Thalidomide as a treatment for non-Langerhans cell histiocytosis with cardiac infiltration: a case report

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Introduction: Non-Langerhans Cell histiocytoses (non-LCH) are a group of disorders characterized by proliferation of histiocytes with absence of Langerhans cells. Majority of non-LCH manifest cutaneously, but may also have systemic manifestations such as the heart. In this case, we described our experience with thalidomide in treating an extracutaneous manifestation of non-LCH.

Methods: This is a case of a 35 year old male, diagnosed with non-Langerhans cell histiocytosis who presented with heart failure symptoms. 2D-Echo revealed a homogenous echogenic density measuring 5.57cm x 3.12cm with an area of 13.7cm2 from the base of the right atrium superiorly to the mid right ventricular free wall, inferiorly abutting the tricuspid annular area. A cardiac MRI was done revealing infiltrative intrapericardial enhancing soft tissue mass below the level of the right pulmonary artery, located between the SVC and ascending aorta, surrounding the right atrium and extending into the right AV groove. Patient was initially treated with thalidomide 200mg/day which was subsequently decreased to 50mg/day due to elevation of creatinine. A repeat 2D-Echo after 2 months showed a decrease in size of the echogenic density measuring 5.55cm x 2.63cm with an area of 12.3cm2. Medications were continued while a repeat 2D-Echo is scheduled upon follow-up. Erdheim-Chester disease, a specific form of non-LCH was also a consideration hence BRAF mutation testing will be done.

Result: Thalidomide is an immunomodulatory drug that is used for treatment of several inflammatory skin diseases including Langerhans and Non-LCH. Thalidomide acts by inhibiting the production of TNF-alpha and IL-6 hence its anti-inflammatory and antineoplastic properties.

Conclusion: In conclusion, our case suggests that thalidomide can be used for extracutaneous manifestations of non-LCH base on its partial regression. Further testing of thalidomide for treatment of extracutaneous non-LCH lesions is required to support the use of this treatment option.