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Retrospective Analysis of DIPNECH and Carcinoid Tumorlets Progressing to Invasive Pulmonary Carcinoid Tumors

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BACKGROUND: There is a spectrum of lesions, ranging from noninvasive diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) to invasive carcinoid tumorlets, that are potentially precursor lesions to pulmonary carcinoid tumors. As rare entities, their diagnosis, prognosis and management remain unclear.

METHODS: A retrospective analysis was performed on patients from our institution with DIPNECH and/or carcinoid tumorlet from 2000 to 2020. Progression of pulmonary nodule(s) associated with DIPNECH/tumorlets to carcinoid tumor was defined radiographically as growth to a size > 5mm. DIPNECH diagnoses were based on either clinical-radiographic features or pathologic review.

RESULTS: A total of 69 patients was identified. Most patients were women (87%), elderly (median age 68 y/o; 44-86), Caucasian (62.3%) and Asian (13.0%); and 52.2% had a smoking history. Symptoms were most commonly cough (62.3%), shortness of breath (50.7%) and no symptoms (15.9%). Overall, 20 patients had at least DIPNECH and 62 patients had at least a tumorlet diagnosed. There was considerable overlap, with 37.8% of patients having more than one diagnosis along the disease spectrum and majority being found within the same pathologic specimen. A total of 10 (14.5%) patients had DIPNECH, tumorlet and carcinoid tumor all found in the same tissue. Treatments for DIPNECH/tumorlets included surveillance (44.9%), inhalers (18.8%), somatostatin analogs (7.2%) and oral steroids (7.2%). Eight (11.6%) patients with DIPNECH and/or tumorlet later developed carcinoid tumor by radiographic criteria, with a median follow up time of 4.7 years. These patients had an eventual pathologic diagnosis of carcinoid tumor.

CONCLUSION: DIPNECH and tumorlets are possible precursor lesions to pulmonary carcinoid tumors. This study is one of the largest examining these rare diseases to help guide future diagnosis and management. We found that these diseases are most common in older women who may or may not smoke, frequently co-occur, and over years may slowly progress to become carcinoid tumors.

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