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Pediatrics

CUTANEOUS LESIONS AMONG SOUTH INDIAN CHILDREN WITH BETA THALASSAEMIA MAJOR.

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ABSTRACT

Objective: To study the cutaneous lesions among south Indian children with Beta thalassaemia major .

Design: Observational, hospital based study.

Method & Material: 67 children between age 1 year to 14 years with diagnosis of beta thalassaemia major were followed up at Paediatric hematology clinic over 4 years for cutaneous lesions.

Results: The common skin lesions were pruritus(49.2%),xerosis (40%) and hyper pigmentations (37.3%).

KEYWORDS : Beta thalassaemia major, children, cutaneous lesions, pruritus, hyper pigmentations.

Introduction

Thalassaemia syndromes are among the most common genetic disorders. Each year, more than 50 000 new patients are born with a severe form of thalassaemia worldwide, especially in the Mediterranean, Middle East, Indian Peninsula and Southeast Asia[1,2] It is believed that the partial resistance of thalassaemia carriers against malaria accounts for its high gene frequency in these tropical regions.[3]

While improvement in the care and quality of life (QOL) of patients with thalassaemia has been remarkable in developed world, unfortunately most of the patients with thalassaemia are born in countries with very limited resources .Indeed, thalassaemia imposes significant burden on healthcare systems of endemic regions so that the lifetime costs of this chronic disease can be beyond the capacity of some resource-poor countries. It is estimated that only 12% of patients with transfusion-dependent β-thalassaemia are properly transfused and of those <40% have access to adequate iron chelation.[2]The burden of thalassaemia on the healthcare system is expected to increase even further in these countries due to a reduction in childhood and infant mortality from communicable diseases, malnutrition and other causes, as a result of improved public health measures.

There are functional and physiological abnormalities in various organs due to progressive, significant anemia and overload of iron due to hemolysis. Although skeletal, cardiac and endocrinological complications in beta thalassaemic children are well known, the cutaneous manifestations are less studied. There is paucity of data regarding skin lesions among children suffering with beta thalassaemia major .Hence our study to determine cutanious complications in children with beta thalassaemia major.

Materials and methods

The study comprises 67 children with diagnoses of transfusion dependent beta thalassemia major over period of 4 years at Pediatrics Hematology clinic in our institute. All 67children were diagnosed as Beta thalassaemia major based on clinical features, hematological parameters as well as HPLC Hemoglobinometry criteria. Children who had pre existing skin disorders before diagnosis and those who refused to participate in study were excluded. All children were regularly followed up at our hematological clinic by Principal investigator and single dermatologist was consulted for skin lesions. The following data regarding age, sex, clinical features, age on first transfusion, frequency of transfusion and type of chelation therapy were entered on Performa. Presence of organomegaly , skeletal deformity and cardiac involvement were also recorded . The blood counts, serum ferritin were monitored as per institute's protocol. The regular chelation therapy is considered if the child had been on chelation drugs for last 6 months.

The Institutional ethical committee clearance was received. The informed consent was obtained from parents or guardians of study group.

Results

The study group included 67 children with beta thalassaemia major. Among them boys were 41(61.1%) and girls were 26(38.8%). The age group varied from 1 year to 14 years. Clinical anemia was found in 60 (89.5%) children, hemolytic facies in 57 (85%) ,Hepatosplenomegaly in 59 (88%) children .Among study group,60 (89.5%) children were on regular chelation therapy with 36(53.7%) children on Deferasirox and 20(29.8%) children on Deferiprone. Mere 10 children (14.9%) underwent splenectomy to reduce transfusion frequency.

Demographic and laboratory characteristics of study group.

Parameter	Mean ± SD
Age (years)	5.8 ±2.3
Weight (kgs)	13.6±6.4
Age of onset of blood transfusion (years)	1.5±0.8
Age of onset of chelation therapy (years)	2.9±1.2
Hemoglobin (g/dl)	5.2±0.71
Serum Ferritin (ng/ml)	2240±600

Distribution of cutaneous lesions among study group (N 67)

Cutaneous lesions	N (%)
Pruritus	33(49.2%)
Xerosis	27(40%)
Hyperpigmentations	25(37.3%)
Pityriaisis alba	16(23.8%)
Scars	13(19.4%)
Contact dermatitis	10(14.9%)
Urticaria	9 (13.4%)
Tinea	7(10.4%)
Hyperhiddrosis	4(6%)
Impetigo	3(4.4%)

Discussion

The improved standard of care regarding frequent blood transfusions coupled with better availability of iron chelation drugs and stem cell therapy have contributed for increased life span of children with beta thalasasemia major all over the world. Hence increased incidence of known and hitherto unknown complications among them.

The most common cutaneous lesion among our study group was pruritus 49.2%. Sameh SF et[4] had reported the prevalence of 37% among Egyptian children and 37.2% among Turkish children by Dogramaci et al[5] . The proposed hypothesis was stimulation of mast cells by cutaneous iron deposit to release histamine causing pruritus. Xerosis (40%) was next common cutaneous lesion among ours study group where as it was 22%[4] in Egyptian children and 34% among Turkish study group.[5]The prevalence of xerosis depends on prevailing environmental conditions.

Generalized hyper pigmentations were third most common finding accounting for 37% of children. Fekri et al [6] reported frequency of hyper pigmentation 65.3% in their study and Sameh SF et al at 31.5%.[5] The study carried out Egyptian children with beta Thallasemia major shown prevalence of pityriasis alba 18.5% where as our study group reported 23.8%. It was believed to be related to the deficiency of serum copper and other minerals in thalassemia major children leading to inadequate functioning of melanocytes.[7] .The prevalence of other cutaneous lesions like contact dermatitis , urticaria

Tinea and impetigo were found at 14.9%,13.4%,10.4% and 4.4% respectively among our children. This could be explained by the various immunological abnormalities like splenic dysfunction, iron deposits in reticular system and generation of auto RBC antibodies due repeated blood transfusion in children suffering with beta thalassaemia major. These children also suffer with malnutrition.

Our study had following limitations: 1 we did not correlate serum ferritin level and other micro nutrients like copper, selenium and zinc levels and cutaneous lesions due to economical constrain .2 We did not study skin lesions due chelating agents themselves in our study. 3 We did actively search the skin lesions among study population during follow up period.

Conclusion

The children suffering with transfusion dependent beta thalassemia major had various skin lesions among them most common were pruritus, xerosis and hyper pigmentations. The awareness of such skin lesions prompt us to actively seek them during follow up. Hence early remedial treatment which will help in better quality life for children.

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