Effect of Splenectomy on Blood Picture in Children with Thalassemia Major

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Abstract

Background: Beta thalassemias are a group of inherited autosomal recessive hematological disorders, it results in red blood cell destruction with symptoms of anemia. Red blood cell destruction, repeated blood transfusion and bad compliance to routine use of iron chelator lead to iron accumulation in the heart, liver and endocrine organs, Clinical picture of this disease is microcytosis and hemolytic anemia that need repeated blood transfusion which may lead to irreversible damage to organs and tissues due to iron accumulation, about 90% of patients with thalassemia had undergone splenectomy by age 15 years with a result in stoppage of transfusion requirement in most thalassemia intermedia patients and with a decrease in the need for red cell transfusions in thalassemia major patients.

Aim of study: To study the effect of splenectomy on different blood picture parameters in children with β- Thalassemia Major.

Methods: A prospective study that was conducted on 40 confirmed homozygous β- thalassemia major patients before and after splenectomy with complete physical examination was performed for all patients by assessing anthropometric measurements, vital signs, presence of pallor, jaundice, liver status. Complete blood picture with blood indices by Coulter Counter in addition to reticulocytes count.

Results: After splenectomy there is higher mean level of hemoglobin with 5.3% increase, higher platelet count with 110% rise in the mean platelet count and higher total leucocytic count with 42.5% increase and decreased reticulocytic count with a drop of 41% in the mean level of reticulocytes with all results statistically highly significant.

Conclusion: There is improvement of blood picture parameters after splenectomy which includes increasing of hemoglobin level and platelet count with decreased level of reticulocytic count. The increase of total leucocytic count after splenectomy is explained by the increased rate of infection.

Key words: Thalassemia Major – Blood parameters \_ children- Splenectomy.

تأثير استئصال الطحال على صورة الدم لدى الأطفال المصابين بأنيميا البحر الأبيض المتوسط

المستخلص العلمى

الخلفية العلمية: أنيميا البحر الأبيض المتوسط هي مجموعة من الاضطرابات الوراثية الجسمية بسبب عيوب في تخليق سلاسل الهيموجلوبين ، ينتج عنها تدمير خلايا الدم الحمراء مع أعراض فقر الدم , وعمليات نقل الدم المتكررة والامتثال السيئ للاستخدام الروتيني للأدوية الطاردة للحديد إلى تراكم الحديد في القلب والكبد وأجهزة الغدد الصماء . و فى حوالي 90٪ من مرضى الثلاسيمية يخضعوا لعملية استئصال الطحال عند بلوغهم عمر 15 سنة مما يؤدى إلى توقف متطلبات نقل الدم في معظم مرضى الثلاسيميا الوسطية مع انخفاض الحاجة إلى نقل خلايا الدم الحمراء في مرضى الثلاسيميا الكبرى.

الهدف من البحث: دراسة تأثير استئصال الطحال على عناصر صورة الدم المختلفة لدى الأطفال المصابين بأنيميا البحر الأبيض المتوسط .

المرضى واجراءات البحث: تم إجراء دراسة استطلاعية أجريت على 40 مريض مصابا بأنيميا البحر الأبيض المتوسط بعد تأكيد التشخيص وتماثل الجينات المرضية قبل وبعد استئصال الطحال. وتم عمل صورة الدم الكاملة مع مؤشرات الدم بواسطه الفحوصات وأجهزة التحليل المعملى بالإضافة إلى عدد الخلايا الشبكية.

النتائج: بعد استئصال الطحال هناك مستوى أعلى من الهيموجلوبين مع زيادة بنسبة 5.3٪ وارتفاع في عدد الصفائح الدموية بنسبة 110٪ في متوسط ​​عدد الصفائح الدموية وزيادة عدد كريات الدم البيضاء مع زيادة 42.5٪ وانخفاض عدد الخلايا الشبكية مع انخفاض بنسبة 41٪ في المتوسط مستوى الخلايا الشبكية وجميع النتائج ذات دلالات احصائيه عاليه.

التوصيات:- لوحظ من خلال الدراسة زيادة في عدد كرات الدم البيضاء الكلي بعد استئصال الطحال ووجود خطورة بالاصابه بالعدوى وذلك فينصح باخذ التطعيمات اللازمة قبل استئصال الطحال و الأخذ بسبل الحمايه من العدوى.

الخلاصة:- يوجد تحسن بمعظم عناصر صورة الدم وتشمل زيادة مستوى الهيموجلوبين وعدد الصفائح الدموية مع انخفاض مستوى عدد الخلايا الشبكي و هناك زيادة في عدد كريات الدم البيضاء الكلي بعد استئصال الطحال ويمكن تفسير ذلك من خلال زيادة خطر الأصابه بالعدوى .

الكلمات الدالة: أنيميا البحر الأبيض المتوسط – عناصر صورة الدم - الأطفال - استئصال الطحال.

Introduction

Beta thalassemias are a group of inherited autosomal recessive hematological disorders spread in the Mediterranean region due to defects in synthesis of β chains of hemoglobin, caused by mutation in the HBB gene on chromosome 11 causing asymptomatic to clinically severe hypochromic microcytic anemia (Hayder and Ali, 2017).

Beta thalassemia major is a hereditary synthesis defect in beta chains of hemoglobin; it results in red blood cell destruction with symptoms of anemia. Red blood cell destruction, repeated blood transfusion and bad compliance to routine use of iron chelator lead to iron accumulation in the heart, liver and endocrine organs (Shodikin et al., 2016).

Beta thalassemia major present in various shapes depending on the beta globin chains defect. The most severe form is beta thalassemia major which happens as a conclusion of inheritance of two beta globin chain mutations either in homozygous or compound heterozygous states. Patients with beta thalassemia major require recurrent blood transfusions for survival due to severe anemia. Clinical picture of this disease is microcytosis and hemolytic anemia that need repeated blood transfusion which may lead to irreversible damage to organs and tissues due to iron accumulation (Tolba et al., 2015).

Splenomegaly in patients with thalassemia major is often huge and mainly associated with severe transfusion dependent anemia due to ineffective erythropoiesis and prolonged hemolysis due to RBC membrane affection associated with precipitation of unbound α globin chains., about 90% of patients with thalassemia had undergone splenectomy by age 15 years with a result in stoppage of transfusion requirement in most thalassemia intermedia patients and with a decrease in the need for red cell transfusions in thalassemia major patients. However, these improvements came at the expense of various splenectomy associated complications including sepsis from encapsulated organisms (although this risk has been overcome with new protein conjugate vaccines and antibiotic prophylaxis) (Neal et al., 2013)

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Aim of study

To study the effect of splenectomy on different blood picture parameters in children with β- Thalassemia Major.

Patients and methods

This prospective study was conducted on 40 children (males and females) between 6- 18 years of age with confirmed homozygous β- thalassemia major diagnosis recruited from Hematology Clinic, Childrens university hospitals; Ain Shams and Cairo Universities, Egypt, from December 2016 till December 2017, before and after splenectomy done to those patients. Informed consent was taken from all patients or their legal guardians. excluded patients with acute febrile illness within 72 hours prior to enrollment, serious concurrent illness, chronic renal failure and other hematologic disease comorbidity. Data were collected by reviewing medical records as well as by direct patient interviewing. Thorough history taking was obtained including age, sex, consanguinity, other affected siblings, similar family condition, history of disease related complications e.g. transfusion-related infections, allow sensitization, iron-overload, related cardiac, endocrine and liver disturbances etc, history of concomitant medical conditions e.g. viral hepatitis (HBV, HCV), blood transfusion history including age of onset, duration and frequency of transfusion and history of drug therapy e.g. hydroxyurea, chelation. A complete physical examination was performed for all patients by assessing anthropometric measurements, vital signs, presence of pallor, jaundice, liver status. Complete blood picture with blood indices by Coulter Counter in addition to reticulocytes count. Patients' data were analyzed using SPSS 17.0 for windows 7. Quantitative variables were expressed by mean and SD (Standard deviation), compared using unpaired t-student test and Mann-Whitney test. Spearman rank order test was used for correlating quantitative variables. P value was considered to be significant if less than 0.05.

Results

Table (1) Distribution of gender and age of studied patients

|  |  |  |
| --- | --- | --- |
| N=40 | No | % |
| Gender  Male  Female | 29  11 | 72.5  27.5 |
| Age  Less than 10  10-  15> | 5  14  21 | 12.5  35.0  52.5 |
|  | Mean SD | Range |
| Age (years) | 13.8 3.0 | 7.0-17.0 |

This table shows that 72.5% of the studied patients are male and 27.5% are female with 12.5% of the patients are less than 10 years, 35% of the patients in the age range of 10 to less than 15 and 52.5% of the patients are 15 and above.



Table (2) Comparison between the mean level of blood picture parameters before and after splenectomy among studied patients

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| N=40 | Before  Mean SD | After  Mean SD | Paired t | P |
| Hemoglobin | 7.5 0.3 | 7.9 0.3 | 13.8 | 0.000\*\* |
| WBCs | 8.7 2.7 | 12.4 2.3 | 11.5 | 0.000\*\* |
| Platelets | 203.5 56.1 | 427.6 90.5 | 20.4 | 0.000\*\* |
| Reticulocytes | 8.7 3.0 | 5.1 2.3 | 15.1 | 0.000\*\* |

\*\* P<0.01 highly significant

There is higher mean hemoglobin after splenectomy compared to presplenectomy level and the difference is statistically highly significant with 5.3% increase. There is a higher means increases in the mean WBCs 12.4 compared to 8.7 before surgery and the difference is statistically highly significant with 42.5% increase. This table shows higher mean platelets after surgery 427.6 compared to 203.5 before surgery with 110% rise in the mean platelet count and the difference is statistically highly significant. There is a lower mean reticulocytic count after splenectomy compared to pre-surgery level with a drop of 41% in the mean level of reticulocytes and the difference is statistically highly significant.



Discussion

Forty patients 29 (72.5%) males and 11 (27.5%) females were included in our study. All were children aged 6 to 18 years with 12.5% of the patients are less than 10 years, 35% of the patients in the age range of 10 to less than 15 and 52.5% of the patients are 15 and above all with

established diagnosis of β- Thalassemia Major, the previous studies were done on adults (Darzi et al., 2014). To our knowledge, our study was the first to discuss the influence of splenectomy on different blood picture parameters in children with β- Thalassemia Major.

Regarding blood characteristics, the hemoglobin level is below the normal physiological level in all children with β- Thalassemia Major before and after splenectomy it is probably due to the fact that these patients are subjected to a state of chronic anemia that is caused by frequent hemolysis. On the other hand, there is higher mean hemoglobin after splenectomy was 7.9 g/dl compared to presplenectomy level 7.5 g/dl and the difference is statistically highly significant with 5.3% increase which agrees with previous studies (Ammar et al., 2014).

In our study there is higher mean platelets after surgery 427.6 compared to 203.5 before surgery with 110% rise in the mean platelet count and the difference is statistically highly significant and There is a higher mean increases in the mean WBCs 12.4 compared to 8.7 before surgery and the difference is statistically highly significant with 42.5% increase this may be explained by the fact that the spleen is the organ removing aging or abnormal RBC, foreign invaders, and other cells including platelets, WBCs from the circulation. Splenectomy raises circulating platelets and WBC, which may contribute to a high risk of thrombosis and certain infections including meningitis, pneumonia and sepsis which agrees with previous studies (Ruchaneekorn et al., 2013, Cappellini., 2007 and Vento et al., 2006).

It was reported in our study that there is a lower mean reticulocytic count after splenectomy compared to presplenectomy level with a drop of 41% in the mean level of reticulocytes and the difference is statistically highly significant, suggesting the role of spleen in reticulocyte pooling which comes in agreement with previous studies done by Ruchaneekorn et al., 2013 and Khuhapinant et al., 1994.

Conclusion

Ther is an improvement of blood picture parameters includes increasing of hemoglobin level and platelet count with decreased the level of reticulocytic count. There is increase of total leucocytic count after splenectomy with the increased risk of sepsis and infection after splenectomy with the necessity of presplenectomy vaccination.

Recommendation

we noticed that there was increase of total leucocytic count after splenectomy with the increased risk of sepsis and infection after splenectomy with the necessity of presplenectomy vaccination and to perform the ways of prevention of infection with early treatment of infection post splenectomy.

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