P-17
DIPNECH: The Mayo Experience

Daniel Almquist[1]; Arriana Cabrera[1]; Heidi Kosiorek[2]; Mohamad Sonbol[2]; Thorvardur Halfdanarson[2]; Helen Ross[1]; Dawn Jaroszewski[1]; Harshita Paripati[1]

[1]Mayo Clinic Arizona; [2]Mayo Clinic

BACKGROUND: Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) is a pulmonary disorder with neuroendocrine cell proliferation associated with Lung NET. Optimal diagnostic and treatment strategies have yet to be well defined. Herein, we aim to describe the Mayo Clinic experience with DIPNECH.

METHODS: A retrospective analysis was performed of patients diagnosed with DIPNECH within Mayo Clinic between January 2000-Febuary 2019. Cases were identified from clinical databases at Mayo Clinic. Data on demographics, disease characteristics, time of diagnosis, surgery, and last follow up date were extracted. Extent of symptom burden, treatment approaches, disease progression, and disease-free survival (DFS) were evaluated by chart review.

RESULTS: A total of 59 patients were identified with a median age of 63(43-81) years. The cohort was predominantly female (93.2%) and non-smoking (76.3%). Most patients (86.4%) had symptomatic disease with chronic cough being the most common (71.2%) followed by exertional dyspnea (44.1%). Imaging typically showed bilateral lung nodules (93.2%) with mosaic attenuation noted 69.5% of the time. Surgical resection was frequently completed to confirm diagnosis (94.9%). Most patients received inhaled glucocorticoids combined with a beta agonist (79.7%). Oral steroid use was seen in 49.2% of patients whereas a somatostatin analog was used in 15.3% following the diagnosis of DIPNECH. These medical interventions led to symptom relief in 23.7% of the patients. The median follow up for all patients was 19.5months. Progression of tumorlets was seen in 48.7% of patients with only 7(17.9%) patients progressing to a diagnosis of carcinoid tumor. The 3-year DFS was 90.5%.

CONCLUSION: DIPNECH remains a rare disease more commonly diagnosed in women in their early 60's. DIPNECH appears to have an indolent course with obstructive symptoms being the most common finding. Overall, DIPNECH appears to have a low risk of progressing to carcinoid tumors based on the observation of this study.