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# PURE RED CELL APLASIA : A CASE REPORT

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**ABSTRACT** Pure cell aplasia is a rare bone marrow failure that affects erythroid lineage characterized by normocytic normochromic anemia with reticulocytopenia in the peripheral blood and absent or infrequent erythroblasts in the bone marrow. It can be congenital or acquired. Acquired can be primary when no cause is identified or secondary-due to underlying or associated pathology. Herein we report a case of a 28 year old female with Primary Acquired Pure Red cell aplasia. The patient presented with severe anemia (Hb-1.9gm%) and low reticulocyte count 0.1%. Bone marrow aspiration shows normocellular marrow with Decreased erythropoiesis with M:E ratio of 20:1..Patient was started on oral prednisolone and improvement was seen and the patient became transfusion independent.

**KEYWORDS :** Pure Red Cell Aplasia , Reticulocytopenia

## INTRODUCTION

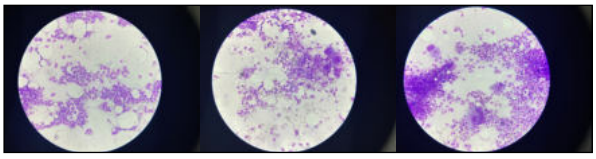
Pure red cell aplasia is a syndrome presents with anemia due to failure of bone marrow erythropoiesis characterized by normochromic normocytic anemia with severe reticulocytopenia and marked reduction or absence of erythroid precursors from bone marrow. It is a rare disorder which may be congenital or acquired. Congenital PRCA (DIAMOND BLACKFAN ANEMIA) is a disease of ribosomal biogenesis, classical features include Craniofacial Dysmorphism and thumb abnormalities. Acquired may be primary or secondary. Primary acquired PRCA is an autoimmune disorder in which an immune mechanism interrupts erythroid differentiation. Secondary PRCA associated with autoimmune/ collagen vascular disorders, lymphoproliferative disorders-chronic lymphocytic leukemia, infections like Parvovirus B19 infection, HIV, Hepatitis, Tuberculosis, hematological malignancy, non hematological malignancy of which thymoma is best known, Drug induced and Pregnancy.

## CASE REPORT

A 28 year old female housewife, presented to medical OPD with complaints of easy fatiguability and generalized weakness for 2 months and 2 bouts of vomiting in 2 days. Patient denied fever, blood loss, jaundice, weight loss, loss of appetite, Joint pains, rash, lump in abdomen, drug ingestion. She has no known prior illness and There was no history of any blood transfusion and family history was nil significant.

Physical examination revealed severe pallor, No signs of icterus ,enlarged lymph nodes. Her vitals were BP 100/70mmHg,PR 106bpm,RR 24 breaths/min,SPO2-99%@RA,TEMP - afebrile. Flow murmur was heard on auscultation, no hepatosplenomegaly, no signs of heart failure. Her initial blood tests showed severe anemia with Hemoglobin - 1.9gm%, Hematocrit -5%, Total RBC-0.54millions/cumm , Reticulocyte count-0.1%, MCV-92.1fl, MCH-35.5pg, MCHC-38.6gms%, Total Leukocyte count-8340 with DLC -Neutrophils-86, lymphocytes-10, eosinophils-2, monocytes-2, basophils-0, platelets-2.13lakhs, ESR-120mm/1st hr. Peripheral smear shows NORMOCYTIC NORMOCHROMIC ANEMIA OF SEVERE DEGREE WITH RELATIVE NEUTROPHILIA. Other abnormalities include deranged LFT with Total Bilirubin-1.3mg/dl with direct bilirubin 0.4mg/dl, SGOT-253 IU/L, SGPT-239 IU/L, ALP-86 IU/L, S.Total proteins-6.3gm/dl with albumin 3.6gm/dl.RFT and CUE were normal. Bone marrow aspiration showed Decreased erythropoiesis with only occasional early erythroblasts showing dysplastic changes and increased myelopoiesis M:E ratio of 20:1.Megakaryocytes were increased in number with hyper and hypolobated forms.

As PRCA is suspected further investigations were done to know the etiology. None of the common causes of PRCA was found-no infection Hepatitis B, Hepatitis C, Hepatitis A, HIV I and II serology were negative, no evidence of parvovirus B19 infection; no evidence of neoplasm in thorax abdomen; Direct Coombs test Positive, indirect Coombs test negative so Hb electrophoresis was done which turned out to be normal. Other investigations include ANA -negative, RA factor -negative, Stool Occult blood-negative, thyroid profile was normal as well as iron levels. Karyotyping found no abnormality.



Initially 4 units PRBC was transfused and then oral prednisolone was initiated with a dose of 1mg/kg/day. Response was observed after 3 weeks of starting treatment with an increase in reticulocyte count 1.5% and hemoglobin was 11.3 ,platelet and WBC were within normal range. LFTs were repeated and showed S.TotalBilirubin-0.9, Direct-0.2,Indirect 0.7,SGOT-18,SGPT-36,ALP-72,S.total proteins-6.9.Tapering of steroids was started and tapped off over 6weeks. During this period hemoglobin was maintained between 10-12g/dl and the patient became transfusion independent.

## DISCUSSION

In the case reported above, the patient presented with severe normocytic normochromic anemia with low reticulocyte count but without evidence for general bone marrow failure, which gave a concern about Pure Red Cell Aplasia. Positive Direct Coombs test indicates excessive immune complexes production but reticulocytopenia instead of reticulocytosis makes autoimmune hemolytic anemia diagnosis least likely which was further confirmed by normal Hb electrophoresis. Bone marrow aspiration supports our diagnosis of pure red cell Aplasia with Normocellular bone marrow with decreased erythropoiesis and but with occasional dyserythropoiesis which raised a concern about early presentation of myelodysplastic syndrome as PRCA but age of onset, peripheral smear without any dysplastic changes, normal karyotyping and response to oral steroids makes MDS least likely diagnosis.

PRCA is very diverse disease with no standardized therapeutic strategy. First line for Acquired PRCA(primary or idiopathic) is immunosuppressant's which include steroids, cyclosporine A , Rituximab, Antithymocyte globulin. Steroids are the preferred immunosuppressant. Most of the patients require Maintenance therapy to prevent relapse. Immunosuppression is less effective in relapsed PRCA than Naive PRCA and patient may develop refractory PRCA which is treated with Monoclonal antibodies like Alemtuzumab and Rituximab. Secondary PRCA can be managed by removing the primary insult. Stem cell transplantation should be considered in congenital PRCA.

## CONCLUSION

PRCA is a rare disorder of bone marrow failure and should be

suspected in isolated anemia with severe reticulocytopenia. Bone marrow aspiration helps to confirm our diagnosis. Investigations should be done to know the secondary causes of PRCA. Treatment for primary PRCA is Immunosuppression.

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