Octreotide LAR in Patients with Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH): A Case Series

Elizabeth Byron, MD¹; and Jonathan Strosberg, MD¹

Background: Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) is a rare primary pulmonary disease associated with proliferation of neuroendocrine cells in the lung and multifocal neuroendocrine tumorlets. Although typically a benign condition, DIPNECH causes chronic, progressive cough and dyspnea which can adversely impact quality of life. There have been no treatments described in the literature.

Methods: Three patients with DIPNECH associated with chronic cough have been treated at the Moffitt Cancer Center with octreotide LAR. Their charts were reviewed to assess symptomatic response to treatment.

Results: The three patients were women, ages 55-60, who presented with a biopsy-proven diagnosis of DIPNECH. They complained of similar symptoms including chronic cough with associated mild phlegm production and dyspnea with exertion. They had received previous treatment with steroids and/or inhalers; however their symptoms did not improve. The patients were started on treatment with octreotide LAR 20-30mg every 4 weeks. Since initiation of therapy, all three women have noticed a subjective partial improvement in their cough and dyspnea. The longest period of response that we have documented is 18 months. Also of note, one of the patients stopped octreotide due to loss of insurance coverage, and her cough increased while off the medication.

Conclusions: Octreotide LAR treatment may improve chronic respiratory symptoms associated with DIPNECH. Prospective trials focusing on symptom-control are warranted.

¹Moffitt Cancer Center, Tampa FL