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AUDIT OF BETA-THALASSEMIA CASES AT NISHTAR HOSPITAL MULTAN

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ABSTRACT

Background: Beta-thalassemia is one amongst the common genetic disorders in our country. Therefore, it's consider necessary to review and probe it to cover all aspects of this disorder. Cooley's anemia is one amongst the foremost common genetic disorders in Pakistan and over 5000 new patients have added within the pool annually. This familial sickness has each medical and social implications, and thus there's a need to assess the magnitude of β-Thalassemia trait amongst members of the family of thalassemia major patients. **Objectives:** the target of the study was to seek out the distribution of β-thalassemia in several age groups, prevalence of transfusion connected microorganism infections and serum ferritin levels. **Methodology:** This cross-sectional descriptive study done at Nishtar Hospital Multan. Peripheral blood smears were studied for abnormal red blood cell morphology findings of microcytosis, hypochromia, tear drops, target cells and RBC indices i.e. haemoglobin, MCH, MCV, MCHC and Hb electrophoresis was done. All cases of thalassemia major and thalassemia intermedia were included in the study. Patients who were voluntarily participated were divided into three different age groups, i.e. 0-5 years, 5-10 years, and 11 years and above. All patients were screened for HCV, HBV, and HIV. Some of the cases were tested for serum ferritin levels. **Results:** We studied 250 cases, out of which 58% (145) were male and 42% (105) were female. Male to female ratio was 1.38:1. After doing getting detail history, examination and lab results we found that 235 (94%) patients were having thalassemia major and 6% (15) with thalassemia intermedia. 196 cases were from rural areas and 65 from urban areas. After studying viral markers we found that 45 cases were positive for anti-HCV antibody and 10 cases positive for HBsAg. The means of serum ferritin levels in thalassemia major (2564) and thalassemia intermedia cases (1674). **Conclusion:** Majority of patients were suffering from β-thalassemia major, because we selected cases having clinical sign and symptoms of disease and thalassemia intermedia patients may present normal and was missed during selection. Patients suffering from β-thalassemia major were mostly male, while in thalassemia intermedia sex distribution was almost equal. The overall prevalence of β-Thalassemia in rural areas was high. 18% of β-thalassemia patients were positive for anti-HCV antibody. There was significant difference in means of ferritin levels of thalassemia major and of thalassemia intermedia patients.

KEYWORDS: Beta Thalassemia, Thalassemia Major, Thalassemia Intermedia.

INTRODUCTION

Diverse mutations in globin genes are major cause of thalassemia, which are considered major genetic disorders worldwide.[1] Beta-thalassemia probably is the most common single gene disorder causing a major genetic health problem around the globe. According to a study almost 70,000 infants are born with β-thalassemia worldwide each year, and 270 million people are carriers of haemoglobinopathies.[2-3] β-thalassemia is most commonly present among populations in all Mediterranean countries, as well as in Southeast Asia, India, Africa, Central America and the Middle East2. Another study done in KPK showed that the prevalence of beta thalassemia in Pathan population was 7.96% and in Punjabis it was 3.26%.[4] However no central registered data is available in Pakistan, but it is estimated that 5000-9000 children with β-thalassemia are born per year. About 9.8 million carriers are estimated in general population with a carrier rate of 5–7%.[5-6]

Trends of cousin marriages, high birth rate, low educational level, early marriages, lack of awareness, week basic health model has increase number of children with transfusion dependent. Sometimes patients may present with milder symptoms in β-Thalassemia intermedia, though have both mutant alleles.[7-8] Prenatal screening and genetic counselling can decrease the number of thalassemia cases. Identification of B-thalassemia carriers and provision of prenatal diagnosis

of homozygous conception followed by termination of pregnancy will allow couples at risk to avoid having children with B-thalassemia disease 9-10. We have focused on three parameters in this study i.e. blood transfusion related viral infections, urban-rural distribution and serum ferritin levels. The objective of this study was to show the distribution of Beta-thalassemia in among patient presented to Nishtar Hospital Multan, according to the age group, prevalence of transfusion related viral infections and serum ferritin levels.

METHODOLOGY

This cross-sectional descriptive study done at Nishtar Hospital Multan. Peripheral blood smears were studied for abnormal red blood cell morphology findings of microcytosis, hypochromia, tear drops, target cells and RBC indices i.e. haemoglobin, MCH, MCV, MCHC and Hb electrophoresis was done. All cases of thalassemia major and thalassemia intermedia were included in the study. Patients who were voluntarily participated were divided into three different age groups, i.e. 0-5 years, 5-10 years, and 11 years and above. All patients were screened for HCV and HBV. Some of the cases were tested for serum ferritin levels. Anti-HCV and HBsAg testing were carried out by ICT method. The normal values of serum ferritin were taken as (range) 40-350 µgm/L for males and 14-150 µgm/L for females. The data was entered and analyzed by using SPSS version 16.

RESULTS

We studied 250 cases, out of which 58% (145) were male and 42% (105) were female. Male to female ratio was 1.38:1. After doing getting detail history, examination and lab results we found that 235 (94%) patients were having thalassemia major and 6% (15) with thalassemia intermedia. 196 cases were from rural areas and 65 from urban areas. After studying viral markers we found that 45 cases were positive for anti-HCV antibody and 10 cases positive for HBsAg. The means of serum ferritin levels in thalassemia major (2564) and thalassemia intermedia cases (1674).

Table I: Age and sex distribution of study subjects.

Age Groups (years)	Gender		Total
	Male	Female	
0 – 5	84 (33.6%)	56 (22.4%)	140 (56%)
5 – 10	40 (16%)	35 (14%)	75 (30%)
10 and above	21 (8.4%)	14 (5.6%)	35 (14%)
Total	**145 (58%)**	**105 (42%)**	**250 (100%)**

Table III: Blood transfusion related viral infections.

Viral infection	No. of cases
Anti-HCV positive	55 (19.4%)
HBsAg positive	10 (4%)

Table III: Serum ferritin levels.

Parameters	Thalassemia Major	Thalassemia Intermedia
Mean S. Ferritin (µgm/l) (x)	2775	1519
Number of cases (n)	235	15

DISCUSSION

In current study, the ratio between male-to-female thalassemia cases was almost 1.38:1. This was almost similar to a study carried out at Karachi that showed the prevalence ratio of thalassemia major in male-to-female as 1.4:1.[11] In another study carried out at Isfahan, Iran, the above ratio was exactly the same, i.e. 1.38:1 for thalassemia major.[12] In current study, 94% cases were registered as of thalassemia major and 6% as thalassemia intermedia. Thus the prevalence ratio between thalassemia major and thalassemia intermedia was 15:1. The prevalence rate for thalassemia intermedia was 10.38% in Bengal, India.[13] The male-to-female ratio in thalassemia intermedia patients was almost 1:1 in our study. Findings of Hassan M Yaish, Et, al,[13] showed equal sex distribution in thalassemia intermedia.

The rural to urban ratio of thalassemia was 2.8:1 in our study. This was most probably due to the more happening of cousin marriages in rural areas. Thalassemia is a hereditary disorder which runs in families.[14] However, ignorance, poor maternity care and illiteracy in rural areas could be the important contributory factors which need immense consideration. This can be reduced by nationally conducted screening/population surveys and establishing special care centers in major cities where facilities for diagnosis, genetic counselling, prenatal diagnosis and genetic studies using modern techniques of restriction enzyme analysis and treatment and health education of population are available.

The prevalence rate of HCV infection in βthalassemia cases in present study was 18%. A study conducted in 2014 showed prevalence rate of HCV in healthy subjects was 3.5%.[15] However, the prevalence rate in current study was remarkably lower than those of found in other studies in Pakistan.[16] A study conducted in Lahore showed prevalence rate of HCV infection in thalassemia patients was 41%.[16] Another study was conducted at Government College University, Faisalabad which revealed that anti-HCV antibody was positive in 65% beta thalassemia patients.[17] In Egypt in 2012, the anti-HCV positivity in β-thalassemia patients was 19.5%.[18]

For HBV transfusion related infection, the prevalence rate in current study was 4%. According to a study conducted in Rawalpindi the HBV prevalence rate was 3%.[12] In Indian study the transfusion related HBV infection in β-thalassemia patients was 9%.[13] In present study, serum ferritin levels were remarkably higher in thalassemia major patients with gross variations. The

higher levels of serum ferritin were recorded in the middle age group (5 – 10 years) in both male and female cases. The mean serum ferritin level in thalassemia major cases was 2564 µgm/l. The result was comparable with a study conducted at Rawalpindi in 2004 in which mean serum ferritin level was found to be 3390 µgm/l.[16] The mean serum ferritin level in thalassemia intermedia these cases was 1674 µgm/l. An Indian study conducted in 2014 which showed the mean serum ferritin level as 486.54 ± 640 µgm/l.[17]

CONCLUSION

Majority of patients were suffering from β-thalassemia major, because we selected cases having clinical sign and symptoms of disease and thalassemia intermedia patients may present normal and was missed during selection. Patients suffering from β-thalassemia major were mostly male, while in thalassemia intermedia sex distribution was almost equal. The overall prevalence of β-Thalassemia in rural areas was high. 18% of β-thalassemia patients were positive for anti-HCV antibody. There was significant difference in means of ferritin levels of thalassemia major and of thalassemia intermedia patients.

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