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Riociguat Use in a Patient with Sickle Cell Anemia Related Chronic Thromboembolic Pulmonary Hypertension

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Introduction: Chronic thromboembolic pulmonary hypertension is a devastating complication of sickle cell anemia and it has limited treatment options. Riociguat is the only approved medical treatment modality in chronic thromobembolic pulmonary hypertension, but it has not been approved in sickle cell anemia related chronic thromboembolic pulmonary hypertension yet. Riociguat use in such sickle cell anemia related chronic thromboembolic pulmonary hypertension cases is so limited that to the best of our knowledge, only a recent case-series paper of 6 six sickle cell anemia related chronic thromboembolic pulmonary hypertension patients treated with riociguat showed an improvement in clinical and biochemical markers. We aimed to represent our first riociguat experience in a sickle cell anemia related chronic thromboembolic pulmonary hypertension patient to raise an awareness of a new treatment modality and the curative properties, tolerability, and possible adverse effects of the drug.

Case Presentation: A 59 years old man with history of sickle cell anemia had concomitant coronary artery disease and diabetes mellitus. He had history of deep vein thrombosis and pulmoner embolism confirmed with doppler ultrasonography and dynamic thorax computerized tomography respectively in 2013 and was treated with rivaroxaban over a period of 6 months. He was free of pulmonary symptoms for 4 years. He presented in 2017 with NYHA-III dyspnea on exertion for last 1 year. Echocardiogram revealed dilated right sided chambers with septal flattening and pulmonary arter hypertension. (SPAP: 105 mm Hg). Right heart catheterization was performed mean pulmonary arterial pressure of 55 mmHg. the six minute walk test distance was 240 meters and he was desaturated at the end of the test. He was diagnosed as sickle cell related chronic thromboembolic pulmonary hypertension with abnormal V/Q scan (multiple pulmonary segmental perfusion defects) with abnormal right heart catheterisation results and compatible echocardiography findings despite anticoagulation for six months. He was started on riociguat 0.5 mg three times daily with dose titration gradually to 2.5 mg three times daily. Repeated ECHO showed no remarkable change but his 6MWT was 350 meters after 1 year treatment. He is still under clinical stable follow-up, his dyspnea is NYHA-II. He has a good compliance with the drug and no adverse effect has been detected in the first year of treatment.

Conclusion: Riociguat use in sickle cell anemia related chronic thromboembolic pulmonary hypertension needs further larger, prospectively follow-up designed studies.

Keywords: Riociguat, chronic thromboembolic pulmonary hypertension, echocardiography