October 2017

Iron overload assessment in beta thalassemia major - is T2* magnetic resonance imaging the answer?

Natasha Ali
Aga Khan University, natasha.ali@aku.edu

Electronic Physician (ISSN: 2008-5842) http://www.ephysician.ir
October 2017, Volume: 9, Issue: 10, Pages: 5609-5610, DOI: http://dx.doi.org/10.19082/5609

Iron overload assessment in β Thalassemia Major – is T2* Magnetic Resonance Imaging the answer?

Natasha Ali

Assistant Professor of Haematology, Department of Pathology & Laboratory Medicine/Oncology, Aga Khan University, Karachi, Pakistan

Type of article: Letter to Editor

Abstract
This is a Letter to Editor and does not have an abstract.
Keywords: β Thalassemia, Iron overload, Magnetic Resonance Imaging

Dear Editor,

The initial reports in β thalassemia major were published by Lucarelli et al, spanning a decade and performing the procedure in over 1000 patients. An overall 20 years' thalassemia free survival was reported as 73% (1). It was in the 1980s when the Pesaro group recommended a classification for determining outcome of transplant in patients aged 17 years or less. The variables included: compliance to iron chelation, hepatomegaly and portal fibrosis as documented by liver biopsy. Based on the number of risk factors present, patients were grouped into class I (no risk factors) class II (any one or two risk factors) and class III (all risk factors). According to the categories, the disease free survival was 94%, 77% and 53% for class I, II, III respectively (2). The prognostication grid is very subjective with respect to the presence of liver enlargement and the adequacy of chelation. Moreover, not every patient is willing to undergo an invasive procedure like liver biopsy to determine the extent of portal fibrosis. Allogeneic stem cell transplantation remains to be the only curative treatment option in patients with β thalassemia major (3). The major predictor for a favorable outcome in allogeneic transplant for β thalassemia is the extent of iron overload prior to the procedure (4). T2* Magnetic Resonance Imaging (T2* MRI), is a non-invasive methodology which can be utilized to determine cardiac and liver iron overload (5). For cardiovascular iron overload, T2* MRI is a highly reproducible and sensitive technique. A study done from our center by Alvi N et al., classified 70% of the patients with Thalassemia major as NYHA (New York Heart Association) class II at presentation using this technique. T2* of <20 milliseconds was seen in 62.6% while 47% showed a value of 10 milliseconds or less. There was no correlation of serum ferritin with the value of T2* MRI (p value: 0.464) (6). A recent cross sectional study on 162 subjects with β thalassemia major was done to assess cardiac and liver iron overload simultaneously. Approximately 85% of the subjects had normal cardiac iron stores. However, 70.4% had severe liver iron overload. There was a weak correlation (r=0.28) between cardiac iron stores and serum ferritin levels. The liver iron concentration and serum ferritin showed a considerably more significant correlation (r=0.37) (7). As compared to several other studies, this study also demonstrated that liver siderosis precedes cardiac iron overload. T2* MRI of liver is a non-invasive modality in identifying the degree of iron deposition. Hamidieh AA et al. (8) has used T2* MRI of cardiac and liver in post-transplant patients with β thalassemia major. The pre and post-transplant values were compared and the technique proved to be beneficial in monitoring patients post-procedure. In summary, liver pathologies and inflammatory conditions falsely increase serum ferritin levels, making it an unreliable predictor of liver iron concentration. The gold standard is liver biopsy but it is an invasive procedure and cannot detect cardiac iron status (9). T2* MRI of cardiac and liver used in combination, is a non-invasive, reproducible diagnostic modality for prognostication of iron overload in patients with β thalassemia prior to undergoing allogeneic stem cell transplant. The previous classification requiring liver biopsy can be altered to utilize this technique as a replacement. Moreover,

Corresponding author:
Assistant Professor Haematology, Department of Pathology & Laboratory Medicine/Oncology, Aga Khan University, Karachi, Pakistan. Tel.: +923009202079, Email: natasha.ali@aku.edu
Received: May 16, 2017, Accepted: September 21, 2017, Published: October 2017
iThenticate screening: August 21, 2017, English editing: October 12, 2017, Quality control: October 14, 2017
This article has been reviewed / commented by Five experts

if performed in combination with cardiac magnetic resonance, the assessment of liver and cardiac iron can be done simultaneously.

Conflict of Interest:
There is no conflict of interest to be declared.

References:

1) Angelucci E, Baronciani D. Allogeneic stem cell transplantation for thalassemia major. Haematologica. 2008; 93(12): 1780-4. doi: 10.3324/haematol.2008.001909. PMID: 19050069.
2) Lucarelli G, Galimberti M, Polchi P, Angelucci E, Baronciani D, Giardini C, et al. Bone marrow transplantation in patients with thalassemia. N Engl J Med. 1990; 322(7): 417-21. doi: 10.1056/NEJM199002153220701. PMID: 2300104.
3) Angelucci E, Matthes-Martin S, Baronciani D, Bernaudin F, Bonanomi S, Cappellini MD, et al. Hematopoietic stem cell transplantation in thalassemia major and sickle cell disease: indications and management recommendations from an international expert panel. Haematologica. 2014; 99(5): 811-20. doi: 10.3324/haematol.2013.099747. PMID: 24790059, PMCID: PMC4008115.
4) Angelucci E. Hematopoietic stem cell transplantation in thalassemia. Hematology Am Soc Hematol Educ Program. 2010; 2010: 456-62. doi: 10.1182/asheducation-2010.1.456. PMID: 21239835.
5) Wood JC. Magnetic resonance imaging measurement of iron overload. Curr Opin Hematol. 2007; 14(3): 183-90. doi: 10.1097/MOH.0b013e3280d2b76b. PMID: 17414205, PMCID: PMC2892972.
6) Alvi N, Tipoo FA, Imran A, Ashraf MN, Qidwai A, Khursheed M, et al. Burden of Cardiac Siderosis in a Thalassemia-Major Endemic Population: A Preliminary Report From Pakistan. J Pediatr Hematol Oncol. 2016; 38(5): 378-83. doi: 10.1097/MPH.0000000000000574. PMID: 27164519.
7) Wahidiyat PA, Liauw F, Sekarsari D, Putriasih SA, Berdoukas V, Pennell DJ. Evaluation of cardiac and hepatic iron overload in thalassemia major patients with T2* magnetic resonance imaging. Hematology. 2017; 22(8): 501-7. doi: 10.1080/10245332.2017.1292614. PMID: 28218005.
8) Hamidieh AA, Tayebi S, Moeininia F, Shamshiri AR, Behfar M, Alimoghaddam K, et al. T2(∗) MRI changes in the heart and liver of ex-thalassemic patients after hematopoietic stem cell transplantation. Hematol Oncol Stem Cell Ther. 2014; 7(3): 103-8. doi: 10.1016/j.hemonc.2014.06.002. PMID: 24998835.
9) Eghbali A, Taherahmadi H, Shahbazi M, Bagheri B, Ebrahimi L. Association between serum ferritin level, cardiac and hepatic T2-star MRI in patients with major β-thalassemia. Iran J Ped Hematol Oncol. 2014; 4(1): 17-21. PMID: 24734159, PMCID: PMC3980017.