CASE REPORT

Cyclosporine A responsive pure red cell aplasia in a child with systemic lupus erythematosus

Niranjan Biswal · Parameswaran Narayanan · Ginigeri Chetan

Abstract PRCA is a rare complication in SLE. Response to various drugs like steroid, Intravenous Immunoglobulin (IVIG), recombinant human erythropoietin and plasmapheresis vary in different cases. Cyclosporin A may be effective in cases even unresponsive to other modalities of therapy.

Keywords PRCA · SLE · Cyclosporine-A

Abbreviations

SLE – Systemic lupus erythematosus.

CY-A – Cyclosporine-A

PRCA – Pure red cell aplasia

Introduction

Pure red cell aplasia (PRCA) is a rare hematological condition in children. Acquired PRCA due to virus human parvovirus-B 19 (i.e. HPV B 19) is transient and usually recover spontaneously. However it may be secondary to various other causes like viruses, connective tissue disorder systemic lupus erythematosus (SLE), drugs and malignancy which may last for a longer duration. Requiring frequent packed cell transfusion and supportive therapy [1].

PRCA is a severe conditions requiring intervention to correct the anemia which has direct influence on recovery from the primary condition, response to drugs and secondary infection due to reduction in cell mediated immunity, because of anemia [1]. Anemia is the commonest abnormality seen in SLE. It may be due to inflammatory cytokines, iron deficiency, autoimmune hemolytic anemia, PRCA [2]. PRCA associated with SLE is a rare condition reported only in 32 cases [3].

We report a case of PRCA secondary to SLE, who responded to cyclosporine within a week after a failure to respond to therapy with erythropoietin and high dose steroid therapy.

Case report

A 9-year-old girl presented to us with severe anemia and CCF. She was diagnosed to have SLE at the age of 5 years. She received prednisolone, hydroxy chloroquine and paracetamol for initial 2 years. She was taking prednisolone 10 mg daily for the last 6 months. and was symptom free for two and half years. She became pale and was unable to walk.

N. Biswal¹ (✉) · P. Narayanan² · G. Chetan³

¹Professor

²Assistant Professor

³Senior Resident

Department of Pediatrics, JIPMER,
Pondicherry - 605 006, India

Ph: 0413-2273139

Fax: 0413-2272067

E-mail: drnbiswal@yahoo.com

nbiswal@jipmer.edu

She was a healthy looking child with no evidence of malnutrition. she had severe pallor, palpable spleen 2 cm below the costal margin and congestive heart failure. Other systems were normal. Her renal, hepatic parameters were normal. 2 D echocardiography revealed mild cardiomegaly and normal ejection fraction with no vegetation. Investigation revealed, Hemoglobin (Hb) =2 gm/dl. Total leucocyte count=8000/mm³ and differential count = N=52%, L=43%, E=5%, platelet count 185000/ dL. Bone marrow aspiration showed hypo cellular marrow with complete absence of erythroid progenitor cells. Her hematological picture was consistent with PRCA.

She was treated with frequent blood transfusions (approx 2 units of packed cell every week) for 3 weeks. Her Hemoglobin never rose above 6 gm/dl and fell to 3 gm/dl in a week after transfusion. She was treated with pulse methyl prednisolone without success. She was treated with. Human recombinant erythropoietin (Wepox) in a dose of 100 I.U./Kg /dose three times a week for 8 weeks with minimal rise in Hb, but no change in requirement for transfusion. Her Hb rose to 4 gm/dl after 2 weeks and reticount increased to 4 %, which lasted for 2 weeks and later came down to 1 %. With an Hb level of 3 Gm% without any further rise in hemoglobin.

Hence she was treated with cyclosporine A (CY-A) in a dose of 2.5 mg /kg/dose twice a day. Her Hb rose to 7 Gm% after 1 week of therapy and steadily increased there after up to 16 gm% after 4 months of therapy. It was steady at 16 gm% at the completion of 4 months of therapy and at this point cyclosporine was tapered and stopped over a month. she developed hypertension after two weeks of therapy with Cy-A and required antihypertensive therapy. Subsequently she had maintained an Hb level of 11–14 gm % level steadily even after 4 years of discontinuing the CY –A. Her latest hemoglobin level was 13 gm/dl in June, 2006. However she developed nephritic range proteinuria after 1 year of Cy-A treatment and was treated with cyclophosphamide pulse therapy every month for 5 cycles with low dose prednisolone (10 mg/day). This is probably the longest remission response period for PRCA to cyclosporine therapy.

Discussion

Anemia of chronic disease occurs in patients during the period of active disease. It had been described with human parvovirus B-19 infection or with out it in cases with SLE [1].

Withdrawal of the offending drug like procainamide, phenytoin is absolutely necessary. Successful treatment high dose IVIG, pulse methyl prednisolone and plasma-pheresis had been reported. A few cases of PRCA with

SLE had maintained acceptable level of Hb for up to 2 years after Cy-A therapy [3]. Oral CY-A had been effective in treating previously immunosuppressed and untreated patients with severe bone marrow failure in adults [4]. Despite high levels of endogenous erythropoietin (Ep) PRCA responded only to human recombinant Ep after unsuccessful therapy with high dose glucocorticoid and IVIG [5]. However, there had been no controlled study to recommend a single mode of management in this group of patients. This case had a previous history of steroid and hydroxyl chloroquine treatment for SLE and a quiescent phase after that. This case had a previous history of steroid and hydroxyl chloroquine treatment for SLE and a quiescent phase after that. She did not respond to erythropoietin therapy and continued to have very low hemoglobin in the range of 3–4 grams and required frequent transfusion.

Her hemoglobin level continued to rise after 7 days of therapy with cyclosporine A and maintained at 11–14 gm % level steadily even after 4 years of discontinuing cyclosporine –A.

This is probably the longest remission response period for SLE with PRCA treated with cyclosporine-A.

Conclusion

Anemia associated with PRCA, which is unresponsive to conventional blood transfusion, IVIG, high dose steroid and erythropoietin therapy, may respond satisfactorily to CY-A and can maintenance at an acceptable level of hemoglobin for a longer period.

References

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