LUPINE PUBLISHERS

ISSN: 2644-1306

Theranostics of Respiratory & Skin Diseases

DOI: 10.32474/TRSD.2019.01.000112

Case Report

Persistent Pulmonary Symptoms in two Adolescents: Could It be a Lung Tumor?

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Received: May 27, 2019 **Published:** June 14, 2019

Abstract

We report on two adolescents with primary endobronchial tumors. They both presented with subacute onset of respiratory symptoms, poor response to conventional medical therapy and pulmonary function test findings suggestive of intrathoracic obstruction. Due to prompt bronchoscopic evaluation, timely resection of the tumors was performed.

Keywords: Atypical carcinoid tumor; Primary endobronchial tumor; Pulmonary neuroendocrine tumor; Intrathoracic obstruction

Introduction

Primary endobronchial tumors in childhood are extremely rare. Because of low clinical suspicion and the variable ways of presentation, they are often misdiagnosed as benign conditions resulting in delayed diagnosis [1]. Herein we report two cases of primary endobronchial tumors in adolescents who presented to the Emergency Department of a tertiary pediatric hospital.

Case 1

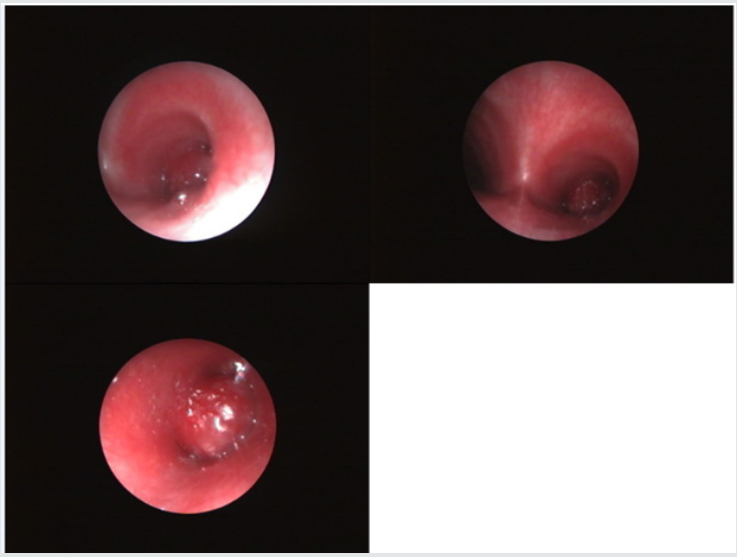

Figure 1: Pictures from bronchoscopic evaluation of case 2. The bronchial tumor occupies completely the right mainstem bronchus.

A 14-year-old female presented with acute onset of cough, fever and chest pain. She had a one-month history of cough treated with broad-spectrum antibiotics and bronchodilators. Several hours prior to admission, she developed fever up to 39.5 oC with chills and chest pain with radiation to her back on deep inspiration. Her past medical and family histories were unremarkable. She was a well-developed adolescent female in no acute distress, 98% oxygen saturation in room air and normal breath sounds on auscultation. Laboratory studies revealed leukocytosis (WBC, 16.6 x 10^3/μl) with a predominance of neutrophils (80%), elevated C-reactive protein (CRP, 89 mg/L) and erythrocyte sedimentation rate (ESR, 75mm). A chest radiograph revealed left upper lobe atelectasis and infiltrate, and a small effusion in the left lung lingula (Figure 1).

A new course of antibiotics and bronchodilators was given with prompt clinical and laboratory response. Blood cultures remained negative. Despite systematic bronchodilation, she developed diffuse wheezing on auscultation. A flow-volume curve obtained at that time revealed a pattern compatible with intrathoracic airway obstruction. Chest computed tomography (CT) confirmed the radiographic findings: atelectasis and infiltrate of the left upper lobe.

Due to the persistence of the airway obstruction, she underwent bronchoscopic evaluation where a lesion obstructed the left upper bronchus completely and the left main stem bronchus by 60%. A radial endobronchial ultrasound demonstrated intraluminar invasion (2cm) resulting in post obstructive atelectasis. The biopsy of the lesion was compatible with an atypical carcinoid tumor of the lung. Our patient underwent left upper lobe lobectomy. More than 20 lymph nodes examined were disease-free. Three years post operatively she remains asymptomatic with negative follow up chest CT and bronchoscopy.

Case 2

A 15-year-old male presented with a two-day history of fever up to 40 oC with chills, mild cough and right-sided chest pain. He had an eight-month history of persistent cough despite the use of inhaled bronchodilators. Two months prior to presentation he was hospitalized with right lower lobe pneumonia. A rightward mediastinal shift found on chest X-ray (CXR) at that time prompted a chest CT scan that revealed situs inversus with dextrocardia and the presence of a mass obstructing the right mainstem bronchus. No further investigation was performed as his parents declined consent. There were no further remarkable findings from his past medical and family history.

Upon admission, he was a well-developed male in no acute distress. Oxygen saturation measurement was 98% in room air. Significant findings on physical examination were reduced air entry in the right lung field and heart sounds best heard on the right side of the chest. Laboratory studies revealed elevated WBC count (15 x 103/μl) with a predominance of neutrophils (84%) and an elevated CRP 178mg/L. A right middle lobe density was found on CXR examination. Blood and sputum cultures were negative.

Despite treatment with antibiotics, steroids, bronchodilation and physiotherapy, he remained symptomatic with decreased breath sounds in the affected side and a flow-volume curve pattern was compatible with intrathoracic airway obstruction. Bronchoscopic evaluation revealed a lesion obstructing completely the right mainstem bronchus. Following cardiothoracic consultation, it was decided to proceed with laser photoresection of the lesion in a specialized medical center in the United Kingdom. Histologic evaluation of the lesion set the diagnosis of typical carcinoid. He underwent repeated bronchoscopies for the removal of the lesion. The patient was placed on close monitoring protocol.

Discussion

Our cases underline the importance of early diagnosis of endobronchial lesions in pediatric patients. Sub-acute onset of respiratory symptoms, poor response to standard medical treatment, along with findings on pulmonary function tests suggestive of intrathoracic obstruction prompted further radiological and endoscopic investigation in only a few days. A small series review from France revealed a mean delay between symptoms appearance, diagnosis and surgical treatment of 5.6 months [2]. Isolated reports however describe pediatric patients who were treated as asthmatics for years (2 -5.5 years) prior to diagnosis [3,4].

Endobronchial lesions should be considered in children with persistent pneumonia or respiratory symptoms despite adequate treatment. Primary bronchial tumors seldom occur in children, and represent a rare cause of pulmonary obstruction. The most frequent histologic subtype is well-differentiated neuroendocrine tumor (NET), or carcinoid [5].

Symptoms of the obstructive nature of the tumor are variable; small pediatric patient series report the following presenting symptoms: recurrent pulmonary infection, pneumonia, cough, haemoptysis, chest pain, weight loss and persistent wheezing [1,2,5]. A significant asymptomatic percentage of adult patients (as high as 25%) has been reported [6,7]. Evidence of atelectasis/collapse on chest X-ray and CT heighten the index of suspicion. Rarely symptoms such as flushing, diarrhea, palpitation, shortness of breath consistent with carcinoid syndrome may occur. Those symptoms are caused by serotonin release from the tumour. In pediatric patients, carcinoid syndrome is extremely rare [8]. Our patients did not exhibit symptoms of carcinoid syndrome.

The lung is the second most common site of occurrence of well-differentiated NETs, after the gastro-entero-pancreatic axis. The 2004 World Health Organization classification recognizes four major histopathologic types of lung NETs: low-grade typical carcinoid, intermediate-grade atypical carcinoid, high-grade small cell and large cell carcinoma. Among pulmonary NETs, typical and

atypical carcinoid tumors of the lung are generally indolent, but capable of very late recurrence or metastasis [9].

The incidence of these lesions and their outcomes are still largely unknown, although an increased incidence of NETs in the last decade has been noticed worldwide. Review of the scant relevant literature suggests a predominance of low-grade typical carcinoids. Patients with atypical disease are three times more likely to succumb from metastasis/locally advanced tumor at presentation or recurrent disease following resection than patients with typical disease [10].

Despite their malignant potential, complete conservative resection prior to tumor spread is the treatment of choice for the management of pediatric bronchial carcinoids. Bronchoscopic removal of the tumor is not considered as first line treatment but it has been reserved for cases in which surgical intervention carries an unexceptably high risk [11,12] and remains an appealing approach that needs further evaluation [13]. Relapses can be successfully surgically treated and can occur even after many years, highlighting the importance of long-term follow-up, probably for life [14].

In accordance with previous reports, our patients belonged to adolescent age-group; therefore they were able to perform the spirometric evaluation, which was characteristic of obstruction [15-17]. The problem is difficult to solve when the patient is younger and suffers recurrent episodes of wheezing and respiratory infections. A high degree of suspicion is necessary in order to decide which patient needs further evaluation.

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DOI: 10.32474/TRSD.2019.01.000112