Life Threatening Exacerbation of Tracheobronchopathia Osteochondroplastica in a 25 Year Old Female

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Introduction Tracheobronchopathia Osteochondroplastica is a rare idiopathic disorder of the anterior and lateral trachea. It is characterized by the calcification and ossification of the luminal cartilage of the trachea and bronchi with formation of submucosal osseous nodules leading to narrowing of the airway. Case Presentation: A 25 year old African-American female with past medical history of vitiligo is hospitalized after six weeks of cough and dyspnea. The cough had progressed from dry to producing a white sputum along with intermittent wheezing and throat discomfort. Previously, several courses of corticosteroids were prescribed with minimal improvement. Social history revealed frequent marijuana use. Her family history was significant for sarcoidosis and asthma. Presenting vitals were within normal limits. Physical exam revealed stridor with course bronchial breath sounds. CT chest revealed a small nodular opacity in the lumen of the trachea and mild diffuse thickening of the trachea and mainstem bronchi with subtle mucosal nodularity. Pulmonary function testing revealed possible small airway disease. During the hospital course, she was treated with antibiotics and inhaled corticosteroids with minimal improvement. Extensive infectious disease investigation revealed only rhinovirus infection. On hospital day 6, oxygen desaturation began. High flow oxygen was started, but was ineffective. Heliox therapy and dexamethasone were then initiated. Direct laryngoscopy revealed multiple subglottic nodules. Bronchoscopy revealed smooth, pink nodules and subglottic narrowing of the airway that spared the posterior tracheal wall (image 1). Tracheal nodule biopsy revealed respiratory epithelium with squamous cell metaplasia. Therefore, a clinical diagnosis of Tracheobronchopathia Osteochondroplastica was made. Antibiotics were discontinued. Dexamethasone was augmented with dapsone and NSAIDs. The patient made a full recovery. Discussion: Tracheobronchopathia Osteochondroplastica typically presents with cough, hemoptysis, and dyspnea but is often asymptomatic until the patient develops a respiratory insult. The disease usually develops slowly and rarely progresses to life threatening airway obstruction. Typically, the condition presents in the 5th or 6th decade of life. The diagnosis is usually determined by bronchoscopy. Although the cause of the ailment is not understood, one study found high number inflammatory cells on histopathology. Both the acuity of the patient’s exacerbation and her youth make her an especially rare case. The patient’s history of autoimmune disease, response to anti-inflammatory drugs, and history of smoking marijuana supports the theory that the disorder is linked with chronic inflammation. Further investigation into whether autoimmune diseases are linked to Tracheobronchopathia Osteochondroplastica is warranted.