Pulmonary artery size as a predictor of outcomes in idiopathic pulmonary fibrosis

Pulmonary hypertension (PH) is a well-recognised complication of idiopathic pulmonary fibrosis (IPF) and is predictive of a worse outcome. The prevalence of PH in the setting of IPF has not been well described in the literature, but has ranged from 32 to 85%.[1,2] Recognising underlying PH in a patient with IPF is a challenging diagnostic dilemma because of the nonspecific clinical symptoms and unreliable non-invasive ancillary tests. The treatment of PH and the need for transplantation in patients with IPF is based on factors that include disease severity, functional status and the degree of hypoxaemia.[3,4] Given the high mortality rate and propensity for acute decompensation, PH in IPF patients should be evaluated early owing to the prognostic implications and potential need for transplant early in the disease course. Shin et al.[5] performed a retrospective review of 98 patients with IPF who attended a tertiary-care centre between 2008 and 2013. The objective was to establish whether there was an independent relationship between pulmonary artery size and outcomes (either lung transplant or death) over a 5-year period. The pulmonary artery and ascending aorta diameter ratio (PA:A) was calculated after being measured by high-resolution computed tomography (HRCT) of the chest. The independent influence of different variables on overall outcomes was evaluated using the Cox proportional hazards model. The patients were divided into two groups based on those with a PA:A ratio ≤1 and a PA:A ratio >1. The baseline characteristics were not significantly different between the two groups except for the forced vital capacity (FVC) % predicted, forced expiratory volume in 1 second % predicted and diffusing capacity of the lung for carbon monoxide (DLCO) % predicted. The mean pulmonary artery diameter and PA:A ratio were 32.8 mm and 0.94, respectively. Patients with a PA:A ratio >1 had higher risk of death or transplant compared with a PA:A ratio ≤1 (p<0.001). A PA:A ratio >1 was also an independent predictor of outcomes in the adjusted outcomes analyses (hazard ratio 3.35, p=0.002). This ratio also appeared to perform better in predicting outcomes than the well-established gender, age and physiology (GAP) index – the latter referring to pulmonary function parameters, including FVC and DLCO.[6] This study highlights that an HRCT PA:A ratio >1 in IPF patients is associated with worse outcomes and may be helpful in risk stratification and prognostication.

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