Case Report

Evan’s Syndrome in HIV Infection

Shashikala A Sangle*, Rishi V Lohiya**

Abstract

Haematological complications are common in HIV patients and it can be because of infection per se or secondary to opportunistic infections and antiretroviral therapy. Evan’s syndrome i.e. autoimmune haemolytic anaemia with thrombocytopenia is however a very rare occurrence inspite of high Direct Coomb’s test positivity in HIV patients. We are reporting one such rare case of Evan’s syndrome in HIV patient probably first such reported case from India.

Introduction

Haematological complications are common in HIV infected patients, with anaemia being the most common. However autoimmune haemolytic anaemia or Evan’s syndrome (autoimmune haemolytic anaemia with thrombocytopenia) is rarely seen inspite of high prevalence of direct Coomb’s test positivity in this group. We are reporting one such very rare case of HIV with Evan’s syndrome probably first reported from India.

Case Report

A 32 years male known case of HIV infection for one year not registered with ART centre and not on any treatment presented with complaints of generalised weakness, easy fatigability and dark coloured urination for 15 days. There was no history of any drug intake, fever, cough or jaundice. Patient had hempe zoster over chest 2 years back which resolved in 30 days. There was no history of blood transfusion or any other major illness in past.

On examination patient was afebrile, weight 43 kg, pulse 96/min, BP 106/60 mm of Hg with extreme pallor, mild icterus, no pedal oedema/ rash/ petechiae/ herpes zoster scar/ oral candidiasis or lymphadenopathy. Per abdominal examination revealed mild hepatosplenomegaly. Other systems examination including musculoskeletal system was normal. Fundoscopy revealed superficial splinter haemorrhages.

Investigations

Hb – 3.0 gm %
PC – 60,000 /cu.mm
MCV – 104 µmol/dl

Peripheral blood smear - Anisopoikilocytosis with schistocytosis, nucleated RBCs +, with shift to left of myeloid series, metamyelocytes +, agglutinated clumps of RBCs +.

Reticulocyte count – 20%
Corrected retic count – 2%

Direct Coomb’s test was negative.

Indirect Coomb’s test was negative.

Bone marrow studies revealed M:E ratio of 1:4 with erythroid hyperplasia with megakaryocytosis. No acid fast bacilli seen.

Sr. LDH – 250 IU/L
Blood urea level – 20 mg/dl
Sr. bilirubin – Total- 2.7 mg/dl
SGPT – 28 IU/L
Sr. Proteins – 6.9 gm % (Albumin- 4.3, Globulin- 2.6)

USG abdomen s/o mild hepatosplenomegaly. No e/o abdominal lymphadenopathy.

Chest X ray - normal

CD4 count - 26/ cu.mm (4%)
Total lymphocyte count 758/ cu.mm.
Repeat CD4 – 87/ cu. mm. (7%) on ART
CD4:CD8 ratio- 0.102

ANA negative
Anti CMV antibodies
IgG > 250AU/ml (positive > 6)
IgM- 0.37AU/ml (negative)

Thus patient was diagnosed as a case of HIV infection with haemolytic anaemia with thrombocytopenia with positive direct Coomb’s test and reversal of CD4: CD8 ratio and bone marrow examination s/o hypercellular marrow with large megakaryocytes with no evidence of malignancy indicating autoimmune haemolytic anaemia with thrombocytopenia i.e. Evan’s syndrome.

Treatment - Patient was transfused with triple saline washed RBCs and was started on steroids (prednisolone 1 mg/ kg), injectable vitamin B12 and antiretroviral therapy. Patient responded to treatment and follow up hemogram after one week of treatment was suggestive of Hb 7.4 gm%, TLC – 13000/cu.mm, platelet count of 78,000 / cu.mm, corrected retic count of 1.5% and Hb 9.8 gm %, TLC – 11,000/cu.mm. with platelet count of 90,000 at the end of two weeks.

Discussion

The precise aetiology of anaemia in HIV infection is often inapparent and is frequently attributed to hypoproliferative marrow secondary to infection and inflammatory sequelae of HIV related illness. Myelofibrosis has also been associated with HIV infection and derangement of immune regulation of erythropoiesis has been postulated as one of the causes. While the most common cause of thrombocytopenia in HIV infection is documented to be due to immune destruction. Evan’s syndrome is simultaneous occurrence of autoimmune haemolytic anaemia and thrombocytopenia. The precise cause is not yet known, but the immunology does differ from patients with just ITP with thrombocytopenia.

Several studies have demonstrated that direct Coomb’s test is common in patients with HIV with incidence as high as 21 % but frank haemolysis has been reported very infrequently. The mechanism through which HIV patients have increased red cell associated antibodies remains unidentified. Presently accepted postulate is benign adherence of overabundant IgG to red cell associated antibodies.

HIV associated immune thrombocytopenia has been reported due presence of antibodies directed against membrane proteins as well as adherence of immune complexes to platelets. Reticulocytosis may not be present in all cases. Erythroid...
hyperplasia without reticulocytosis may be due to presence of antibodies directed against normoblasts or reticulocytes.4

In conclusion, this case and other reports6 confirm that clinically significant autoimmune haemolytic anaemia and thrombocytopenia do occur in HIV patients with or without reticulocytosis.7 Although patients with autoimmune haemolytic anaemia may constitute a small percent of patients with HIV, anaemia, and positive Coomb’s test, lack of reticulocytosis may lead to underdiagnosis of such condition. Thus high suspicion may help in early diagnosis especially if they are to be started on Zidovudine based therapy which is known to induce reduction of erythropoiesis.

References