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Original Research Article

Idiopathic aplastic anemia: a rare case report in Jammu and Kashmir region, India

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ABSTRACT

Idiopathic aplastic anemia is a pancytopenia disorder that is a rare but life threatening for both mother and fetus during pregnancy. Association of aplastic anemia with pregnancy is unclear but considered to be interrelated. Bone marrow transplantation is the most effective treatment for adult aplastic anemia but is inadvisable to perform during pregnancy because of the teratogenic effect of immunosuppressive agents or radiation therapy to the growing fetus. Supportive care, withdrawal from offending drugs and involving erythrocytes and platelets transfusion is a promising way to save the life. Here author present a case report of 36-year-old lady with idiopathy aplastic anemia. In this case medical investigation revealed severe anemia of unknown origin. The patient was treated with hematinics, blood transfusion and glucocorticoids. A healthy baby was delivered without evidence of hemolysis at her eight month and one week of pregnancy, the patient recovered and discharged with normal incidence. Being a rare case, it becomes a necessity to report such life-threatening disorder and management. Moreover, to our knowledge this is the first case reported of its kind from Jammu and Kashmir Division of India.

Keywords: Erythrocytes, Idiopathy aplastic anemia, Immunosuppressive agents, Pregnancy, Platelets transfusion

INTRODUCTION

One of the leading causes of maternal and perinatal deaths worldwide, especially in malaria endemic region during pregnancy and puerperium remains the severe hemolytic anemia.¹⁻⁵ Estimated incidence rate of idiopathy/Acquired aplastic anemia is (AA) reported to 1-2 case per million per year.⁶ This rare and life-threatening disorder consist of pancytopenia especially for women during pregnancy and is considered to be either acquired or congenital.⁷⁻¹⁰ Inciting factors like infection, drugs or organic compound as well as radiation are considered to be responsible for acquired aplastic anemia.¹¹⁻¹³ The pathogenic mechanism underlying this disorder is likely to be an immune mediated. In genetically predisposed

individual overproduction of bone-marrow inhibiting cytokines elicited by abnormal T-cell remains the underlying mechanism.¹⁴ Complaints of jaundice, profound anemia, massive splenomegaly with recurrent infections are presented in such patients.¹⁵ However, it does not compromise with fertility and pregnancy of women.^{13,16} But pregnancy complicated by such disorder is a great challenge for obstetricians.¹⁷ As its association with significant fetal, neonatal, and maternal morbidity and mortality is well reported. Although the causal relationship between AA and pregnancy has not been conclusively established.¹⁸ Prescribing the use of Immunosuppressive agents or involving particular procedure of hematopoietic stem cell transplantation are inadvisable for treating the disorder during pregnancy as

they have potential toxicity effect to growing fetus.^{11,13} There is no agreement about the optimal supportive care, treatment regime and even no clear guidelines are provided for the management of aplastic anemia disorder during pregnancy. However, new guidelines on the diagnosis and management of adult AA have been recently published by the British Society for Standards in Hematology. Thus, the best treatment options are erythrocytes and platelet transfusions to save the precious life.^{10,19}

CASE REPORT

A 34-year-old female with O positive blood group was presented to an outpatient Department of Gynecology and obstetrician of Hera Nagar District Hospital of Jammu and Kashmir, India. The patient had some health issues during her second pregnancy towards the end of First trimester during 11th week of gestation. Patient complained few symptoms of generalized weakness, restless and breathless, pale skin color and persistent

vomiting and nausea. Medical investigation revealed observation of low-grade fever, whitish discoloration of nail buds and Tachycardia (high pulse rate) more than 120 bpm and even hypo tension (60-65/90-100). There was no such complication on her earlier pregnancy, where she delivered a healthy male baby at the age of 30. Pathophysiological study revealed Iron, folate and B12 levels normal. Auto immune haemolytic anemia was ruled out by coombs test and observed to be negative, her biochemical investigation including glucose-6phosphate dehydrogenase (G6PD), hemolysis thalassemia profile, serum heptoglobins analysis all ruled out to be normal. Her renal function test was normal. However, LFT serum glutamic-oxaloacetic transaminase (SGOT) and serum glutamic pyruvic transaminase (SGPT) were increased to 128 and 142 U/L respectively as compared to normal range of 5-40 U/L. Hemoglobin (Hb) HPLC/ electrophoresis fallen within normal range. However, her reticulocytes counts were raised. Her complete blood counts during different phases of pregnancy are shown in Table 1.

Table 1: Complete blood counts.

Duration of pregnancy	RBC counts (mil/mm ³)	Normal range	Mean corpuscular Hb conc.(gm/dl)	Normal range	Red cell distribution width (RDW %)	Normal range
3 rd month	3.35	3.80-4.80	31.80	32.00-35.00	17.80	11.50-14.50
4 th month	2.73		31.05		19.23	
6 th month	2.20		30.08		23.50	
8 th month	2.58		31.9		22.9	

On observation of peripheral blood film (PBf) no fixed pattern could be concluded (Table 2). Every possible cause of anemia during pregnancy was ruled out but

nothing significant was found. A care full history of patient reviled suffering from polycystic Ovary ovarian Disease (PCOD).

Table 2: Peripheral blood film.

Duration of pregnancy	Peripheral blood film observation
Late 2 nd trimester early 3 rd trimester	Dimorphic anemia
Early 3 rd trimester	Roleux formation
Mid 3 rd trimester	Microcytic and hypochromic anemia

She was undergoing medicine ginette 35 taken for just 15 days approx while she conceived for her second pregnancy. No bleeding or spotting reported during the tenure of second pregnancy. Although candidiasis infection of reproductive tract occurred at 24th weeks of gestation for which medication ziprase 200 (salt cefaxime) was taken under supervision of gynaecologist. Regular checkup was followed for every 15-20 days. Iron

and calcium tablets were prescribed during pregnancy but her hemoglobin level kept falling onward fifth month of pregnancy. Inject able iron (Ferric 5CC on 100cc of normal saline given i/v. and vit B12 were administered by the mid of second trimester that resulted in four times iron overload in the patient. In such cases of gestational anemia, the only possible modality of management is repeated blood transfusion. So, it was decided for first

blood transfusion at her seventh month of gestation and second blood transfusion on third day of first transfusion, a drastic improvement was observed in the patient. This was followed by weakly transfusions continued till the end of her eighth month of gestation. Doctor decided to deliver the baby on completion of eight months and one week to avoid any risk to the growing fetus. Patient was induced for normal vaginal delivery and one transfusion was received a day before delivery. First four transfusions were given twice weekly. That's makes total of six units of packet cells given, and transfusion was repeated six times during pregnancy. Patient gave birth to a healthy female baby weighing 2.127 kg with no more transfusion after delivery. Patient improved and symptoms completely resolved within two months of delivery. Hemoglobin levels started improving 10 grams within one and half months with supplements hembpack and coral calcium and cholecalciferol (vitamin D3). Hb levels were regularly monitored after delivery for a period of three months.

DISCUSSION

First case of aplastic anemia in pregnant woman was reported by Ehrlich et al. Although, true etiology of this rare but life-threatening disorder is uncertain but pregnancy associated severe aplastic anemia is thought to be a major risk factor.¹² High level of pregnancy hormone including the placental lactogen, and estrogen erythropoietin are increased. However, increased level of placental lactogen and erythropoietin stimulates hemopoiesis. On the contrary increased level of estrogen level during pregnancy inhibits hemopoiesis.^{9,11-13} The increased level of sex hormones are considered to be the trigger point whose level fall to normal range after delivery and such spontaneous recovery has been reported earlier.²⁰⁻²² In this case also the patient recovered after delivery. Her hemoglobin levels started improving and attained 10 grams within a month time. As our patient had earlier pregnancy with no such complications it seems to be acquired aplastic anemia and as in most cases the pathophysiology of acquired aplastic anemia is immune mediated where role of sex hormones such as estrogen and progesterone in affecting the immune response had been previously mentioned.^{7,23} However, increased sex hormone level do not cause autoimmune disease alone but such abnormal hormone levels provide the stage for other factors like genetic, infections, etc. to trigger aplastic anemia.²⁴ A careful review of the history of our patient looks to be adult onset and none identifiable inciting factors could be concluded. Exact reason for the onset of disease at her second pregnancy could not be ruled out. Case history revealed patient with PCOD and had infection at the time of pregnancy. Candidiasis infection trigger the onset of disorder is matter of further investigation as the chemical composition of drugs/organic compounds/ antibiotic as well as infections acts as inciting factors to trigger acquired aplastic anemia.¹¹⁻¹³ So, the first choice of treatment recommended is blood transfusion maintaining

hemoglobin levels ≥ 8.0 gm/dl and platelets count $\geq 20,000$ /cmm.²⁵ In this case decision of inject able iron and vit B12 caused four times iron overload in the patient, but after providing the patient with two units of bloods transfusion and a of total six blood transfusion, drastically improved the patient condition and rescued the mother from this rare life threatening disorder.

CONCLUSION

Successful outcome of potential life-threatening idiopathy aplastic anemia till date remains withdrawal from offending drugs, supportive care, repeated blood and platelet transfusion. Every possible cause in our case was ruled out but nothing significant was found. In such cases of gestational anemia, the only possible modality of management is repeated blood transfusion. Our case reports the successful recovery of patient from this rare, life threatening disorder.

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