

Primary mucoepidermoid carcinoma at the carina of trachea presenting with wheezing in an asthmatic child mimicking an attack of asthma

A case report

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Abstract

Background: Asthma is a very common disease, but primary tracheal tumors are extremely rare in children. Wheezing is not pathognomonic, but is the typical presentation of asthma and could also be found in patients with tracheal tumors.

Clinical Findings: This report describes a 12-year-old boy with a previous history of frequent asthma attacks and experienced responses to antiasthma treatment. He was admitted to the hospital due to persistent wheezing and progressive dyspnea. Hyperinflation in the bilateral lungs was detected on chest x-ray, but without other significant findings. Chest computed tomography revealed a mass at the carina. Tracheal mucoepidermoid carcinoma was diagnosed by histopathological assessment.

Conclusion: Despite the rarity of tracheal tumors, chest computed tomography scans should be performed in the first place for children presenting persistent wheezing and having poor response to antiasthma treatment to rule out the other alternative diagnosis. Coexistence of other diseases such as tracheal tumor in asthmatic patients should be considered.

Abbreviations: CT = computed tomography, GINA = global Initiative for *Asthma*, MEC = mucoepidermoid carcinoma.

Keywords: asthma, carina of trachea, children, mucoepidermoid carcinoma, wheezing

1. Introduction

Asthma is one of the most common diseases affecting children.^[1] Wheezing, usually resulting from airway obstruction, is the typical clinical presentation of asthma but not a pathognomonic feature. Primary tracheal tumors are extremely rare in children.^[2] The initial clinical presentations of tracheal tumor are mostly

nonspecific respiratory symptoms, such as cough, dyspnea, hemoptysis, and wheezing that may mimic asthma. Because of tumors that originate in the trachea are rare in children, many patients were initially misdiagnosed with asthma. Mucoepidermoid carcinoma (MEC) is also rare tumor that arises from the serous and mucous cells of salivary glands and the respiratory systems. Less than 10 children with tracheal MEC have been reported in literature.^[3–8] This article describes a 12-year-old boy with wheezing and a history of asthma who was finally diagnosed with MEC at the carina of trachea after an initial misdiagnosis of asthma. To our knowledge, this is the first case of tracheal tumor with coexisting asthma.

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2. Consent

Institutional Review Board approval is not required for case report. The patient's father signed the necessary of documents to consent to the use of patient's data for teaching and publication.

3. Case report

A 12-year-old boy originally showed symptoms of wheezing and dyspnea when he was 5 years old. The previous chest x-ray revealed hyperinflation in both lungs. An elevated percentage of eosinophils (>15%) was noted in the peripheral blood. The total serum IgE level was higher than 2400IU/mL, and allergen test revealed hypersensitive to dust mites. After the diagnosis of asthma was made, anti-asthma treatment using bronchodilators and steroids led to rapid improvement in symptoms. However, he had many attacks of asthma thereafter, with the severity of moderate persistent categorized according to GINA classification.

Before this report, he was admitted into the hospital because of shortness of breath, dyspnea on exertion, and frequent



Figure 1. Computed tomography of the chest. A hypervascular mass with a diameter of about 26mm was noted at the carina of trachea (arrow).

hemoptysis over a 3-month period. Physical examination demonstrated obvious wheezing breath sound with apparent subcostal and intercostal retractions. Complete tests on blood count and blood biochemistry were performed, and all were within the normal ranges. A chest x-ray showed hyperinflation in both lungs, low-set diaphragm, and vertical heart without other significant findings. Exacerbation of asthma was impressed, and bronchodilators and steroids were administered. However, the symptoms became worse, and the saturation of oxygen decreased soon later.

Computed tomography (CT) of the chest was then performed. A hypervascular mass with a diameter of about 26mm was detected at the carina of trachea (Fig. 1). Calcified mass was well detected using precontract-enhanced CT, and it resulted in severe

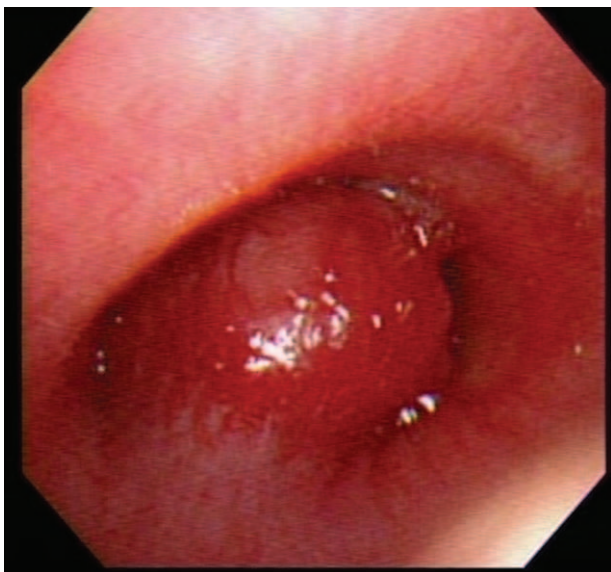


Figure 2. Bronchoscopy demonstrates a smooth, well-circumscribed, and pedunculated mass on the carina with tracheal obstruction.

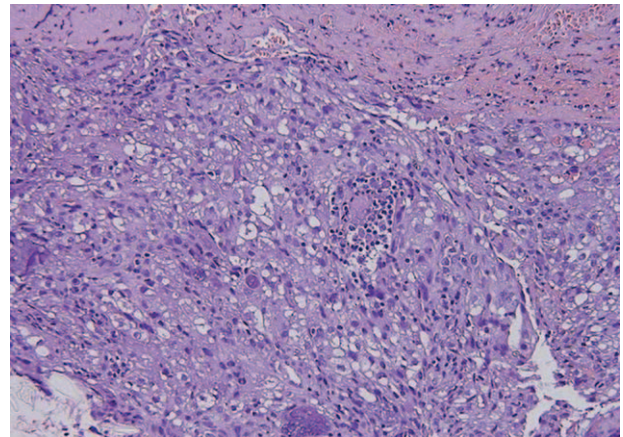


Figure 3. The section of the tracheal tumor reveals polygonal tumor cells with abundant cytoplasm in a nest arrangement. Mucous cells alternating with squamous cells were noted (Hematoxylin and Eosin stain, $\times 400$).

compression of the main airways. Pneumomediastinum and subcutaneous emphysema were also noted. A smooth, well-circumscribed, and pedunculated mass on the carina with tracheal obstruction was found by bronchoscopy (Fig. 2). Endoscopic partial resection of the tumor was performed. Pathologically, polygonal tumor cells with cellular polymorphism and abundant cytoplasm arranged in nests were demonstrated (Fig. 3). Immunohistochemical staining revealed positive for cytokeratin but negative for S-100, P63, smooth muscle actin, and TTF-1. Finally, a diagnosis of MEC was made. Due to the risks of total excision of the tumor located in the carina, his parents refused to receive further surgical intervention after partial removal of tumor by bronchoscopic resection. This patient has been doing well without respiratory distress for one year of follow-up.

4. Discussion

Asthma is one of the most common diseases in the world, and can affect people of all ages.^[1] Asthma attacks occur frequently if no adequate control. Unlike some biomarkers for other diseases,^[9] there is no useful biomarker specific for asthma. While an increase in the percentage of eosinophils, tests for allergic antigens, and an elevated serum level of total IgE are helpful. A diagnosis is usually made mainly based on the patient's history and physical examination findings. Thus a diagnosis of asthma was made according to these typical phenomena found in this patient and he had good responses to antiasthma treatment in each asthma attack except the last one. As asthma is very common, coexistence of other diseases in asthmatic patients should be considered, especially in those who respond poorly to regular therapy of asthma.

Although wheezing is not pathognomonic for asthma, but commonly found in patients with asthma. Luminal narrowing anywhere along the respiratory tract can cause wheezing.^[10] A certain of nonrespiratory conditions such as gastroesophageal reflux or vascular ring may also cause symptoms that mimic asthma.^[10] Besides, coexistence of other diseases with asthma could increase the severity. The present patient had a history of asthma and typical presentations, including wheezing, dyspnea, and evidence of hyperinflation on the chest x-ray. But in this episode, he responded poorly to the anti-asthma medication that

has been effective after the attacks of asthma in the past years. Therefore, it should be considered that other diseases might be linked to this situation. Wheezing in a patient with asthma may be caused by other conditions in case of poor response to antiasthma treatment or patient's history and/or physical examination suggest an alternative diagnosis.

Common causes of tracheal obstruction are congenital anomalies, mass, and infection. Primary tracheal tumors are very rare in children, and even in adults.^[2,11] Unlike in adults, approximately two-thirds of primary tracheal tumors in children are benign.^[2] The presenting symptoms are related to the location of the mass within the trachea, including cough, hoarseness, dyspnea, hemoptysis, wheeze, and stridor. When the tumor is small and located behind the carina, hyperinflation resulting from air trap is usually the only abnormal finding on the chest x-ray. As the present patient a misdiagnosis of asthma, the cancer was delayed in diagnosis. CT scan is the best modality to detect tracheal lesions.^[12] Romão et al^[4] recommend that any school-aged child with new-onset or severe asthma-like obstructive airway symptoms should be worked up early with a chest CT scan. Increased radiation exposure by using multidetector CT with automatic exposure control devices presents a particular concern for pediatric patients. However, strategies such as optimized scanning protocols according to age- or weight-based adjustments for children, decreasing the unnecessary examination and scanning phase, and user education for pediatricians and radiological technologies can decrease the risk from radiation exposure.^[13] Although bronchoscopy is more invasive than CT scan, it is commonly used to obtain tissue for histopathologic diagnosis and is useful to assess the resectability of the tumor.

MECs arise from the serous and mucous glands of salivary glands and the respiratory systems. In the central airways, MECs are more common in the bronchus than in the trachea.^[14-15] To the best of our knowledge, less than 10 children with tracheal MECs have been reported in the literature. In which, most were located about 2 to 4 cm above the carina,^[3-8] and the carina was involved in only two.^[4,8] Complete surgical resection has been recommended for primary malignant tumors of the trachea. However, it is very difficult to reconstruct when the carina was involved. Stem-cell-based tracheal replacement is promising^[16-18] and could be a treatment modality in the future. In the present patient, his parents refused a complete surgical resection after partial resection of the tumor by bronchoscope and he has been doing well after one year of follow-up. Total tumor resection with airway reconstruction may not be needed for MECs located in the carina, but long-term follow up is warranted.

5. Conclusions

Respiratory symptoms of persistent wheezing in children are mostly likely diagnosed as clinical problems of asthma. Although frequent asthma attacks are common, other coexisting diseases in asthmatic patients should be considered in case poor therapeutic response of antiasthma treatment is detected, and a chest CT scan should be performed to assess the possibility of rare tracheal tumor for an asthmatic child.

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