CASE REPORT

A 52 years old female presented with severe respiratory distress due to recurrent pleural effusion for 3 months. Physical examination revealed severe pallor, bilateral pedal edema, tachycardia, tachypnoea, bilateral diminished breath sound, hepatosplenomegaly and bony tenderness in the back. There was no history of fever and jaundice. The patient was normotensive and non-diabetic. She did not receive any blood transfusion. There was no significant past history or family history.

Haematological investigation showed normocytic normochromic anaemia (Haemoglobin - 6 g/dl) with increased total leucocytic count (37900/mm³). Differential leucocytic count showed neutrophilic predominance. Atypical plasma cells were seen in peripheral smear constituting 28% of the white blood cells. [Figure-1A] Platelet count was reduced(43000/mm³) and ESR elevated (86 mm in 1 hour).

Bone marrow examination revealed cellular marrow with depressed trilineage haemopoiesis. Plasma cells are increased in number and constitute about 46% of the nucleated marrow cell population. Most of them were atypical, bi and tri-nucleated forms and some of the cells showed lympho-plasmacytoid appearance [Figure-1B].

Blood biochemical investigation findings were as follows- Fasting blood sugar – 80 mg/dl, serum urea – 41mg/dl, serum creatinine – 1mg/dl, serum calcium – 8.3 mg/dl, corrected serum calcium-9.6mg/dl, LDH – 461 IU/L (Normal – 180 to 360 IU/L), total protein – 8 gm/dl, albumin – 4.4 gm/dl and globulin – 3.6 gm/dl. Albumin : globulin ratio was altered.

Serum protein electrophoresis (SPE) and urine protein electrophoresis (UPE) were negative for M – proteins.

Serum immunofixation electrophoresis (IFE) and urine immunofixation electrophoresis for M-protein were also negative. Free light chain assay (FLC) was not done in our institution.

Viral screening for Hep-B, Hep-C, HIV-I, and HIV2- were negative.

Pleural fluid was grossly haemorrhagic. On microscopy, total cell count was 1100/cumm with 40% plasma cells, 56% lymphocytes and 04% polymorphs. [Figure-1C] Total protein in fluid was 3gm/dl and LDH level was elevated (856 IU/L).

Bone marrow biopsy revealed hypercellular marrow with depressed erythropoiesis, granulopoiesis and megakaryopoiesis. There was marked infiltration by atypical plasma cells having bizarre nuclei. No evidence of granuloma or fibrosis was present [Figure-1D].

Fig 1A: Peripheral smear showing plasma cells (Leishman stain, Oil immersion); Fig 1B- Bone marrow aspiration showing atypical plasma cells (Leishman stain, Oil
immersion); Fig 1C- Deposit smear from pleural fluid showing atypical plasma cells (Leishman stain, Oil immersion); Fig 1D-Photomicrograph of bone marrow trephine biopsy showing atypical plasma cells (H&E stain-40x).

Flowcytometric immunophenotyping of the marrow showed 38% CD38+ve cells [Figure-2A] with cytoplasmic kappa light chain expression [Figure-2B]. CD19 and CD56 were negative [Figure-2C].

Complete skeletal survey did not reveal any osteolytic bone lesion.

DISCUSSION

Plasma cell leukemia (PCL) is a rare and aggressive variant of multiple myeloma with poor prognosis. It is characterized by presence of plasma cells in the peripheral blood is more than 2x10^9/litre or more than 20% of the circulating white blood cells [1]. In our case plasma cells constituted 28% of the white blood cells in peripheral smear.

PCL classified as primary and secondary. Primary PCL is the de novo appearance of PCL without prior occurrence of multiple myeloma and when PCL occurs as a terminal event in multiple myeloma – it is known as secondary PCL [4]. Primary plasma cell leukemia manifests with a short history of anemia, bleeding episodes, generalized weakness, weight loss, hepatosplenomegaly and absence of osteolytic lesions [5]. Our case had similar clinical presentation except bleeding manifestation. Osteolytic bone lesions were absent.

Our patient presented with respiratory distress and recurrent pleural effusion. This is a very unusual presentation of primary plasma cell leukemia. The diagnosis was more difficult due to non-secretory nature of this plasma cell leukemia.

Nonsecretory myeloma accounts for about 1% of cases of multiple myeloma. Clinical, radiological and haematological features are similar to those found in typical (secretory) myeloma except for absence of monoclonal protein in serum and urine and lower incidence of renal insufficiency [1]. Absence of a monoclonal protein in the serum and urine of patients with non-secretory PCL may be a result of an inability to excrete immunoglobin, low synthetic capacity for immunoglobin, increased intracellular degradation and rapid extracellular degradation of abnormal immunoglobins [6]. Immunofluorescence or immunoperoxidase studies show monoclonal cytoplasmic immunoglobulin as in our case which showed cytoplasmic kappa light chain expression.

The molecular basis of non-secretory myeloma could be due to loss of light chain production [7] or mutations that causes absence of cysteines required for disulfide bonds. This results in abnormal misfolded light chains which get retained in the plasma cells due to misfolding and are lysed.

Immunophenotyping was required to establish the diagnosis because of the uncommon clinical manifestation and non-secretory nature of the plasma cell leukaemia. Flowcytometric immunophenotyping of the marrow revealed 38% CD38+ve cells with cytoplasmic kappa light chain expression which supports the diagnosis of non-secretory plasma cell leukemia.

This case is being reported because of its extreme rarity and unusual clinical presentation.

REFERENCES