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A rare primary lung tumor in children: Carcinoid tumor

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The authors stated that the written consent was obtained from the parents of the patient presented with images in the study.

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Abstract

Bronchial carcinoids are rare childhood malignancies, comprising only a small proportion of pediatric lung tumors. This report details the case of a 9-year-old girl with persistent pneumonia that lasted one month and remained unresponsive to antibiotic therapy. Radiological imaging revealed atelectasis of the superior lobe of the left lung, while computed tomography showed an endobronchial lesion in the left main bronchus. An endobronchial biopsy confirmed the diagnosis of a typical bronchial carcinoid tumor. The tumor was completely resected using bronchoscopy, and cryotherapy was used to prevent the tumor's recurrence. No evidence of tumor recurrence was observed in subsequent follow-up bronchoscopy. This case underscores the importance of considering bronchial carcinoid tumors in the differential diagnosis of pediatric patients with recurrent pneumonia or wheezing that is resistant to standard treatments. Through early identification and minimally invasive management, such as bronchoscopy and cryotherapy, it is possible to achieve successful outcomes and avoid more extensive surgery. Our findings add to the scarce literature on pediatric bronchial carcinoid tumors and highlight the instrumental role of prompt and accurate diagnosis in improving prognosis.

Keywords: bronchial carcinoid, childhood, bronchoscopy, biopsy, cryotherapy

Introduction

Bronchial carcinoids account for fewer than 0.1% of pediatric malignancies, rendering them extraordinarily rare. Despite representing 42-63% of primary lung malignancies in children, their prevalence significantly declines to 1-2% among adults [1].

Bronchial carcinoids are divided into two subgroups according to their likelihood of malignancy. Typical carcinoids, which are more commonly seen within the pediatric population, tend to have lower malignant potential [2].

In children, bronchial carcinoids are often located endobronchially, leading to airway obstruction, which can mimic benign respiratory conditions. This overlap in clinical presentation often results in misdiagnosis and a delay in achieving an accurate diagnosis and initiating proper treatment. Differential diagnoses to consider include asthma, recurrent pneumonia, bronchial adenoma, foreign body aspiration, and congenital airway malformations such as bronchogenic cysts. Due to the nonspecific presentation of symptoms like wheezing, cough, or recurrent infections, these tumors frequently are misdiagnosed, which leads to delays in timely and appropriate treatment. Diagnostic evaluation, including imaging and histopathological examination, is essential to differentiate bronchial carcinoids from these more prevalent respiratory disorders [3].

In our case presentation, we aimed to discuss a typical bronchial carcinoid tumor case that presented as pneumonia unresponsive to treatment, guided by current literature.

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Case presentation

A 9-year-old girl was admitted with a 1-month history of pneumonia that was unresponsive to treatment and progressive dyspnea. She had been intermittently using inhaled steroids for dyspnea throughout the past year. Her vital signs, as well as her growth and development, showed no abnormalities. She had reduced breath sounds in her left lung compared to the right, and no other abnormalities were observed during the systemic examination. The complete blood count and acute phase reactants, along with other laboratory analyses, were all normal.

The chest X-ray revealed reduced aeration, especially in the superior lobe of the left lung (Figure 1). The contrast-enhanced thoracic CT showed a soft tissue lesion occupying the lumen of the left main bronchus approximately 2 cm distal to the carina (Figure 2). We also observed a complete collapse of the left superior lobe with compensatory expansion of the left inferior lobe. Flexible bronchoscopy revealed a soft tissue mass with mild vascularity on the mucosal surface, causing obstruction approximately 1-2 cm distal to the left main bronchus bifurcation (Figure 3). A punch biopsy was performed on the mass. The histopathological examination revealed a tumorous formation featuring small round nuclei, scant cytoplasm, and a solid-trabecular pattern (Figure 4). This formation tested positive for CD56 (Figure 5) and diffusely for synaptophysin (Figure 6). It had a Ki67 proliferation index of 5% (Figure 7), consistent with a typical carcinoid tumor diagnosis. The Ga-68 DOTA-TATE positron emission tomography-computed tomography (PET/CT) scan found no meta-stability irregularities. The mass was completely excised by bronchoscopy using electrocautery snare and cryoprobe. The patient's symptoms fully resolved after the procedure, and both follow-up bronchoscopies performed in the 2nd and 4th postoperative months showed normal findings with no traces of the lesion. During the 6-month and 1-year post-treatment assessments, the patient was asymptomatic and both the physical examination and imaging results were unremarkable. All of the patient's initial symptoms had subsided and not returned.

Figure 1: X-ray: Decreased aeration particularly in the upper zones of the left lung

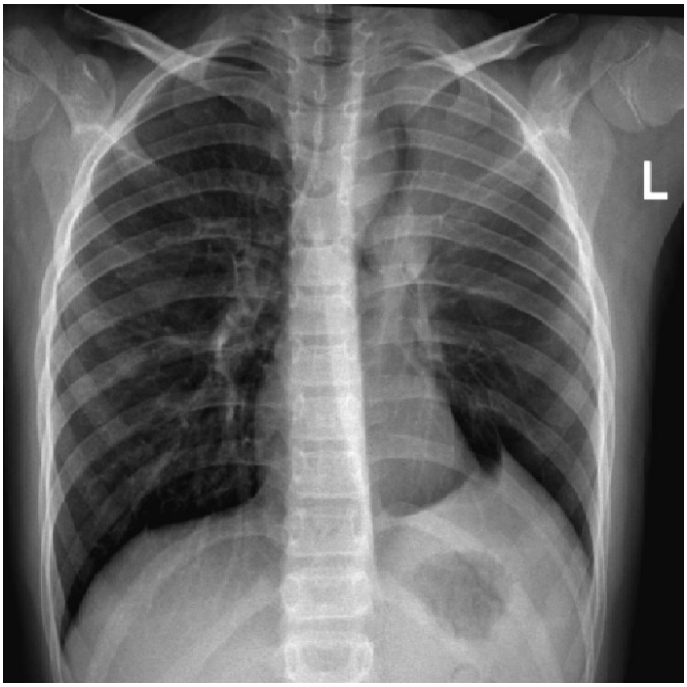


Figure 2: CT: Soft tissue density filling the lumen in the left main bronchus approximately 2 cm distal to the carina (Thick black arrow)

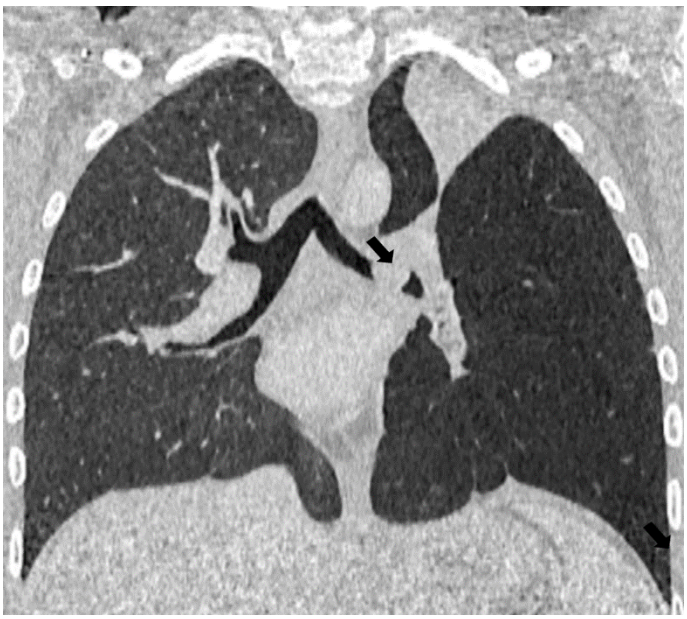


Figure 3: Soft tissue mass with mild vascularity on the mucosal surface, obstructing the lumen

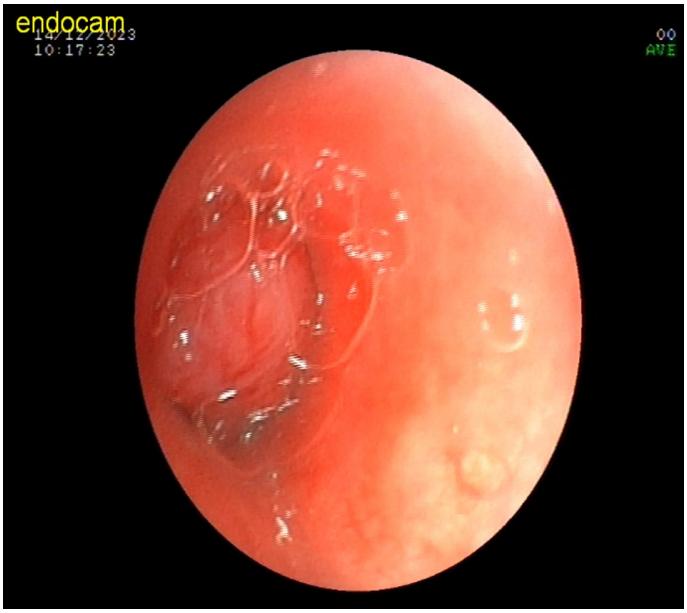


Figure 4: Tumoral formation with small round nuclei, scant cytoplasm, forming a solid-trabecular pattern (Hematoxylin&eosin x40)

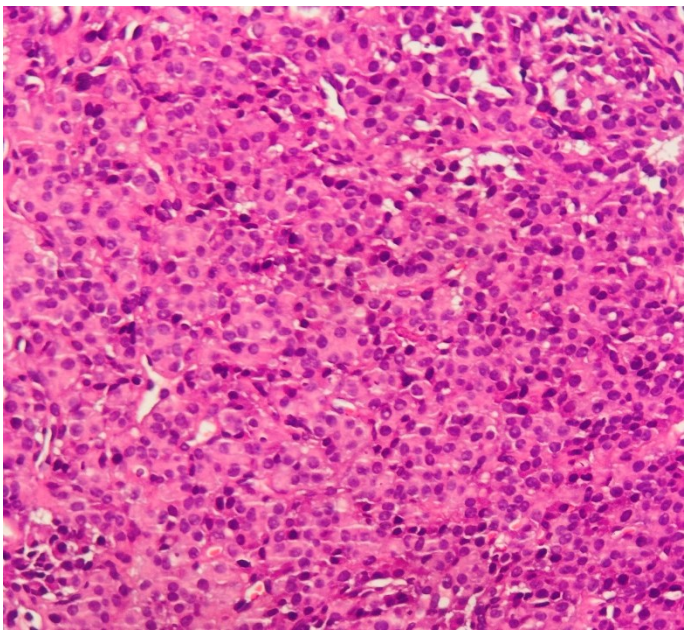


Figure 5: Synaptophysin is diffusely positive in tumor cells (Immunohistochemical analysis X20)

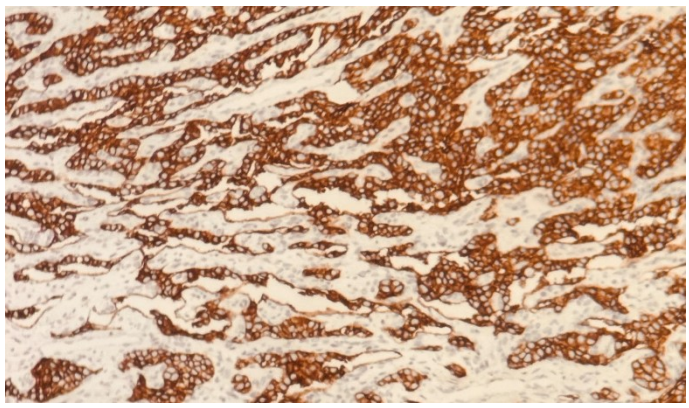


Figure 6: CD56 is diffusely positive in tumor cells (Immunohistochemical analysis X20)

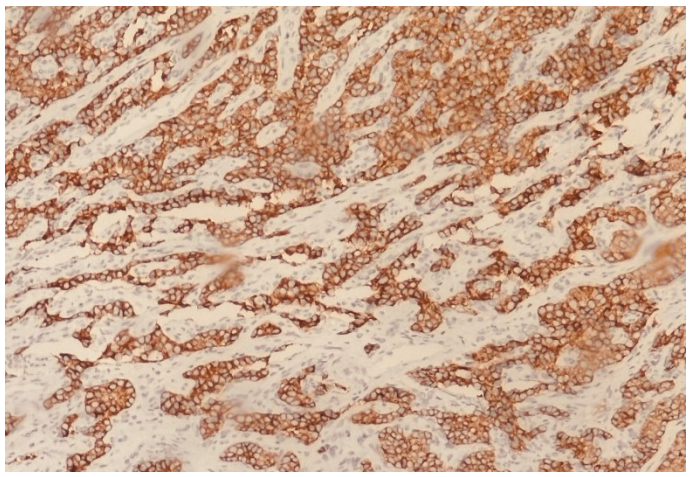
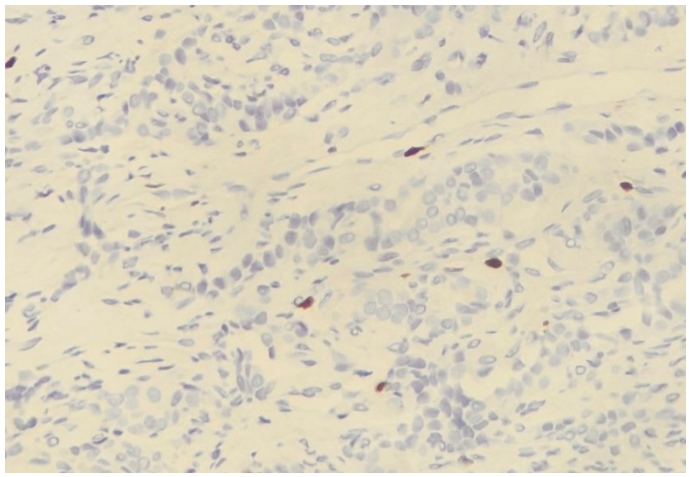


Figure 7: Ki-67 proliferation index is 5% (Immunohistochemical analysis X40)



Discussion

Bronchial carcinoid tumors are well-differentiated malignant neoplasms that originate from Kulchitzky cells, located in the basal layer of the bronchial tissue. These tumors represent 70-80% of all primary malignant lung tumors in children. Typically, carcinoid tumors are centrally located, while atypical carcinoids are more often found peripherally. Both subtypes have the potential to spread to regional lymph nodes, liver, bone, and brain [3]. There are limited studies on childhood bronchial carcinoids, although surgical resection has reported favorable outcomes, particularly for typical carcinoids. Atypical carcinoids, however, exhibit aggressive behavior in adults, but their incidence and prognosis in children remain poorly understood [4].

A review of published pediatric cases indicates that surgery is predominantly the primary treatment modality, leading to favorable survival outcomes in many reports. For instance, Rizzardi et al. [5] detailed the experiences of 15 pediatric patients; only one did not survive due to metastasis. Similarly, Geramizadeh et al. [6] documented positive outcomes in the majority of their cases, excluding two patients who succumbed to metastases. Despite these mortalities, the overall prognosis for bronchial carcinoids in pediatric patients remains generally encouraging, as Rojas and colleagues [7] noted a 95% overall survival rate in a sample of 133 individuals. The collected data underlines the critical role of early detection and intervention, an approach employed in our case management.

Carcinoid tumors typically manifest as polypoid masses within the bronchial tree, involving intraluminal, mural, and extrabronchial extensions. These can result in complete bronchial obstruction, which presents with respiratory symptoms such as shortness of breath and sibilant breath sounds. More severe repercussions, like lung collapse and obstructive pneumonia, may also occur. Symptoms like cough, dyspnea, and stridor, which are common in respiratory infections and pneumonia, can result in delayed diagnosis and treatment. Endobronchial growth often mimics bronchial obstruction and is frequently initially misdiagnosed as asthma [1]. In our case, the patient had been experiencing intermittent dyspnea over the past year, intermittent use of inhaled corticosteroids, and had undergone two recent treatments for pneumonia within the last month.

Radiological imaging plays a pivotal role in the diagnostic process, and atelectasis is the most commonly observed finding on chest X-rays. However, around 10% of cases might present with a normal chest X-ray. CT is crucial for detecting endobronchial lesions and lymphadenopathy; nonetheless, bronchoscopy remains the gold standard procedure for diagnosis, relieving obstructive symptoms, and attaining histopathological confirmation through a biopsy, as exemplified in our case [8].

In cases of suspected bronchial carcinoid, additional imaging with Ga-DOTA-TATE-PET should be undertaken to evaluate lymph node involvement and/or secondary tumors in distant sites [1]. For non-metastatic tumors, complete surgical removal is the preferred treatment, with lymph node excision recommended for atypical carcinoids due to their increased susceptibility to malignant transformation [2].

The aim of surgical treatment is the complete excision of the lesion while preserving as much of the parenchyma as possible. The anticipated outcome is generally favorable, with an 82% 10-year survival rate [3]. This aligns with the larger body of literature on pediatric bronchial carcinoid tumors, which indicates overwhelmingly positive outcomes following surgery, as reported in numerous studies. For instance, Hancock et al. [9] detailed three pediatric cases, all of which survived post-surgery. Gaissert et al. [10], and Al-Qahtani et al. [11] reported similar recovery rates, noting 100% survival in their patient populations.

Cryotherapy is often used as a safe and effective supplementary therapeutic approach for localized endoluminal typical carcinoid tumors. The high sensitivity of carcinoid tumors to freezing and the preservation of tracheobronchial cartilage tissue provides advantages over other adjunct therapies, minimizing local recurrences and long-term complications such as bronchial stenosis, which can occur with alternative adjunct therapies. Evidence of successful results has been shown with adjunct cryotherapy following bronchoscopic resection in adult

studies by Bertoletti et al. [12]. In our case study, adjunct cryotherapy was administered following bronchoscopic resection, and no recurrence was identified in follow-up bronchoscopies.

Conclusion

Although rare in childhood, bronchial carcinoids should still be considered during the diagnostic evaluation of children presenting with persistent pneumonia episodes or wheezing. Early diagnosis and management are crucial. Imaging and bronchoscopy play key roles in both the diagnostic and therapeutic approaches.

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