahan on 9 Oct 2005 for renal failure.

His renal scan showed renal failure and a decrease in function and blood flow of two kidneys, more

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pronounced in right kidney. On physical examination, patient had a mild spelenomegaly. Laboratory data showed low platelet (103,000/µl), high white blood cells (25,100/µl), anemia (Hb= 8.9 g/dl), high blood calcium (10.2 mg/dl), and allbuminuria following urine analysis. Renal biopsy showed cast nephropathy (Figure 1). After 5 days of hospitalization, patient indicated for renal dialysis and then dialysed for several times. In the course of hospitalization, patient admitted to CCU two times for heart problems (rapid response, atrial fibrillation, and ventricular tachycardia on the base of his cardiomyopathy following blood infusion.

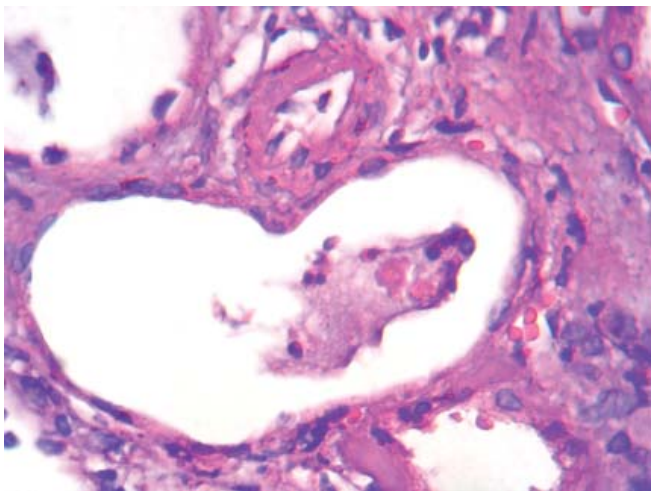

Figure 1. Typical cast within tubule with giant cell (H & E,×400)

After report of cast nephropathy in biopsy, protein electrophoresis was done for patient and it revealed gamma peak. By skull graphy, some fine lytic lesions were seen (Figure 2) and peripheral blood smear showed late normoblasts and rouleaux formation and in differential count revealed 63% immature plasma cells (Figure 3).

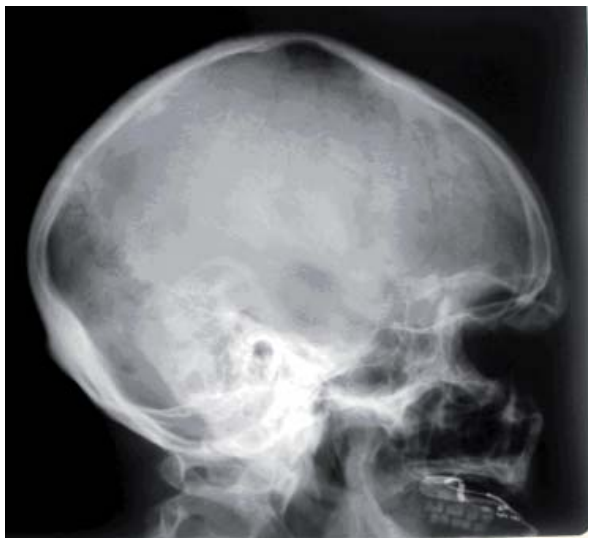

Figure 2. Fine lytic lesions of skull

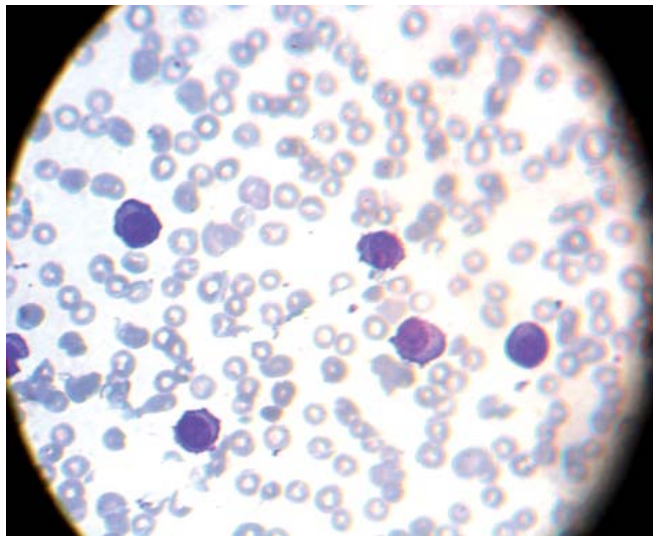

Figure 3. Immature plasma cells in peripheral blood smear (×400)

Bone marrow aspiration showed marked increase in cellularity and infiltration of immature cells and 90% of nucleated cells with fine chromatin and prominent nucleoli and eccentric nucleolus and perinuclear clearing (Figure 4).

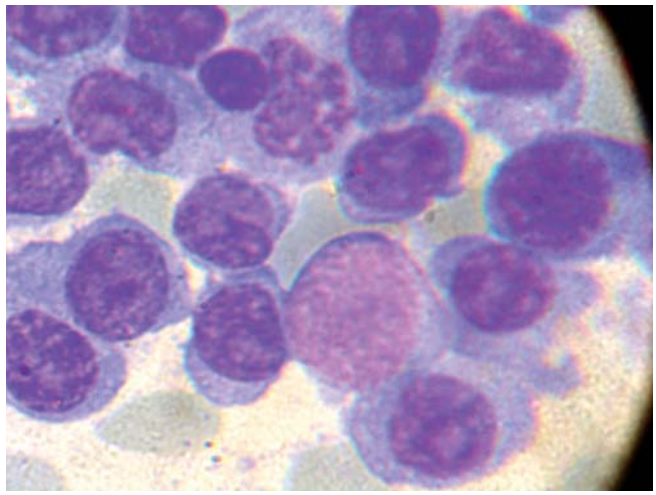

Figure 4. Immature plasma cells proliferation in bone marrow aspiration (×1000)

Our patient was diagnosed to have multiple myeloma and plasma cell leukemia based on the findings of peripheral smear and bone marrow aspiration. This report is unique for the concurrent diagnosis of multiple myeloma and plasma cell leukemia in a patient.

Patient hospitalized in Al-zahra hospital on 26 Oct 2005 for daily plasmapheresis and expired after 2 weeks.

Discussion

Plasma cell leukemia (PCL) is a rare neoplastic disease characterized by plasma cell proliferation in

the bone marrow with invasion of peripheral blood and internal organs (6). By definition, our patient had PCL. This condition can be considered as the leukemic variant of multiple myeloma. Its incidence ranges from 2% to 4% of all myelomas (2, 7, 8). There are two forms of PCL. The primary form arise de novo in patient with no previous incidence of multiple myeloma (which constitutes 60% of cases) and secondary form which consists of a leukemic transformation in previously recognized multiple myeloma (6). Hepatosplenomegaly and lymphadenopathy are more common in primary than in secondary plasma cell leukemia.The lytic bone lesions are more common in patients with secondary plasma cell leukemia (100% versus 60%) (9). Our case had splenomegaly and few lytic bone lesions. Anemia with hemoglobin level less than 9 mg/dl occurs in 80% of cases of plasma cell leukemia versus 35% of cases of multiple myeloma (9). In our case, the hemoglobin was low i.e. 8.9 mg/dl. Rouleaux formation is usually evident in the peripheral blood smear which was also seen in our case.

Thrombocytopenia with platelet count less than 100×10^9/L occurs in 50% of patients with PCL versus only 10% of those with multiple myeloma. Leukocytosis ranges from 20 to more than 100×10^9/L with 20% to 100% of plasma cells. In our case, there was leukocytosis. Our case showed moderate thrombocytopenia. An elevated BUN and/or creatinine occur in 75% of cases of PCL versus only 40% of cases of multiple myeloma (9). Values of these investigations had elevated levels in our case.

Most patients with PCL have a monoclonal IgG heavy chain or light chain in the serum and Bence Jones proteinuria occurs in about 80% of cases. Our case showed elevated M component of Gamma fraction in protein electrophoresis. Also, our case showed proteinuria. Primary PCL has a rapid course with short survival, whereas the secondary form may be associated with a more indolent clinical course and survival is variable (10).

About cardiac problems in our patient, it can be associated with his other disease (PCL). There are some reports about occurrence of PCL and some cardiac disease like restrictive cardiomyopathy and pericardial effusion simultaneously in a patient (11.12).

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