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Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH) and the Role of Somatostatin Analogs

Aman Chauhan, MD[1]; Robert A. Ramirez, DO[2]

[1]PGY IV Resident Internal Medicine and pediatrics; LSUHSC New Orleans
[2]Department of Hematology and Oncology, LSUHSC New Orleans

Background: DIPNECH is a rare pre-neoplastic condition that often presents with a variety of non-specific pulmonary symptoms and sometimes seen in conjunction with pulmonary carcinoid tumors. There is no published data on use of somatostatin analogs in patients with DIPNECH. We review the long term outcomes of somatostatin analog therapy with regard to symptom control in patients with DIPNECH.

Methods: Retrospective study out of our registry of over 2000 neuroendocrine tumors identifying 184 pulmonary neuroendocrine tumors. Out of this there were 5 histopathologically confirmed cases of DIPNECH. Appropriate institutional review board permission was taken for this analysis.

Results: All 5 patients were females, with a mean age at diagnosis of 65.5 years. Median follow up period was 9 years. Cough was the presenting complaints in all 5 patients described as mostly dry, except for one patient who had productive early morning cough. Other symptoms seen in one patient included wheezing, flushing and fluctuating blood pressure. No one reported weight loss, hemoptysis and shortness of breath. One of our patients had a benign thyroid nodule and two patients had previous history of breast cancer. All five of our patients were histopathologically diagnosed with an open lung biopsy. 4 out of 5 patients were started on somatostatin analog. All four patients reported significant improvement in cough. One patient reported mild abdominal discomfort and diarrhea as side effects but remained on treatment. Chromogranin A levels were followed and declined in all but one patient following initiation of somatostatin analog.

Conclusions: From our single institution review of pulmonary neuroendocrine tumors we found only 5 cases of DIPNECH, which reaffirms the rare nature of this pathology. It primarily affects females over 60 years with dry cough as the most common presenting symptom. Most of our patients responded to treatment with a somatostatin analog and had significant improvement in their presenting symptoms. Trending chromogranin A levels seemed to correlate with clinical improvement while the patients were on a somatostatin analog. Somatostatin analogs were well tolerated. Further research is needed in this rare condition, however, a trial of somatostatin analogs should be considered in the treatment of patients with DIPNECH with responders being treated long term.