A STRANGE CASE OF PULMONARY HYPERTENSION

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PH is a haemodinamic and pathophysiological state that can be found in multiple clinical conditions. Chronic hemolytic anemia as sickle cell disease (SCD) is complicated by PH with 30% of prevalence and worse prognosis. PH in SCD is multifactorial including vasoconstriction, proliferation, obstructive remodelling of the pulmonary vessel wall; inflammation and thrombosis in situ play a role in the pulmonary vascular disease. We have observed a 44-yo woman affected by sickle cell who developed dyspnea (WHO-FC III) without chest pain or fever. Transthoracic echocardiography revealed moderate-severe tricuspid with a gradient of 70 mmHg with an assumed right atrial pressure of 20 mmHg estimated by inferior vena cava, right atrial and ventricular dilatation, reduced TAPSE 16 mm, Ryan index 0.8 with normal size and ejection fraction of left ventricular. Electrocardiogram provided right axial deviation. Computed angiography was normal and ventilation/perfusion lung scan showed an elevated probability for CTEPH. BNP was 921 pg/ml and lung diffusion capacity for carbon monoxide was slightly decreased. Prophylactic transfusion during crisis to decrease the frequency of these and new direct anticoagulants were started. After two weeks the patient was in WHO-FC II and transthoracic echocardiography showed a slight tricuspid regurgitation with a pressure gradient of 40 mmHg with TAPSE of 20 mm and Ryan index was normal. In the new guidelines SCD is moved into Group 5 and our patient was not subjected to specific therapy for pulmonary hypertension. DOAC has showed a reasonable alternative to VKA for VTE.

Figure 1. CT scan showing the presence of large pleural effusion, bilateral lung edema and a hypodense area (arrows) in the right hemithorax.

Figure 2. ECG showing sinus rhythm, complete right bundle branch block and RV strain pattern.