Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH): A Case Presentation and Overview of Documented Cases (Poster).

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Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH): A Case Presentation and Overview of Documented Cases

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Introduction

- Pulmonary neuroendocrine cells (PNECs) span the entire respiratory tract, synthesizing amines, peptides, and cytokines (serotonin, gastrin, chromogranin A, and bombesin). They are known to undergo reactive hyperplasia in response to chronic injury.
- The World Health Organization (WHO) recognized diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) as a primary, pre-invasive form of PNEC proliferation with a spectrum ranging from tumorlet to carcinoid tumor formation within the terminal and respiratory bronchioles (1999).
- DIPNECH can lead to varying degrees of obstructive ventilatory defects secondary to fibrosis and proliferation of PNECs beyond the basement membrane.
- High resolution CT (HRCT) screening has led to the increasing recognition and understanding of the disease, accounting for 130 cases described in current literature.

Case

A 70-year-old Caucasian female life-long non-smoker with a chronic cough of 25 year duration had been unsuccessfully treated for presumed allergic rhinitis, postnasal drip, and cough variant asthma. Bronchoscopy was unrevealing while a HRCT scan of the chest discovered two pulmonary nodules with moderate air trapping on expiratory imaging (Figure 1).

The patient underwent wedge resection with pathology revealing carcinoids with tumorlets and PNEC hyperplasia (Figure 2). These tumors were hormonally inactive and a full workup disclosed no metastases. Her current treatment consists of cough suppression with an opiate.

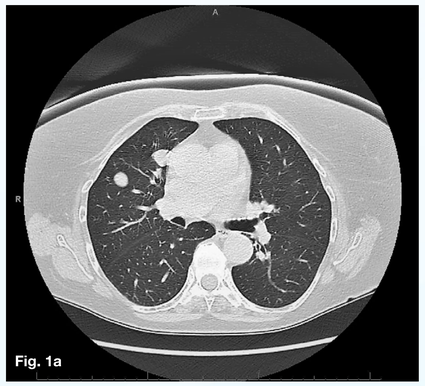


Figure 1a: HRCT - Right upper lobe pulmonary nodules.

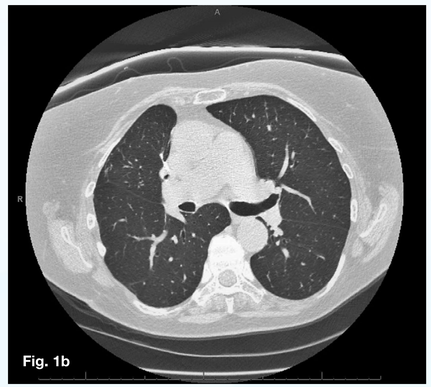
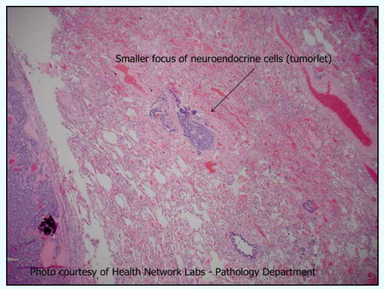
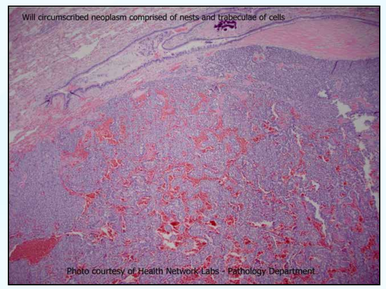
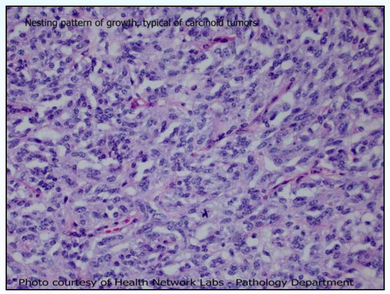


Figure 1b: HRCT – Expiratory image demonstrating air trapping (Post right upper wedge resection).







Discussion

Review of the literature revealed that our patient presented with typical features of DIPNECH (Table 1). The majority of patients are symptomatic and are misdiagnosed for years.

- Transbronchial biopsy and lavage have shown a low yield while PFT testing reveals an obstructive pattern in most symptomatic patients.
- HRCT scans should be obtained with the addition of expiratory imaging to aid in recognition of the mosaic pattern of air trapping suggestive of constrictive obliterative bronchiolitis.
- The current gold standard of diagnosis remains a surgical lung biopsy with histopathologic confirmation.
- Typical disease course is indolent and non-progressive with a small percentage of cases experiencing clinical deterioration. The majority of patients have a favorable long term survival with observation alone.
- While there is no known definitive treatment for DIPNECH, limited data reports improvement with systemic/inhaled steroids. Somatostatin analogs such as Octreotide have inconsistently shown stabilization of disease progression.
- The use of cytotoxic agents is not recommended and may be detrimental. Elevated serum biomarkers and ipsilateral lymph nodal metastasis have been reported but did not predict a poorer outcome.

Table 1. Features of DIPNECH (N=109)

Sex	Female predominance of 92%
Age	34 - 78 years (mean 57; median 62)
Ethnicity	Caucasian predominance suggested
Tobacco History	33% active/former smokers
Symptomatic	55% - 89%
Symptom Type	Cough (71%), dyspnea (63%), wheezing (25%)
Duration	6 months to > 2 decades
PFT	Obstructive (63-86%), restrictive (13%), mixed (17%), nurmal (17%)
HRCT Findings	96% air trapping, 62% one or more pulmonary nodules, 29% ground glass, 21% bronchiectasis
Biopsy Results	88% tumorlets, 40% carcinoid tumors, 44% constrictive bronchiolitis
Prognosis (2-5yr)	41-45% stable course, 26-32% decline on PFT, 35% improved with treatment (inhaled steroids 67%, oral steroids 46%, octreotide 17%)