International Conference 'Advances in Pneumology'
Bonn, 17-18 June 2011

PULMONARY ARTERIAL HYPERTENSION IN SARCOIDOSIS

S. Pabst[1], N. Grau[1], C. Hammerstingl[1], J. Kreuz[1], C. Grohé[2], D. Skowasch[1]

[1]Medical Clinic II, University of Bonn, Germany;
[2] Evangelische Lungenklinik Berlin-Buch, Germany

Background: Sarcoidosis is a systemic granulomatous disease with unknown aetiology. Lungs and lymph nodes are commonly affected. In up to 5% of the cases, a cardiac involvement may lead to serious complications such myocardial infarction, chronic heart failure and arrhythmias. Also cases of pulmonary hypertension (PH) and pulmonary arterial hypertension (PAH) are described. However, the exact prevalence of PAH in patients with sarcoidosis is unclear. **Patients and methods:** 111 patients with proven sarcoidosis were recruited in the outpatient unit of the Division of Pneumology, University Hospital Bonn, Germany, from January 2010 to October 2010. All patients were studied prospectively by transthoracic echocardiography (TTE) for the presence of PH. In case of a systolic pulmonary arterial pressure (PAPsyst) ? 30mmHg (plus estimated central venous pressure) PH was assumed and a right heart catheterization (RHC) followed if there were no other reasons for PH such as chronic heart failure, diastolic dysfunction or severe valve insufficiency. In the statistical analysis (SPSS 17), the echocardiographic and invasive haemodynamic data were correlated with the clinical course of sarcoidosis, chest x-ray, lung function testing and soluble interleukin-2 receptor as a biomarker for the activity of sarcoidosis. **Results:** In 23 of the 111 patients PH was assumed in TTE. Three patients presented with severe mitral insufficiency III° and IV°, in eight patients PH was supposed to be caused by chronic heart failure or relevant diastolic dysfunction, two patients declined undergoing RHC. Of the ten patients investigated with RHC four showed a precapillary pulmonary arterial hypertension (PAH, PAPmean ? 25mmHg and PCWP < 15mmHg), in one patient a postcapillary hypertension (PH, PAPmean ? 25mmHg and PCWP ? 15mmHg) was diagnosed and in five patients PH could not be confirmed (PAPmean ? 25mmHg). Three of the four patients with PAH had a radiologic stage III and IV. In three of the four patients with PAH a significantly reduced transfer factor for carbon monoxide (TLCO) < 45% as a surrogate for severe involvement of the lungs was found. All patients with PAH had a chronic course of sarcoidosis lasting ? 13 years. **Conclusion:** This is the first study prospectively investigating a large cohort of patients with sarcoidosis for the prevalence of PH and PAH. The prevalence of PAH was found to be at least 3.6% (4/111) and therefore exceeds the prevalence of PAH in the normal population by far. A chronic and progressive lung involvement due to sarcoidosis seems to be the most evident risk factor for developing a sarcoidosis PAH.