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Original Research Article

Observation of Autoimmune Hemolytic Anemia in Children in a tertiary care centre of Odisha

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Abstract: Fourteen children with autoimmune hemolytic Anemia are described. Twelve had acute and two had insidious onset with positive direct coombs test. Pallor was the main presenting complaint followed by hepatomegaly, fever, splenomegaly, jaundice and haemoglobinuria. Out of 14 cases 10 belonged to primary group and 4 to secondary group. Secondary causes were SLE and Lymphoma. Oral Prednisolone produced remission in all cases but 4 had relapses after initial response. 4 out of 10 primary cases needed steroid in a tapering dose for 16-24 wks, 6 cases responded to a short course of 3-4 wks of steroid with no relapse till yet. Average Blood Transfusion given before diagnosis of AIHA was three. Average time duration to diagnosis of AIHA was two days in our setup.
Keywords: Autoimmune hemolytic Anemia, Steroid, Blood transfusion.

INTRODUCTION

Autoimmune hemolytic anemia is a condition where antibody directed against autologous red cells most frequently demonstrated by a positive direct coombs test [1]. As its annual incidence is 1:80,000 it is very rarely thought in any case of anemia but it has excellent prognosis if promptly diagnosed and treated [2]. Thus this study was done to study the clinico- hematological profile and treatment outcome of AIHA in children with brief review of the literatures.

MATERIALS & METHODS

This is a prospective study involving 14 patients <14 yrs of age with positive direct coombs test (DCT) diagnosed as AIHA during May 2014 to 2017. Age, Sex and clinical presentations of all cases were studied. In all these cases complete blood count, peripheral smear comment, Recticulocyte count, G-6 PD Activity, Liver function test, Renal function test, some special test like sepsis screening and DCT was done. Bone marrow study, Chest Xray, ANA, CT scan of Abdomen and thorax, Lymph node biopsy were undertaken to identify the secondary causes. The patients were treated with blood transfusion, steroid IV/Oral as per the clinical conditions. Number of Blood transfusion taken in our unit and by referred hospitals before diagnosis, Average time to diagnosis of AIHA and number of blood transfusion after diagnosis were documented. The patients were put on regular follow up every 4 weeks. The dose and duration of therapy was recorded.

RESULTS

The patients included 8 male and 6 female whose age at the onset of the disease ranged from 5 to 13 years. 12 had acute onset and 2 had insidious onset of disease (Table No.1).

Table-1 Age, Sex and mode of onset of disease

Age at onset	Clinical characteristics
Mean +_ SD Range	10.5 ± 2.76 yr. 5-13 Yr
Sex	
Male	8
Female	6
Mode of onset	
Acute	12
Insidious	2

All patients had pallor as presenting complaint followed by hepatomegaly, fever, splenomegaly, jaundice, hamoglobinuria, lymphadenopathy (figure-1). Out of 14 cases 10 had primary and 4 had secondary causes (Table-2). 8 cases had Hemoglobin <5 gm/dl at the time of admission and six had more than 5 gm/dl but less than 10gm/dl. Leucocytosis was seen in 4 cases.

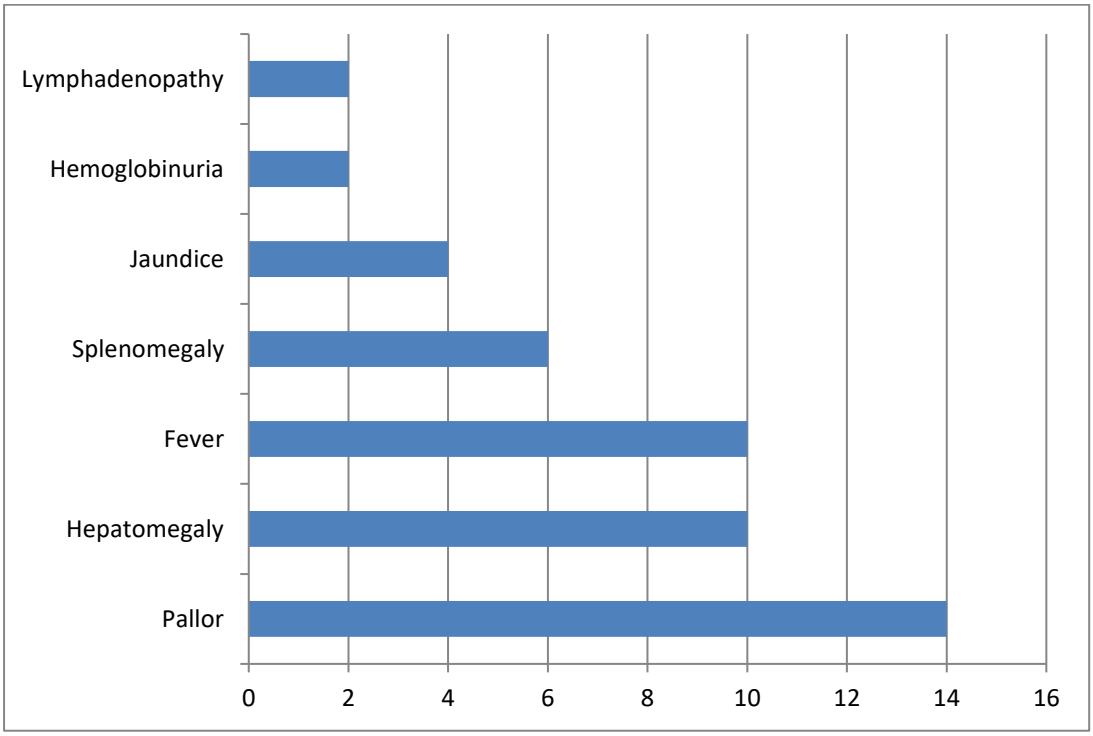

Fig-1: Clinical presentation of the cases

Table-2 Distribution of Cases

Primary	10
Lymphoma	2
SLE	2

Table-3: Hematological findings at the time of diagnosis

Cases	Age (Years)	HB(gm/dl)	Retic count	WBC/mm3	Platelet count*	Peripheral smear comment	Bone marrow study	Coombs test
1	14	4.6	7	12,400	Normal	Microcytic	Erythroid hyperplasia	+ve
2	6	5	9	20,500	Normal	Dimorphic anemia	Erythroid hyperplasia	+ve
3	11	2	5	22,800	Normal	Neutrophillic Leucocytosis	Infective	+ve
4	13	2	6	9,800	Normal	Dimorphic	Megaloblastic	+ve
5	9	8	4	6,800	Normal	Spherocytosis	Not done	+ve
6	12	8	4	7,800	Normal	Spherocytosis	Not done	+ve
7	9	6	5	11,200	Normal	Spherocytosis	Not done	+ve
8	12	8	4	7,800	Normal	Spherocytosis	Not done	+ve
9	9	8	4	6,800	Normal	Spherocytosis	Not done	+ve
10	13	2	6	9,800	Normal	Dimorphic	Megaloblastic	+ve
11	11	2	5	22,800	Normal	Neutrophillic Leucocytosis	Infective	+ve
12	6	5	9	20,500	Normal	Dimorphi anemia	Erythroid hyperplasia	+ve
13	14	4.6	7	12,400	Normal	Microcytic	Erythroid hyperplasia	+ve
14	9	6	5	11,200	Normal	Spherocytosis	Not done	+ve

*More than 1,00,000/mm³

Table-4: Treatment Profile of the cases

No	No of BT given given before diagnosis of AIHA	Time to diagnosis of AIHA in our Hospital in days.	Treatment given	No of BT given after Diagnosis
1	6	2	BT+IV methylprednisolone	1
2	3	2	BT+Oral steroid	Nil
3	6	1	BT+Antibiotic+IV mathylprednisolone	1
4	3	2	BT+oral steroid	1
5	Nil	2	BT+Oral steroid	-
6	Nil	2	BT+Oral steroid	-
7	2	1	BT Oral steroid	1
8	Nil	2	BT+Oral steroid	-
9	Nil	2	BT+Oral steroid	-
10	3	2	BT+Oral steroid	1
11	6	1	BT+Antibiotic_IV methylprednisolone	1
12	3	2	BT+Oral steroid	Nil
13	6	2	BT+IV methylprednisolone	1
14	2	1	BT Oral steroid	1

Table-5 Duration of therapy in Primary AIHA

Cases	Duration
6	3-4 wks
4	16-24 wks

Megaloblastic marrow and infective marrow was seen in two cases. Platelet count was normal in all cases (Table-3). Average number of blood transfusion given before diagnosis of AIHA was 3. Average time duration to diagnosis of AIHA was 2 days in our setup. In four cases six blood transfusions were given before diagnosis and average one transfusion was needed after treatment with steroid. All patients received packed red cells. Four patients given IV methyl prednisolone at a dose of 30 mg/kg/day. Ten patients received oral steroid at dosed 1-2 mg/kgday (Table-4). 6 cases responded to short course of steroid (3-4 weeks) with no relapse till yet. 4 cases needed steroid in a tapering doses for 16-24 weeks, two out of these four cases had relapses, now following a chronic course and was planned for alternate therapy (Table-5). Common adverse effects of long term therapy was cushingoid facies, truncal obesity and striae.

DISCUSSION

Autoimmune hemolytic Anemia is an uncommon condition in pediatric practice. 71% cases belonged to primary group. Dacie, Dausect et al reported an occurrence of approximately 70% of idiopathic variety[3]. Male female occurrence was 1.3:1. Buchanan et al in 1976 reported male female ratio of 1.2 to 1.9:1. [4, 5].

All the patients had moderate to severe anemia of acute onset, some of them also had fever, hepatomegaly, Jaundice, Splenomegaly. This spectrum of clinical feature is well recognized in pediatric patient [4, 6].

The most important and useful laboratory test to establish the diagnosis of AIHA is direct antiglobulin test (DAT or Coomb's test) [2, 7]. All patients in our study were DAT positive.

10 out of 14 patients were idiopathic as no cause could be ascertained. Lymphoma and SLE were two secondary causes of AIHA. These are common secondary causes reported in literature [8, 9].

None of our patients had low platelet count i.e.<1,00,000/mm^3. The white blood cell count is usually within normal limit in autoimmune hemolytic anemia but in acute hemolysis, leukocytosis can be noted, and the usual level is above 30,000/mm^3 [10]. In our series 3 pts had leukocyte count above 20,000/mm^3 suggestive of leukocytosis. Bone marrow examination usually reveals erythroid hyperplasia and rarely megaloblastic changes [11]. In our study two cases had megaloblastic changes and two had feature of marrow infection which was later diagnosed as lymphoma by lymphnode excisional biopsy. Four cases showed erythroid hyperplasia and bone marrow was not done in four cases. Recticulocytosis was marked in almost all cases and average count was five.

Six out of fourteen (42.8%) patients presented with severe anemia and heart failure. This is in agreement with the pattern of AIHA because of acute nature of disease in most cases. The same presentation was reported in different studies [12, 13].

There is no literature available about the number of transfusion given before a diagnosis of AIHA is made and exact time taken to a diagnosis of AIHA. In our series average three transfusions given prior to establishment of AIHA with average time duration to diagnosis was two days in our hospital. In four cases six blood transfusions had been given before DAT (Direct Antiglobulin Test) showed positive result. This can be well explained by the facts that poor index of suspicion by treating physicians because of low incidence, lack of proper lab facility, symptomatic management and delayed reaching to tertiary care centre. Possibility of alloimmunisation due to frequent transfusions also cannot be ruled out.

Initially all patients responded to steroid which was similar to response in other studies [14–16]. Four patients were given IV methylprednisolone because of the serious general condition. The others were put on oral steroid. Six patients had relapse, two had subsequently diagnosed as lymphoma. Two cases responded short course of steroid but another two cases had relapses even after one year in tapering dose of steroid and entered into chronic course so planned for IVIG therapy. IVIG though one modality of treatment, the use was deferred in our series due to non affordability, more often acute nature of disease in our study. Two patients who were in chronic course were given IVIG and was marked no relapse till yet. 60% primary cases responded to short course steroid (3-4 weeks). Steroid toxicity though marked in 4 cases, subsided after gradual stopping and changing over to IVIG therapy.

CONCLUSION

Autoimmune Hemolytic Anemia though uncommon, a high index of suspicion and workup in any case of acute, sub-acute anemia and early treatment improves the outcome. Blood transfusion without expected rise of hemoglobin and difficulty in grouping due to lysis are early clues to the diagnosis, prompt treatment and need of lesser BT. This study further suggests study of antibodies to different alloantigen in high transfusion cases.

REFERENCES

1. Packman CH, Leddy jp: Erythrocyte disordersanemias due to increased erythrocyte destruction mediated by antibodies. In Williams wj, Beutler E, Erslev Aj, Lichtman MA, eds. Hematology, 3rd ed. Newyork, Mcgranz Hill company, 1983, 632-647.
2. Schreiber AD, GillFM, Manno cs. Autoimmune, hemolytic anemia. In Nathan DG. Orkin SH, eds. Nathan and oski's Hematology of infancy and childhood 5th ed. Vol. 1Philadelphia, W.B. Saunders Company, 1998; 501-506.
3. Dacie JV: Autoimmune Hemolytic anemia. Arch Intern Med. 1975; 135(10):1293-1300.
4. In Behrman RE, Kliegman RM, jenson HB, eds. Nelson Textbook of Pediatrics, 18th ed. Philadelphia; Saunders, 2000; 1638-1640.
5. Buchanan GR, Boxer LA, Nathan DG. The acute and transient nature of idiopathic immune hemolytic anemia in childhood. J Pediatr. 1976; 88:780–783.
6. Naithani R, Agrawal N, Mahapatra M, Pati H, Kumar R, Choudhury VP. Autoimmune hemolytic Anemia in India: Clinico-hematological Spectrum of 79 cases. Hemaology 2006; 11:73-76.
7. V.Gupta, J.Shukla, B.D. Bhatia. Autoimmune hemolytic Anemia. Indian Journal of Pediatrics, Vol 75-May,2008..
8. Carapella deLucaE. Autoimmune hemolytic anemia in childhood: Follow-up in 29 cases. Vox Sang.1979; 36:13-20.
9. Oliveria MCLA. Clinical course of autoimmune hemolytic anemia: an observational study. J pediatr(Rioj). 2006; 82:58-62.
10. Habibi B, Homberg JC, Schaison G, Salmon C. Autoimmune hemolytic anemia in children: a review of 80 cases. Is J Med. 1974; 56:61–69?
11. Sokol RJ, Hewitt S, Stamps BK, Hitchen PA. Autoimmune haemolysis in childhood and adolescence. Acta Haematol. 1984; 72:245–257.
12. Heisel MA, Ortega JA. Factors influencing prognosis in childhood autoimmune hemolytic anemia. Am J Pediatr Hematol Oncol. 1983; 5:147–152.
13. Hendrick AM. Auto-immune haemolytic anaemia - a high-risk disorder for thromboembolism? Hematology. 2003; 8:53–56.
14. Pullarkat V, Ngo M, Iqbal S, Espina B, Liebman HA. Detection of lupus anticoagulant identifies patients with autoimmune haemolytic anaemia at increased risk of venous thromboembolism. Br J Haematol. 2002; 118:1166–1169.
15. Arndt P, Leger RM, Garratty G. Serologic findings in in autoimmune hemolytic anemia associated with immunoglobulin M warm autoantibodies. Transfusion. 2009; 49:235–242.
16. Salama A, Janvier D, Mayer B, Saison C, Moscatelli H, Aucouturier F, Yilmaz P, Arnaud L, Wild V, Knop S, Cartron J-P. Lethal autoimmune hemagglutination due to an immunoglobulin A autoagglutinin with Band 3 specificity. Transfusion. 2014; 54:1988–1995.