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Letter to Editor

Oncology Section

Anaemia in CD5 Negative Chronic Lymphoproliferative Disorder: Importance of Bone Marrow Examination

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Dear Editor,

Pure Red Cell Aplasia (PRCA) was initially described by Kaznelson in 1922. Acquired PRCA in association with non-Hodgkin's lymphoma has been reported rarely [1-4].

We report a case of 32-year-old P1+0 non lactating vegetarian female who presented with fever, generalised lymphadenopathy for five months. There was no history of abnormal bleeding. On examination, she had pallor, generalised lymphadenopathy and splenomegaly. Her White Blood Cell (WBC) count was 38.68 x 10^9 cells/l with 80% small to medium sized lymphoid cells with clumped chromatin and scant cytoplasm, few of the cells (10%) had nucleoli and nuclear clefting. The platelet count was 273x10^9 cells/l. Haemoglobin was 10.3 g/dl with MCV 89.5 fl, MCH 27.8 pg and RDW of 15.2%. Serum biochemical parameters including serum bilirubin (0.2 mg/dl), liver enzymes, protein, albumin, creatinine and urine routine microscopy were within normal range. Serum LDH was 560 IU/l. HIV, HBsAg, Anti-HCV antibody was negative. The patient had sufficient iron (sFe 165 µg/dl and sFerritin 285.25 ng/dl), folic acid (18.17 µg/l) and vitamin B12 190 ng/l. Bone marrow was hypercellular. Majority cells 85% where abnormal lymphoid cells. Erythroid and granulocytic precursors were reduced. Immunophenotyping of peripheral blood showed kappa restricted clonal B cells positive for CD19, CD20. The cells were negative for CD5, CD23 and CD10. Diagnosed as CD5 negative B chronic lymphoproliferative disorder, the patient was initially started on prednisolone (1 mg/kg/day) and chlorambucil (12 mg OD) for 7 days/month. On follow up her lymph nodes, spleen and WBC count regressed, however there was a persistent decline in Hb [Table/Fig-1] and the patient received a total of 7 unit packed red cell transfusion in three months.

Haematological Parameters	1st month	2nd month	3rd month	4th month
TLC (x 10^3 cells/µl)	38.68	23.79	25.79	14.0
Lymphoid cell %	80	87	84	83
Platelet (x 10^3 cells/µl)	273	181	290	458
RBC count (x 10^6 cell/µl)	3.71	0.66	0.64	0.67
Hb (g/dl)	10.3	2.6	2.4	2.6
MCV (fl)	89.5	83.0	87.3	87.3
MCH (pg)	27.8	38.9	38.6	38.8
S Bilirubin (mg/dl) (uncojugated)	0.2	-	0.9	0.7

[Table/Fig-1]: Follow up values of haematological parameters.

In work up for auto immune haemolytic anaemia Direct Antiglobulin (DAT) Coombs test was negative. Reticulocyte was <1%. The serology for parvo virus B-19 was negative. Repeat bone marrow showed fair number of granulocytic precursors and functional megakaryocytes. Erythroid precursors were <1% of the total nucleated cells. For PRCA patient was tried with anabolic steroids (stanozolol) at the end of fourth month. Patient responded well, her haemoglobin reached 10.6 gm/dl at three months of stanozolol therapy and is under follow up.

PRCA is a form of autoimmune cytopenia and recommendation for diagnosis includes severe normocytic normochromic anaemia with reticulocytopenia and ≤ 1% erythroid precursors in the marrow, negative DAT, absence of other features of haemolysis (normal haptoglobin, unconjugated bilirubin, LDH) and no parvo-B19 infection by PCR assay [4]. Our case fulfilled all these criteria.

Several pathophysiological mechanisms [5] for PRCA in lympho-proliferative disorders have been postulated including role of B cell, Tϒ cells and large granular lymphocytes of ϒδ type. It may also occur as a paraneoplastic syndrome of lymphoid malignancy. Immunosuppressive therapy and use of rituximab has been reported to be benefiting these cases [2-4].

Anaemia in lymphoproliferative disorders may have several aetiological mechanisms and such patients should be worked up for red cell aplasia after excluding therapy related anaemia and autoimmune haemolysis. The additional use of immunosuppressive therapy is helpful in achieving response.

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