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CASE STUDY

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Shrinkage of Spleen in Sickle Cell Thalassemia: A rare case report

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Abstract

The most prevalent monogenic gene disorder caused by defective hemoglobin in the blood is thalassemia. Splenectomy (Total/partial) is considered to be the alternative treatment method based on hypersplenism or iron overload and is an inherited-autosomal-recessive disorder. In South Asia, the far east, the Middle East, and Eastern Mediterranean nations, it is more prevalent. Over 40,000 children are born with Thalassemia every year[1].

Keywords: ß-Thalassemia, Sickle Cell Thalassemia, Hypersplenism, Shrinkage of Spleen

1 | INTRODUCTION

HB E-ß-Thalassemia is more prevalent in South-Asia. (1) Approximately 6443 patients with Hb-E-beta thalassemia are born annually in Bangladesh with a prevalence rate of 30-40 percent. In the case of ß- characteristics, the carrier status is 3% and E-trait is 4% (2) . It is a condition with a Hb globin chain defect with fewer normal red blood cells and insufficient Hb to carry oxygen to the body. It produces fewer Hb proteins that are healthy and fewer red blood cells (3) . The most important diagnostic instruments for detecting ß-Thalassemia are the levels of hemoglobin (Hb) and Hb-electrophoresis. Hb levels drop & iron overload commonly occurs. Hb decreases with advancing age, infections, certain physiological conditions, and peculiarities. Due to increased intestinal absorption of iron caused by ineffective erythropoiesis, iron overload takes place (4) , (5) . With the availability of treatments such as blood transfusion and iron chelation, the quality of life (QoL) of patients has been enhanced (5) . Three clinical phenotypes have been categorized on a severity basis and more than 300 ß-thalassemia alleles have been identified1,6. Here

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we include a rare case presentation of ß-Thalassemia with Spleen shrinkage from 2 years of retrospective results, directing research into this significant disease in the near future.

2 | CASE PRESENTATION:

A diagnosed case of ß-Thalassemia (2012), a 19-year-old female Muslim girl admitted to Anwer Khan Modern Medical College Hospital on 28 February 2018 with symptoms of exhaustion for 5-6 months, generalized weakness for 5 months, yellowish eye discoloration for 2-3 months. On examination; - pale, dyspneic, icteric, T-102°F, RR-18 breath/min, Pulse-92 b/min, BP-110/80 mm Hg, no regional lymphadenopathy but liver (+5.5 cm> +6 cm from central margin) and mild-splenomegaly (+7.4 cm > +7 cm from coastal margin) with hypochondrial tenderness and H/O cholecystectomy (2014) and HCV-Positive (2014) and HCV-Positive) (2016). No abnormality is disclosed through other systematic review. Anisochromic microcytic anemia with a full blood count (Hb: 6.4 > 7.4 > 6.90 > 10.2 mg/dl, PBF- Spherocytosis & tear-drop cells), S. ferritin increased (3834 > 1439 > 3429 > 1528 ng/ml), SGPT increased (85 > 48 > 63 > 90 U/L), S. ferritin increased (85 > 48 > 90 U/L), PT-Normal(12 sec>13 Sec>12 sec), INR-1.10. Moderate-Hepatomegaly, mild-splenomegaly & non-visualized gall bladder on USG of Entire Abdomen (Splenic pole to pole diameter-18.8>16.8>16.4 cm; Hepatic line-18.8>19.7>19.9 cm; Pancreas A/P diameter-1.5>1.5>1.5 cm).

Treatment:

Previous studies on Hb-Electrophoresis reported that it is a big case of Hb-E ß-Thalassemia. So, she was treated with iron chelation, 2 units of PRBC (Inj. Desferol). The patient was discharged with considerable improvement with the advice of—

- Neutropenic diet,
- Tab. Deferasirox 500 mg
- Cap. Hydroxiurea 500 mg
- Tab. Folic Acid 5 mg
- Tab. Vit-B complex +Zinc
- Tab. Calcium-Vit-D3
- F/U visits monthly

3 | DISCUSSION

In Thalassemia, the destruction of RBC leads to bilirubin being excreted by liver metabolism. Complex pathophysiology entails inadequate erythropoiesis, apoptosis, oxidative damage, and reduced survival of red cells, causing the life expectancy of patients without blood transfusion to be less than 5 years (6) (7) .In thalassemia and hypersplenism, spleenectomy eliminates the need for blood transfusions (8). Initial splenomegaly and splenic shrinkage over time was the most important clinical finding for this patient. Iron chelation therapy improves the life expectancy of patients with thalassemia. Iron deposition in various organs, such as the heart, liver and endocrine glands, causes damage, such as anemia, cardiovascular, reticuloendothelial and other organ dysfunction. It is possible to avoid monitoring of liver function and care for iron overload/underlying diseases due to viral hepatitis, iron overload, drug toxicity or other causes (9) . There is very little research on splenic shrinkage in patients with thalassemia. In Sickle beta thalassemia, subsequent splenic shrinkages lead to decreased HbF levels with increased intravascular sickling and elevated permanent sickle cell levels, etc (10) . A Nigerian study shows that in the age group of 10-16 years, 33.8 percent of patients find Splenomegaly and 15 percent in adult groups with Thalassemia. The research of Ghana advocates 15 percent of splenomegaly over 10 years, while 9 percent endorse Jamaican studies. Because of the complex splenic structure that causes stasis, anoxia, and sickle erythrocytes to be trapped within the splenic pulp and sinuses, splenomegaly contributes to marked red pulp congestion and blood sequestration. There are several infringements, chronic vaso-occlusion, fibrosis. Thus, due to the resultant siderofibrotic mass, also known as auto-splenectomy, regression of splenic size to the impalpable spleen.

Conclusion:

Our results show that spleen shrinkage occurs in patients with thalassemia, including hepatomegaly. Experience with one patient should, however, not be extended to others. A broad group of follow-up interactions of patients for further research and connections should therefore be written.

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Consent:

Written informed consent was obtained for the publication of this case study from the patient.

Conflict of Interest:

In the preparation of this manuscript, the writers declare that they have no conflict of interests or financial participation.

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