Favourable response to PAH specific therapies in pulmonary capillary hemangiomatosis

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Pulmonary Capillary Hemangiomatosis (PCH) is a rare disorder which in the current classification of pulmonary hypertension, PCH (group 1') is linked to but not part of PAH (group 1). In this report, we describe a case of PCH that was successfuly managed by an oral combination therapy with endothelin receptor antagonist and PDE5-I.

A 24-year-old woman was admitted to our department with progressive dyspnea (FC IV), cough and palpitations in the postpartum 3rd month. Echocardiography showed right ventricular overload, right heart chambers enlargement and D-shaped left ventricle, systolic pulmonary arterial pressure (PAP) 118 mmHg. There were no signs of rheumatologic, thromboembolic or systemic disease. Pulmonary function test results were normal, with the exception of very low DLCO (%30 predicted). High resolution computed tomography revealed diffuse centrilobular nodules and lobular ground glass opacities. Right heart catheterization (RHC): mean PAP (mPAP 63 mmHg), pulmonary vascular resistance (20 WU), right atrial pressure (RAP) 12 mmHg), PAWP (6 mmHg), cardiac index 1.5 lt/min/m2.

After volume control with diuretics, treatment with macitentan 10 mg/ day was started. About a month later, tadalafil was added to treatment and the the dose was increased to 40 mg with close monitoring. RHC at the fourth month of therapy: mPAP (44 mmHg), PVR (12 WU) and RAP (8 mmHg) PAWP(6mmHg). Her FC was class II and six minute walk distance was 372 m.

In conclusion, despite the abscence of histological diagnosis, with the computed tomography findings comparable to HCM, as a bridging therapy to lung transplantation, keeping in mind that PAH spesific therapy might be harmful watchful sequential combination revealed clinical and hemodynamic improvement in this patient.