Open Access Case Report

A Case of Renal Artery Thrombosis in Thalassemia Major Patient

Hamid A Chatha[1]
Mohammad Q Zadran[2]
Thiagarajan Jaiganesh[3]

[1]Consultant Emergency Medicine, Tawam Hospital, Al Ain, UAE
[2]Specialist Emergency Medicine, Tawam Hospital, Al Ain, UAE
[3]Consultant Emergency Medicine, Tawam Hospital, Al Ain, UAE

Address for Correspondence
Dr Hamid A Chatha
Consultant Emergency Medicine, Tawam Hospital, Al Ain, UAE
hamidchatha@hotmail.com

Abstract

Thalassemia, an inherited hemoglobin disorder, is the most common hemoglobinopathy in the world. Despite recent advances in the medical field to understand its pathophysiology, the management of thalassemia remains a complex process. Thalassemia patients are susceptible to various complications, including thromboembolic events. More commonly occurring thromboembolic complications include deep venous thrombosis (DVT), portal vein thrombosis, pulmonary embolism, and stroke. However, renal artery thrombosis has never been reported. In this report, we present a case of renal artery thrombosis in a thalassemia major patients.

Key words: Thalassemia, Thrombosis, Renal Artery, Anemia

Introduction

Thalassemia, an inherited haemoglobin disorder, is the most common hemoglobinopathy in the world.[1] It has a high prevalence in Southeast Asian, the Mediterranean region, the Far East and Sub-Saharan Africa. This disorder causes an imbalance of Alpha/Beta-globin production that manifests as chronic hemolytic anemia, ineffective erythrocytosis, and iron overload. The severity of the clinical course divides thalassemia into 2 main types: Thalassemia intermedia (TI) and Thalassemia major (TM). Patients with TI have milder anemia whereas TM patients have severe anemia that requires a lifelong blood transfusion. Despite all the recent advances in the medical field to understand its pathophysiology, the management of thalassemia remains a complex process. Thalassemia patients suffer from many complications due to the nature of their chronic disease. Among all the complications, thromboembolic events such as deep venous thrombosis (DVT), portal vein thrombosis, pulmonary embolism, and stroke have been reported.[2] Here we describe a case of renal artery thrombosis in a thalassemia major patient. To the best of our knowledge, the renal artery thrombosis due to thalassemia has never been reported.

Case Report

A 22-year-old male, with known Beta-thalassemia major, presented to the emergency department with a one-day history of abdominal pain. The abdominal pain was associated with nausea and vomiting. He denied fever, diarrhea or urinary symptoms. The patient was on chronic blood transfusion and had iron overload. Three months before this presentation he had undergone splenectomy due to massive splenomegaly.

In clinical examination his vital signs were stable. On physical exam, he had mild to moderate lower abdominal tenderness without signs of peritoneal irritation. The patient's blood result revealed high white blood cell count of 14.4 X 10^9/L with high absolute neutrophils count of 10.1 X 10^9/L. The rest of the blood results were CRP 3.5 mg/L, Hemoglobin 104 g/L, Platelets 522 X 10^9/L, and Ferritin 2640 mcg/L. He had normal kidney functions and a negative urine dipstick.

Considering clinical examination and investigations, a computerized tomography scan (CT-scan) of the abdomen with contrast was requested to rule out any acute inflammatory process. The CT-scan revealed that the patient had complete occlusion of the left renal artery with non-perfusion of left kidney. The right kidney perfusion was intact. After reviewing his previous radiological investigations, it was found that he had normal left kidney perfusion 3 months ago. This case was discussed with the urology team, considering the left kidney was already atrophic, no surgical intervention was suggested.

Discussion

With the advances seen in the medical field, the life expectancy of thalassemia patients has dramatically improved. Unfortunately, this increase in patient's life expectancy has led to more disease-related

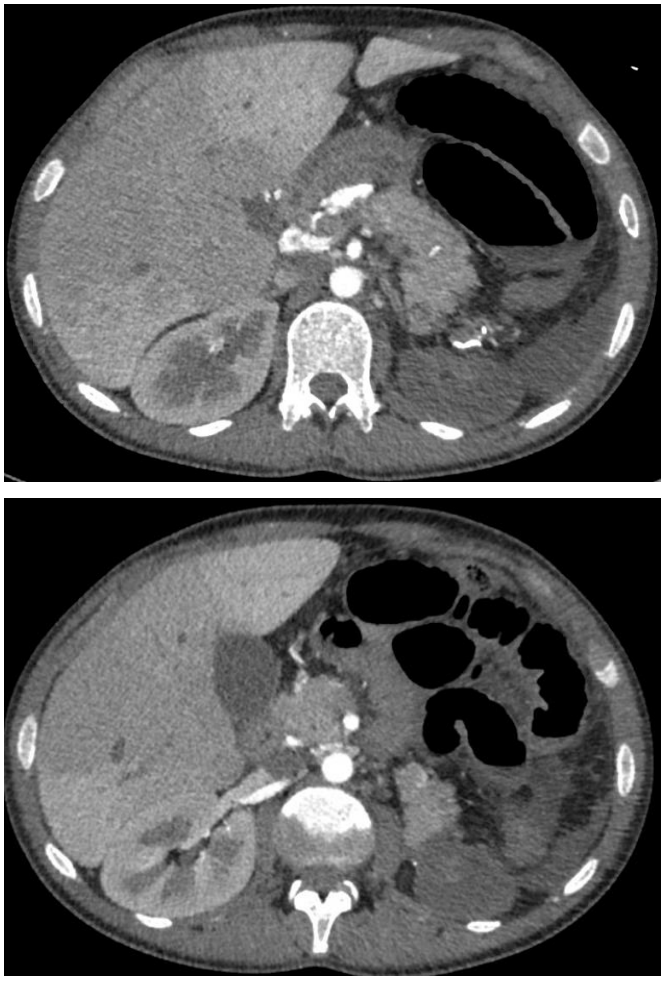

Figure 1. CT-scan images Illustrating complete occlusion of left renal artery with left renal atrophy.

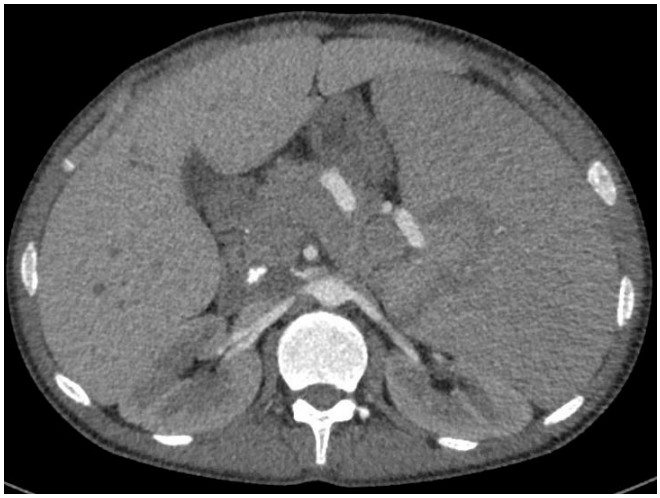

Figure: 2 CT-scan image that was obtained 3 months before above presentation which shows normal left kidney morphology with patent left renal artery.

complications. Thromboembolic events (TEE) are one of the common complications seen in Thalassemia patients. [3] The incidence of TEE has been reported to be 4-5%.[4] TI patients have milder anemia as compared to TM patients, hence blood transfusions are rarely required in TI, while on the other hand, TM patients need regular blood transfusions. Evidence in the last 2 decades has proved that TEE are more prevalent in TI than TM.[5] The onset of serious TEE is about 1 to 2 % in TM patients as compared to 5 % in TI patients.[6] Moreover, venous TEE are more common to occur than arterial TEE in thalassemia patients.[5,7] However, some data has shown that arterial TEE are higher in TM and venous TEE in TI.[5]

Taher et al, reported that chronic hypercoagulable state was found to be 4.38-fold higher in TI as compared to TM.[5,8] In general, the predisposing mechanisms for hyper coagulopathy in thalassemia patients include chronic platelet activation, abnormal expression of adhesion molecules in the vascular endothelium, membrane changes in RBCs, and disorder of the coagulation system.[9] Moreover, splenectomy is an important risk factor that leads to an increase in thromboembolic events. Splenectomy in thalassemia patients promotes a hypercoagulable state because of the decreased ability to scavenge procoagulant red blood cells and activated platelets.[10] Other risk factors were also found to contribute in increasing thromboembolism in thalassemia such as old age, cardiomyopathy, female sex, and diabetes.[5,11] Finally, patients with thalassemia who never received blood transfusions were also found to be at increased risk of developing thrombosis due to their high count of nucleated red blood cells (≥300 × 10^6 cells/L) and platelets (≥500 × 10^9 platelets/L).[7]

Despite the increased risk of thromboembolism in thalassemia patients, there is no available data that shows any benefit with the use of prophylaxis antiplatelet or anticoagulation therapy. A potential role of aspirin use has been suggested to reduce the risk of recurrence in patients after their first thromboembolic episode.[5] Some experts recommend the use of antiplatelets/anticoagulation in thalassemia patients post-splenectomy after doing risk assessment for VTE.[6] Due to the complexity of the disease and weak evidence in favor of thromboprophylaxis, it is important to consider individualized therapy by an expert physician.

As for the case reported, the patient was suffering from TM and had recently undergone splenectomy secondary to splenomegaly. He had high platelets and ferritin levels which further increased the risk for TEE.

He was not known to have any co-existing pathology which could have predisposed to renal artery occlusion.

During a thorough literature search, we did not find a similar case that has been reported. The aim of this case report is to highlight renal artery thrombosis as a complication in post-splenectomy thalassemia patients.

Conclusion

In conclusion, this case report serves as a reminder that thalassemia patients require ongoing monitoring and care, Physicians should have a high index of suspicion so that the diagnosis of such complications can be made early and focused treatment can be initiated, including rare events like renal artery thrombosis.

References

1. Martin A, Thompson AA. Thalassemias. Pediatr Clin North Am. 2013;60(6):1383–91. http://dx.doi.org/10.1016/j.pcl.2013.08.008
2. Taher A, Abou-Mourad Y, Abchee A, Zalouaa P, Shamseddine A. Pulmonary thromboembolism in β-thalassemia intermedia: Are we aware of this complication? Hemoglobin. 2002;26(2):107–12. Available from: http://dx.doi.org/10.1081/hem-120005447
3. Succar J, Musallam KM, Taher AT. Thalassemia and venous thromboembolism. Mediterr J Hematol Infect Dis. 2011;3(1):e2011025. http://dx.doi.org/10.4084/MJHID.2011.025
4. Sahin C, Basaran O, Topal Y, Akin F. A thalassaemia intermedia case with concomitant left atrial thrombus. BMJ Case Rep. 2014;2014:bcr2013203106–bcr2013203106. http://dx.doi.org/10.1136/bcr-2013-203106
5. Taher A, Isma'eel H, Mehio G, Bignamini D, Kattamis A, Rachmilewitz E, et al. Prevalence of thromboembolic events among 8,860 patients with thalassaemia major and intermedia in the Mediterranean area and Iran. Thromb Haemost. 2006;96(10):488–91. phttp://dx.doi.org/10.1160/th06-05-0267
6. Thalassemia.com: Treatment of thalassemia - thromboembolic disease. Thalassemia.com. https://thalassemia.com/treatment-thrombosis.aspx
7. Taher AT, Musallam KM, Karimi M, El-Beshlawy A, Belhoul K, Daar S, et al. Splenectomy and thrombosis: the case of thalassemia intermedia: Splenectomy and thrombosis in TI. J Thromb Haemost. 2010;8(10):2152–8. http://dx.doi.org/10.1111/j.1538-7836.2010.03940.x
8. Taher AT, Otrock ZK, Uthman I, Cappellini MD. Thalassemia and hypercoagulability. Blood Rev. 2008;22(5):283–92. http://dx.doi.org/10.1016/j.blre.2008.04.001
9. Singer ST, Kuypers FA, Styles L, Vichinsky EP, Foote D, Rosenfeld H. Pulmonary hypertension in thalassemia: association with platelet activation and hypercoagulable state. Am J Hematol . 2006;81(9):670–5. Available from: http://dx.doi.org/10.1002/ajh.20640
10. Taher AT, Weatherall DJ, Cappellini MD. Thalassaemia. Lancet. 2018;391(10116):155–67. http://dx.doi.org/10.1016/s0140-6736(17)31822-6
11. Haghpanah S, Karimi M. Cerebral thrombosis in patients with β-thalassemia: a systematic review: A systematic review. Blood Coagul Fibrinolysis . 2012;23(3):212–7. http://dx.doi.org/10.1097/MBC.0b013e3283502975