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Original Research Article

Study of Infections among Children with Multitransfused Thalassemia Major

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Abstract

Background and Aim: Due to the high annual expense of thalassemia treatment in India, the majority of thalassemic children receive insufficient blood transfusions and inadequate iron chelation therapy. Infection is the major cause of morbidity and mortality in thalassemia, so regular screening is required for this disease.

Material and Methods: The investigation was carried out in the Pediatrics department of Tertiary care Hospital of India. The study included 75 patients with thalassemia who were between the ages of 2 and 18 at the time of their diagnosis. According to the supplied proforma, a thorough history (age, sex, history of blood transfusion, history of iron-chelation therapy) was taken. After fully disinfecting the area with the cleaner, 4 ml of blood was drawn from a vein and allowed to clot. Routine tests such the complete blood count (CBC), renal function test (RFT), and liver function test (LFT) were performed using the separated serum. Transfusions and other measures, such as calcium supplements, FA, and iron chelators, were taken.

Results: There were 36 male and 39 female patients that were included in the study; the male to female ratio was 0.80:1. In the current investigation, RVD infection was more common than HCV infection (2.60%) and HCV infection (20.60%) (Table 2). Being an autosomal recessive illness, thalassemia is more prevalent in populations with high rates of consanguinity marriage. In the study conducted today, 4 patients experienced fever, 7 patients had a cough, 2 patients had headaches, and 2 patients had vomiting.

Conclusion: No patient had tuberculosis infection despite increased vulnerability to infections like tuberculosis in thalassemia due to defective immune response, blood transfusion, iron overload, splenectomy, and iron chelation therapy. Patients with thalassemia who frequently need blood transfusions require better methods of eliminating excess iron from their bodies. Therefore, to reduce growth retardation in patients with transfusion-dependent thalassemia, it is crucial to have an effective iron chelation therapy in addition to maintaining haemoglobin levels.

Keywords: Blood Transfusion, Chelation Therapy, Complete Blood Count, Thalassemia Major.

Introduction

Haemoglobin genetic abnormalities known as thalassemias are inherited. The primary cause of thalassemia worldwide is various globin gene mutations. With 50–60,000 new thalassemia patients being born each year, it is estimated that 1.5% of the population is a carrier for -thalassemia. Although it can also be found in populations in Africa, Southeast Asia, and the Middle East, -thalassemia is most common in the Mediterranean region. [1,2]

Over the past ten years, beta thalassemics who are transfusion-dependent have an average life expectancy that has reached the third and fourth decades. Better medical facilities have also changed the characteristics of young children who require blood transfusions. However, as life expectancy has increased, the disease's numerous consequences have come to light. These difficulties may partly result from the underlying disorder and partially from the standard therapy, which involved blood transfusions and a consequent iron excess. [3-5]

While those with beta thalassemia mild experience no symptoms and lead normal lives, those with beta thalassemia intermediate experience moderate anemia, and those with beta thalassemia major experience severe anemia and need blood transfusions. However, frequent transfusions can result in iron overload, which then causes a number of illnesses such as endocrine dysfunction,

cardiomyopathy, and liver disease, which ultimately cause people to pass away. However, patients with beta thalassemia major will die within the first five years of their lives if a blood transfusion is not done. [6,7]

The most precise approach of estimating iron overload is to estimate the direct liver iron content. However, this method wasn't available in our setup. The indirect method, which measures serum ferritin levels, is dependable, simple to use, inexpensive, and has no negative side effects. [8,9]

Bone marrow transplantation is a treatment option for beta thalassemia major, but it is difficult to cure these children due to a lack of resources, affordability, and awareness. As a result, the mainstay of their care today is routine blood transfusions and on-going iron-chelation therapy. [7]

Due to the high annual expense of thalassemia treatment in India, the majority of thalassemic children receive insufficient blood transfusions and inadequate iron chelation therapy. The purpose of this study is to test for blood born infections (HBsAg, HCV, and RVD) and lower the burdens of tuberculosis through active infection identification. [3,10] Through the screening of infections in a cohort of thalassemia patients, this study seeks to achieve this objective.

Materials and Methods

The cross sectional investigation that was conducted for a year is the subject of the current analysis. The investigation was carried out in the Pediatrics department of Tertiary care Hospital of India. Before the study began, the ethical clearance certificate was received after the study protocol was presented to the institute's ethics committee. The study included 75 patients with thalassemia who were between the ages of 2 and 18 at the time of their diagnosis. The universals sampling technique was used to determine the sample size. The following inclusion and exclusion criteria were used in the study:

Patients or the legal representatives of patients who were minors gave their written, informed consent to take part in the study, which was carried out in conformity with the Declaration of Helsinki's guidelines. Major-thalassemia was used as the inclusion criterion for all cases. Children with congenital disorders and chronic illnesses, such as cancer, tuberculosis, chronic hepatitis, congenital heart disease, chronic renal failure, epilepsy, and diabetes mellitus, were excluded from the study.

Children with other hemoglobinopathies, such as haemoglobin J variation, etc., and those with abnormal liver functions or infections present at the time of sample collection were excluded from the study. According to the supplied proforma, a thorough history (age, sex, history of blood transfusion, history of iron-chelation therapy) was taken. After fully disinfecting the area with the cleaner, 4 ml of blood was drawn from a vein and allowed to clot. Routine tests such the complete blood count (CBC), renal function test (RFT), and liver function test (LFT) were performed using the separated serum. Transfusions and other measures, such as calcium supplements, FA, and iron chelators, were taken.

Statistical analysis:

As a result, an excel sheet was created with all of the data. When necessary, appropriate tests were applied for analysis using the statistical package for the social sciences (SPSS) programme. Different parameters' means and standard deviations were computed. To find connections between viruses and research factors, two tests were used. At 0.05 levels, significance value was assessed using a 95% confidence level.

Results

A total of 75 patients who had been given a thalassemia diagnosis were included in the analysis. The included patients' ages ranged from 2 to 18 years, according to the findings. There were 36 male and 39 female patients that were included in the study; the male to female ratio was 0.80:1. According to the Kuppuswamy scale, there were only 37 patients in the lower socioeconomic class and 38 patients that belong to the upper socioeconomic class.

The study comprised 75 thalassemia patients in total. The oldest and youngest patients who were included were determined to be 18 years old and 2 years old, respectively. The age distribution of the patients that were included was as follows: there were 10 patients in the 2 to 5 year age range, 18 patients in the 6 to 9 year age range, 29 patients in the 10 to 13 year range, and 18 patients in the 14 to 18 year range. Most participants were between the ages of 10 and 13 years. None of the patients tested positive for HBsAg.

In the study conducted today, 4 patients experienced fever, 7 patients had a cough, 2 patients had headaches, and 2 patients had vomiting. No significant correlation between infection and other variables, such as splenectomy, S. ferritin, S. creatinine, SGPT, and BT need, was discovered in the current study.

In the current investigation, RVD infection was more common than HCV infection (2.60%) and HCV infection (20.60%) (Table 2). Being an autosomal recessive illness, thalassemia is more prevalent in populations with high rates of consanguinity marriage. Consanguinity was

identified in 40.8% of the patients in this investigation. Latent and active TB infections have not been discovered in any patients. Splenectomy is indicated when thalassemia major patient has a very large spleen causing physical discomfort associated with hypersplenism, an annual transfusion requirement of approximately 200 to 220 ml packed RBC/kg with a hematocrit of 80%, Greater than a 60% increase in consumption of PRBC over 6 months, clinically significant pancytopenia that persists after correction of vitamin B12 and folate deficiency.

Table 1: Socio-demographic data of the included patients

Socio demographic data	No. of patients
Male	36
Female	39
Upper middle	2
Lower middle	11
Upper lower	38
Lower	24

Table 2: Blood transfusion-related infection in thalassemia patient

Parameters	Reactive	Non-reactive	Total
RVD	2	73	75
HCV	17	58	75
HBsAg	0	75	75

Discussion

Patients with -thalassaemia, an inherited genetic condition, require regular transfusions to stay alive. The danger of infection transmission in these individuals is increased by frequent blood transfusions. Due to a lack of awareness and inadequate donor screening procedures, transfusion-transmitted infections have been a significant problem for patients with -thalassemia in poorer nations. [11,12]

The age of patients with -thalassemia in the current study was 11.5 8.8 years. Similar data from earlier research may be seen in the study by Alavi et al. [13], which indicated that patients with -thalassemia were an average age of 11.5 5.2 years. Similar to this, Ansari et al.'s study determined that the average age was 8.5 6.42 years. An average age of 9.7 5.2 was reported by Sanei-Moghaddam et al. All blood donations should be subject to extremely accurate and targeted HBV, HCV, HIV, and syphilis screening, according to WHO recommendations. There should be a regulatory framework for regulating the operations of blood transfusion services, including blood screening, and each nation should have a national programme for blood screening. [14] Patients with thalassemia frequently have blood transfusions, increasing their risk of developing blood transfusion-related infections like RVD, HBsAg, and HCV. In the current study, RVD infection was more common than HCV infection (22.66% vs. 2.66%). According to a study by Mishra et al. on 196 thalassemic patients, the seroprevalence of anti-HCV was high at 100 (51.1%), whereas that of anti-HIV 1/2 was low at 6 (3.1%) and HBsAg was high at 3 (1.5%).

Conclusion

The purpose of this study was to evaluate and screen patients with several transfusions of thalassemia for tuberculosis and other diseases (HBsAg, HCV, and RVD). No patient had tuberculosis infection despite increased vulnerability to infections like tuberculosis in thalassemia due to defective immune response, blood transfusion, iron overload, splenectomy, and iron chelation therapy.

Patients with thalassemia who frequently need blood transfusions require better methods of eliminating excess iron from their bodies. Therefore, to reduce growth retardation in patients with transfusion-dependent thalassemia, it is crucial to have an effective iron chelation therapy in addition to maintaining haemoglobin levels.

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