Atypical Pulmonary Carcinoid Tumor Encapsulated in a Staphylococcus aureus Infection

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Abstract

To the editor: We present a 13-year-old boy with mild persistent asthma and obstructive sleep apnea who initially presented with fever, dyspnea, cough, night sweats and myalgia for 4 days. He reported having an intermittent "hacking cough" for years. Physical examination was significant for diffuse crackles in the right lung base, decreased breath sounds and mild digital clubbing. Initial chest x-ray (CXR) revealed right middle lobe (RML) and right lower lobe (RLL) opacities with right pleural effusion interpreted and treated as community acquired pneumonia. He was followed as an outpatient by his pediatrician and referred to our pediatric pulmonology clinic months later for persistent RLL atelectasis and chronic cough. He was well appearing with similar physical examination findings as described earlier. Laboratory tests including sweat chloride, cystic fibrosis genetic panel, immunoglobulin levels, complement, pneumococcal and tetanus titers were normal.

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We present a 13-year-old boy with mild persistent asthma and obstructive sleep apnea who initially presented with fever, dyspnea, cough, night sweats and myalgia for 4 days. He reported having an intermittent "hacking cough" for years. Physical examination was significant for diffuse crackles in the right lung base, decreased breath sounds and mild digital clubbing. Initial chest x-ray (CXR) revealed right middle lobe (RML) and right lower lobe (RLL) opacities with right pleural effusion interpreted and treated as community acquired pneumonia. He was followed as an outpatient by his pediatrician and referred to our pediatric pulmonology clinic months later for persistent RLL atelectasis and chronic cough. He was well appearing with similar physical examination findings as described earlier. Laboratory tests including sweat chloride, cystic fibrosis genetic panel, immunoglobulin levels, complement, pneumococcal and tetanus titers were normal.

Computerized tomography (CT) of the chest revealed a mass in the right mainstem bronchus causing distal obstruction of the RML and RLL (Figure 1). Flexible bronchoscopy revealed a round, tan-colored, cystic-appearing mass completely obstructing the bronchus intermedius (Figure 2). The mass was attached to the bronchial mucosa and was not resectable. Bronchoalveolar lavage and biopsy via rigid bronchoscope was done and grew methicillin-susceptible *Staphylococcus aureus*(MSSA) without evidence of tumor or other pathology on these initial specimens (Figure 3A). The patient was treated with a course of amoxicillin-clavulanate followed by clindamycin for a total of 6 weeks. A repeat combination flexible and rigid bronchoscopy was performed and repeat biopsy revealed an atypical pulmonary carcinoid tumor (Figure 3B).

Following the diagnosis, he underwent open thoracotomy with complete resection of the RML and RLL. There was local metastasis to two ipsilateral peribronchiolar lymph nodes which were also resected. Pathology revealed a 4.0 x 1.8cm mass and was strongly chromogranin, synaptophysin, CD56 and AE1/AE3 positive. Ki67 was 34%. There was necrosis and 2 mitoses per $2mm^2$. He denied systemic carcinoid symptoms such

as flushing, edema, diarrhea and palpitations. A 24-hour urine collection for 5-hydroxyindoleacetic acid (5-HIAA) and whole-body positron emission tomography (PET) scan were negative.

Neuroendocrine tumors (NET) are rare primary neoplasms that affect the lung, small intestine and rectum. Lung NET are slow-growing masses that arise from enterochromaffin cells in the bronchial mucosa with an incidence of 0.2 to 2 cases per 100,000[1]. Atypical carcinoid tumors comprise <1% of primary lung neoplasms. Criteria for diagnosis include presence of carcinoid features, necrosis (typical carcinoid does not) and the presence of 2 mitosis per $2mm^2$ on biopsy[2]. There are limited case reports in the pediatric population of atypical pulmonary carcinoid[3]. Surgical resection remains the only curative option with a 5-year survival rate of 75% and 10-year survival rate of 64% in locally resectable masses in adults[4]with limited data in children. There is no current consensus on first line chemotherapy and monitoring for disease recurrence.

We present the first case of a pulmonary atypical carcinoid tumor initially presenting as a MSSA infection in a pediatric patient. The initial presentation as an MSSA superinfection encapsulating the tumor delayed the eventual diagnosis, presenting as a diagnostic and prognostic challenge. Given that metastatic disease in carcinoid tumors carry a much poorer prognosis, it is of paramount importance that the underlying malignancy be discovered in a timely manner. There have been case reports describing fungal superinfection with Aspergillus masquerading as pulmonary carcinoid tumors,[5] but there are no described cases of bacterial superinfection. When faced with a bronchial mass that presents as an infection, it is important to treat the presenting infection and repeat the bronchoscopy and biopsy to discern whether there is an underlying condition such as malignancy.

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FIGURES

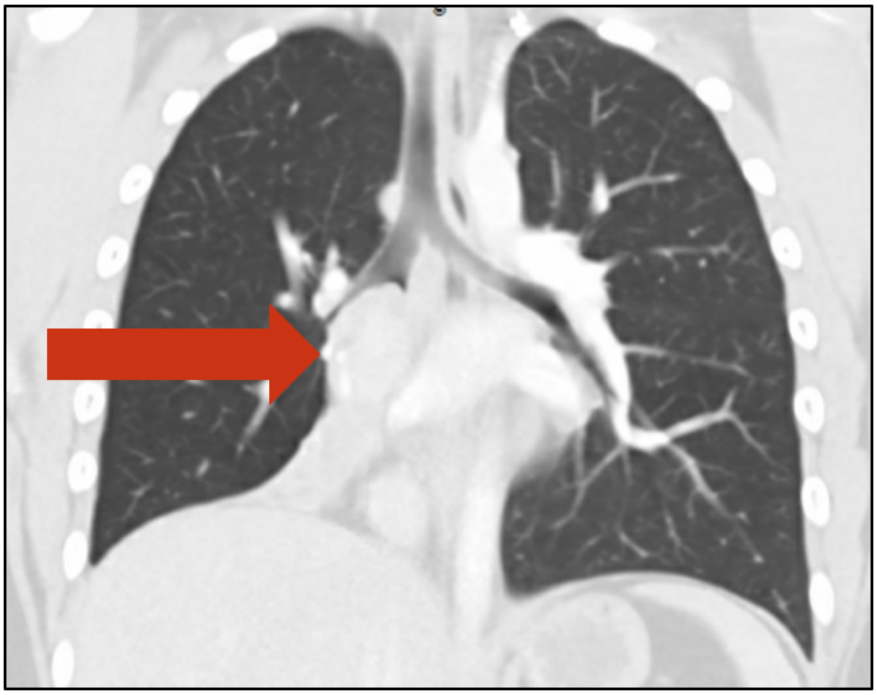

Figure 1. Chest computerized tomography of the mass (arrow) causing airway obstruction with collapse of the right middle and lower lobes.

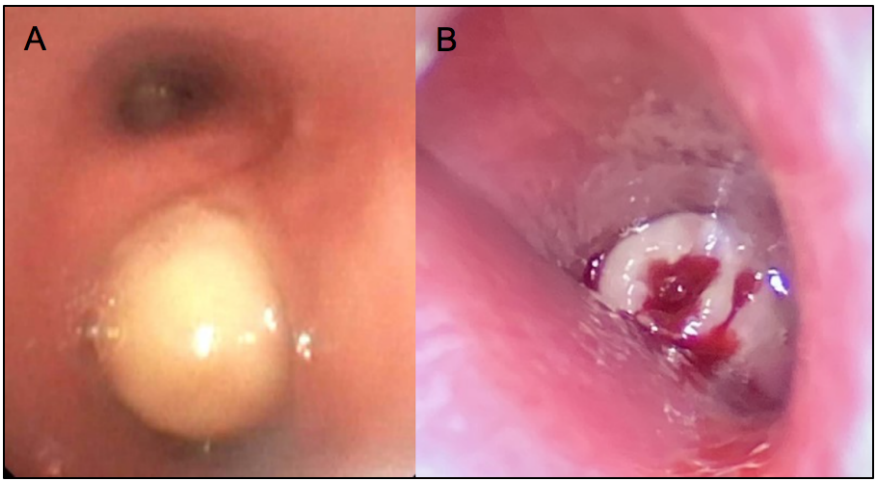

Figure 2. A. Endobronchial mass found in the bronchus intermedius during flexible bronchoscopy. **B.** Biopsy was obtained during subsequent rigid bronchoscopy. The mass was attached to the bronchial wall and was encapsulated in caseous material.

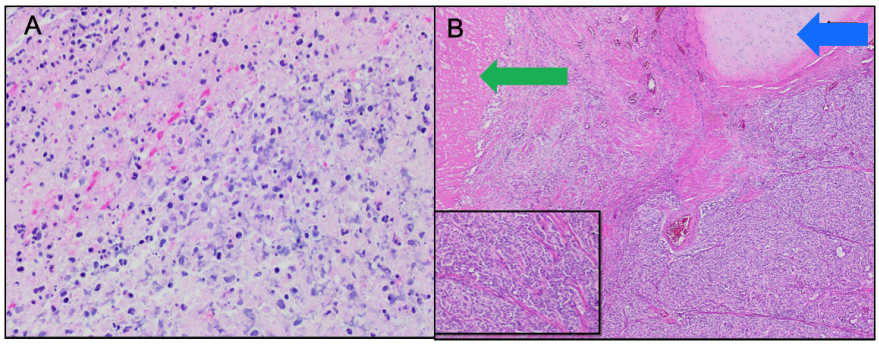

Figure 3. A. Necroinflammatory debris, consistent with *Staphylococcus aureus* bronchitis (H&E, 200X). Gram stain (not pictured) showed gram positive cocci in clusters. **B.**Atypical carcinoid tumor (lower right) adjacent to bronchial cartilage (blue arrow) with focal necrosis (green arrow), (H&E, 40X). Insert (lower left) shows carcinoid tumor characterized by nests and trabeculae of uniform cells with stippled chromatin (H&E, 100X).