TORAKS 2020
10. kongres Hrvatskog torakalnog društva
10th Congress of the Croatian Thoracic Society
16.–19. rujan | September
Hotel Hilton Garden Inn, Zagreb

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FROM BAD TO WORSE: IDIOPATHIC PULMONARY FIBROSIS AND LUNG CANCER (IPF-LC)

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Objective:

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Introduction: Idiopathic pulmonary fibrosis (IPF) is the most common idiopathic interstitial pulmonary disease with a median survival of 2–4 years after diagnosis. IPF itself increases the risk of lung cancer (LC) development. A significant number of IPF patients have risk factors, such as the older age, a history of smoking or concomitant emphysema, which can predispose the patient to LC. There are multiple common genetic, molecular and cellular processes that connect lung fibrosis with LC, such as myofibroblast/mesenchymal transition, myofibroblast activation and uncontrolled proliferation, endoplasmic reticulum stress, alterations of growth factors expression, oxidative stress, and large genetic and epigenetic variations that can predispose the patient to develop IPF and LC. Although IPF-LC represents only a fraction of the IPF population, it is a dreaded complication. IPF-LC appears to have a unique phenotype – predominantly non-small cell lung cancer (NSCLC) in which adenocarcinoma is most prevalent, frequently located in the periphery of the lower lobes adjacent to areas of fibrosis – sometimes termed ‘scar-cinoma’. For the clinician, there are no recommendations for screening or for treatment. The current approved IPF therapies, nintedanib and pirfenidone, are also active in LC. In fact, nintedanib is approved as a second line treatment in NSCLC, and pirfenidone has shown anti-neoplastic effects in preclinical studies.

Case reports: We report our experience with patients who developed IPF and LC. In our Institute registry there are 64 patients with IPF. In three patients we have diagnosticed lung cancer, all of them were non-small cell lung cancer.

Conclusion: Many questions certainly remain for both researchers and clinicians.

Key words: idiopathic pulmonary fibrosis (IPF), lung cancer (LC), non-small cell lung cancer (NSCLC)

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