

Case Report

Isolated Marrow Tuberculosis Presenting as Pure Red Cell Aplasia: A Case Report

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Abstract

Pure Red Cell Aplasia (PRCA) is a rare hematological condition. It has been associated with various viral, toxic or idiosyncratic insults. Tuberculosis involving erythroid precursors in isolation and presenting as pure red cell aplasia has not been described in literature. We report a case of otherwise asymptomatic 22 years male, who presented with pure red cell aplasia refractory to conventional therapy and later diagnosed to have tubercular involvement of bone marrow. He showed complete recovery of anemia with anti tubercular therapy.

Keywords: Pure red cell aplasia; Tuberculosis; Anemia

Introduction

Pure red cell aplasia is a condition of isolated suppression of erythroid precursors. Association of viruses, drugs and radiation has been documented. Tuberculosis has myriad of hematological manifestations. Disseminated tuberculosis has been associated with severe pancytopenia commonly. Most of the hematological manifestations are also complications of anti-tubercular drugs rather than disease itself. We report a unique case of tuberculosis presenting as pure red cell aplasia without any systemic symptom or sign.

Case Summary

A 22 years old male presented with increasing fatigue and tiredness from 2 months. His clinical examination was unremarkable with no lymphadenopathy or organomegaly although marked pallor was evident. There was no history of fever, rash, joint pain, significant weight loss or any medication in recent past. His personal, occupational and family history was non contributory. He was evaluated at another health facility. Lab investigations done there showed normocytic anemia with normal leucocytes and platelet count. Bone marrow aspiration was normocellular with paucity of erythroid precursors. CT chest excluded thymoma. He was diagnosed to have pure red cell aplasia based on bone marrow findings. He was started on oral steroid based therapy [prednisolone 2mg/ kg/day for 2 weeks followed by slow taper] along with red cell transfusion support. Danazol [200mg twice a day] was added as well after 6 weeks in view of no response. He remained transfusion dependent and after 3 months referred to our centre for further management. He was re-evaluated further. His initial lab investigations showed normocytic anemia with normal leucocytes and platelet count (Table). Serology for hepatitis B, hepatitis C and human immunodeficiency virus were negative. Bone marrow aspiration showed normocellular marrow with erythroid hypoplasia. Bone marrow biopsy showed normal cellularity, presence of granulomas along with isolated paucity of erythroid precursors. Ziehl-Neelson staining showed presence of acid fast bacilli (Figure). Thorough systemic assessment was done to find the primary tubercular focus but no other systemic involvement was noted. He was started on four drugs anti Koch's treatment (AKT; rifampicin, isoniazide, ethambutol and pyrazinamide). Steroid and

Danazol were withdrawn. He became transfusion independent after 3 weeks of AKT. He was treated with 2+10 months of AKT (4 drugs AKT for 2 months followed by 2 drugs AKT for 10 months). He is off therapy for more than 1 year and there is no recurrence of anemia.

Discussion

Tuberculosis is an endemic disease in India. Primary organ to involve is lung although it is not unusual to find cases with nervous system, bone, peritoneal or endometrium as primary presentation [1]. Miliary and disseminated tuberculosis is often associated with hematological abnormalities and bone marrow involvement is not uncommon [2]. Bone marrow examination done in severely immune compromised patients can reveal tubercular involvement [3]. Hungund et al. found 46% cases with myriad of hematological changes in clinically proven tuberculosis cohort however none was having erythroid hypoplasia [4].

Pure Red Cell Aplasia (PRCA) is a rare condition of absence of erythroid precursor cells in an otherwise normocellular bone marrow. It can be idiopathic or secondary to systemic illnesses, drugs or viral infections [5]. While primary or idiopathic PRCA needs treatment

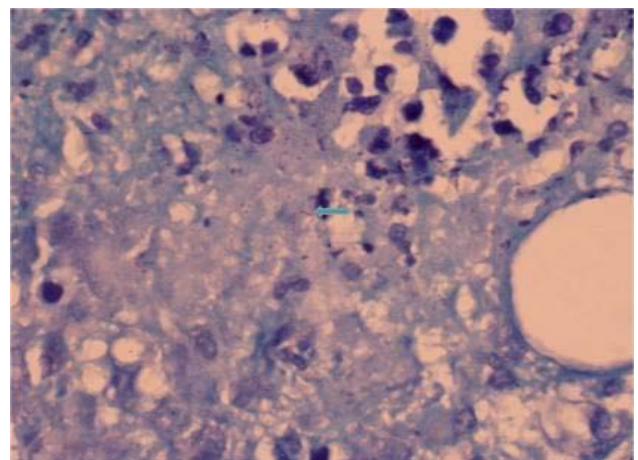


Figure: Ziehl-Neelson -stained sections of trephine bone marrow biopsy showing Acid Fast Bacilli (400X magnification).

Table: Baseline laboratory reports.

Hemoglobin	51g/L
Reticulocytes	0.0%
Red blood cell count	1.9x 10 ⁶ /μL
Mean Corpuscular volume	84.1 fl
White blood cell	17810/μL
Platelets	642 x 10 ³ /μL
LDH	119 U/ml
Serum Bilirubin total	0.45 mg/dl
Serum Bilirubin direct	0.14 mg/dl

with immunosuppressive medicines like steroid, cyclosporine or Rituximab, secondary PRCA can improve with removal of primary insult. Pathogenesis of PRCA secondary to infection is multi factorial and yet not well defined. Infection induced antibodies have been postulated to cross react with erythroid precursor cells causing PRCA [6]. T-cell induced selective erythroid series suppression and direct NK cell cytotoxic actions has been postulated as well [7]. As PRCA secondary to Koch's is rare, exact pathogenesis is unknown.

Index case presented with isolated anemia and diagnosed as PRCA. Tuberculosis presenting with PRCA as a primary presentation is extremely rare and not described in literature. Dutta et al. reported a case of PRCA in setting of symptomatic tuberculosis [8]. However the case described was having preceding AKT induced hepatitis as well and his bone marrow was negative for granuloma or AFB. To our knowledge, this is the first case who presented with PRCA as the sole manifestation of tuberculosis. Index case also showed sustained improvement in Hemoglobin after AKT, supporting the etiology.

Although rare, tuberculosis can present like isolated marrow involvement and PRCA. This case is the first described presentation of tuberculosis as PRCA in otherwise asymptomatic individual. This is an interesting association and an add-on entity in tuberculosis clinical spectrum.

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