

Atelectasis in pediatrics: a case of carcinoid tumor

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Abstract

Carcinoid pulmonary tumors occur in the fourth to sixth decades of life. Usually, typical carcinoid arise a decade earlier when compared to atypical carcinoid (45 years and 55 years, respectively). Typical carcinoid tumors are the most common primary lung neoplasm in children and late adolescents, but there are less than 40 cases described in the literature. The clinical presentation is nonspecific and usually the symptoms are due to bronchial obstruction, sometimes with recurrent pneumonia. Its rarity may delay diagnosis but in most cases a favorable course after treatment is observed. The authors describe the case of a 13-year-old girl diagnosed with a carcinoid tumor located on the intermediate bronchus. The treatment approach included endoscopic laser resection, for obstruction resolution, followed by a right inferior bilobectomy with mediastinal lymph node dissection as definitive treatment. Histopathology confirmed a typical carcinoid tumor with mediastinal ipsilateral lymph node involvement.

Case Report

A 13-year-old girl was admitted to the emergency department complaining of pleuritic chest pain in the right hemithorax associated with dry cough. Symptoms had started 3 days ago during basketball practice. No other symptoms or traumatic events were reported. She was previously healthy and had no relevant medical record. At admission the patient was feverish (38.4°C), eupneic, with a blood pressure of 122/63 mmHg, heart rate of 105 beats/minute, respiratory rate of 18 cycles/minute and peripheral oxygen saturation of 97% on

room air. Thorax examination revealed decreased transmission of vocal vibrations and dullness on percussion associated with diminished vesicular murmur at the lower third of the right hemithorax. Blood tests showed leukocytosis ($20.3 \times 10^9/L$) with neutrophilia (79%) and increased CRP (254 mg/dL). Chest x-ray revealed a right paracardiac triangular opacity that allowed the cardiac silhouette's visualization accompanied by a ipsilateral diaphragmatic rise. These findings were compatible with right lower lobe atelectasis. In this context and for better understanding of the underlying condition, a CT scan was performed. The CT scan confirmed the right lower lobe atelectasis due to a homogeneous endobronchial mass, located in the intermediate bronchus, causing partial obstruction and also mediastinal lymphadenopathy at stations 4R and 7 (Figure 1). Diagnosis of an obstructive pneumonia was admitted and she was admitted to the Pediatrics Department for etiological study and treatment. She was put on antibiotics and had clinical and analytical improvement without radiological resolution. A flexible bronchoscopy demonstrated total obstruction of the intermediate bronchus by a rounded and vascularized lesion with a smooth surface (Figure 2A,B). For diagnostic and therapeutic purposes, the patient underwent a rigid bronchoscopy with NdYAG laser resection of the endobronchial lesion (Figure 2C). Pathological examination revealed a typical carcinoid tumor (positive staining for CK 8/18, CD56, Chromogranin A and synaptophysin without necrosis and with <2 mitosis/10 HPF). She was then referred to our institution for staging and treatment. The patient performed a ^{68}Ga -DOTA-NOC PET/CT which revealed abnormal hyper fixation at the intermediate bronchus (SUV: 5.5) and hyper fixation at some mediastinal lymph node stations (10R, 4R and 7). We admitted the diagnosis of a cT2N2M0 typical carcinoid tumor. Based on those findings, she underwent a right inferior bilobectomy with mediastinal lymph node dissection. The surgery was complicated by local hemorrhage. In the postoperative period she developed dyspnea and desaturation associated with hemoglobin drop (from 13 g/dL to 6.8 g/dL) and a right pleural effusion. A hemostasis revision was performed in which we found no evidence of an active bleeding focus. Pathological examination of the surgical specimen (Figure 3) confirmed the presence of a typical carcinoid tumor in the bronchial wall with mediastinal lymph node involvement (metastasis in 10R, 4R and 7 - pT2N2Mx).

The case was discussed by the multidisciplinary team of thoracic tumors and a sur-

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veillance approach was decided. After 18 months the patient remains asymptomatic and without any evidence of relapse.

Discussion

Primary lung neoplasms are a rare event at pediatric age. Approximately, 75% are malignant and 80% of these correspond to typical carcinoid tumors.^{1,2} Those are considered a low-grade neoplasm with slow growing. They arise from the neuroendocrine Kulshitzky cells found in the basal layer of bronchial epithelium.³ Based on histopathologic criteria, carcinoid tumors are classified in typical or atypical according to the presence/absence of necrosis and normal/raised mitotic index. A low mitotic index (<2 mitoses/ 2 mm^2) associated with the absence of necrosis is consistent with a typical carcinoid. The ratio between typical carcinoid and atypical carcinoid is about 8-10:1.⁴ Generally, typical carcinoid seem to adopt a more benign course than atypical carcinoid; however both types are capable of metastasizing to regional lymph nodes or distantly to other organs such as the liver, bone or brain.⁵ Usually, in the pediatric population, the median age at diagnosis is 10.5 ± 3 years.⁶ Frequently, endobronchial carcinoid tumors tend to occur at bronchial bifurcation.⁷ In 85% of cases they involve the bronchi (75% at the lobar level and 10% at the main bronchi) and in 15% they arise from the lung periphery. Commonly, the

carcinoid tumor corresponds to an endobronchial polypoid mass with intraluminal, mural and extra bronchial components. Based on that characteristic, complete bronchial obstruction can arise contributing to respiratory symptoms like dyspnea and wheezing or further complications as atelectasis and obstructive pneumonia.⁸ Functional tumors presenting with carcinoid syndrome are rare.⁹ Besides their potential for a metastatic and invasive behavior, distant metastasis or local recurrences are rare events, especially in patients with typical carcinoid.

When performing a chest X-ray, the detection of atelectasis is the most common finding, like in our patient, although it can be normal in 10% of cases. The CT scan has more sensitivity in the study of endobronchial abnormalities, as it allows for the detection of the different mass components (intraluminal, mural and extra bronchial), the presence of adenopathies and contrast enhancement.¹⁰ CT scan has a crucial importance in the endobronchial mass approach, but bronchoscopy remains the gold standard for diagnosis and allows the resolution of obstructive symptoms, as observed in our case. Carcinoid tumors tend to be vascularized masses that have high bleeding potential, raising questions about the risk/benefit of a bronchial biopsy. Despite that, the bronchial biopsy obtained by bronchoscopy gives the highest yield of positive diagnosis.¹¹

Carcinoid tumor, as a neuroendocrine tumor, expresses somatostatin receptors, so it can be targeted and visualized with radio labeled somatostatin analogues. The rationale for the employment of ⁶⁸Ga-DOTA-conjugate peptides for the assessment of somatostatin receptor expressing tumors is based on the high affinity of these compounds for somatostatin receptors. PET/CT offers a higher resolution compared to somatostatin receptor scintigraphy.¹² In our case the ⁶⁸Ga-DOTA -NOC PET/CT confirmed abnormal somatostatin expression at the intermediate bronchus and at stations 10R, 4R and 7.

Surgical removal is the treatment of choice for bronchial carcinoid tumors. The aim is to remove the tumor and to preserve as much lung tissue as possible. The surgical approach is dependent on the size and location.¹³ Sleeve or bronchoplastic resection are preferred to more extensive resection of centrally located carcinoids as a parenchymal-sparing approach. Our patient underwent right inferior bilobectomy with mediastinal lymph node dissection due to lesion's location and to lymph node involvement. Endoscopic Laser resection as the only form of treatment seems appealing

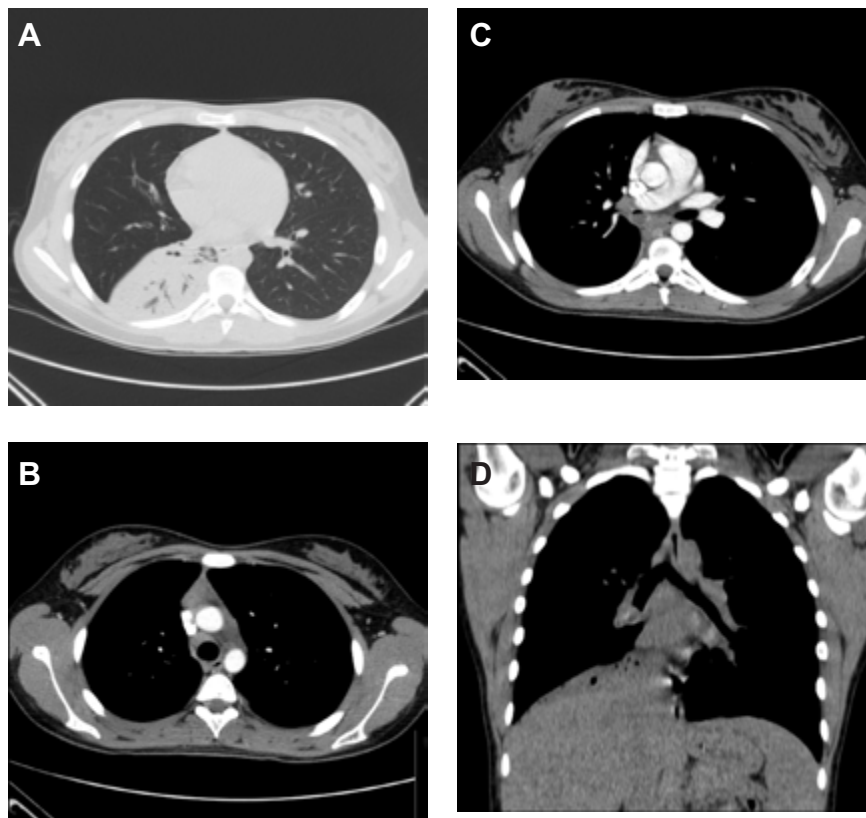


Figure 1. Computed tomography scan at admission: A) axial lung window: right lower lobe atelectasis; B) and C) axial mediastinal window with IV contrast: Enlarged mediastinal lymphadenopathy at 4R,7 and 10R with endobronchial mass causing intermediate bronchus obstruction; D) coronal lung window: right lower lobe atelectasis and intermediate bronchus mass.

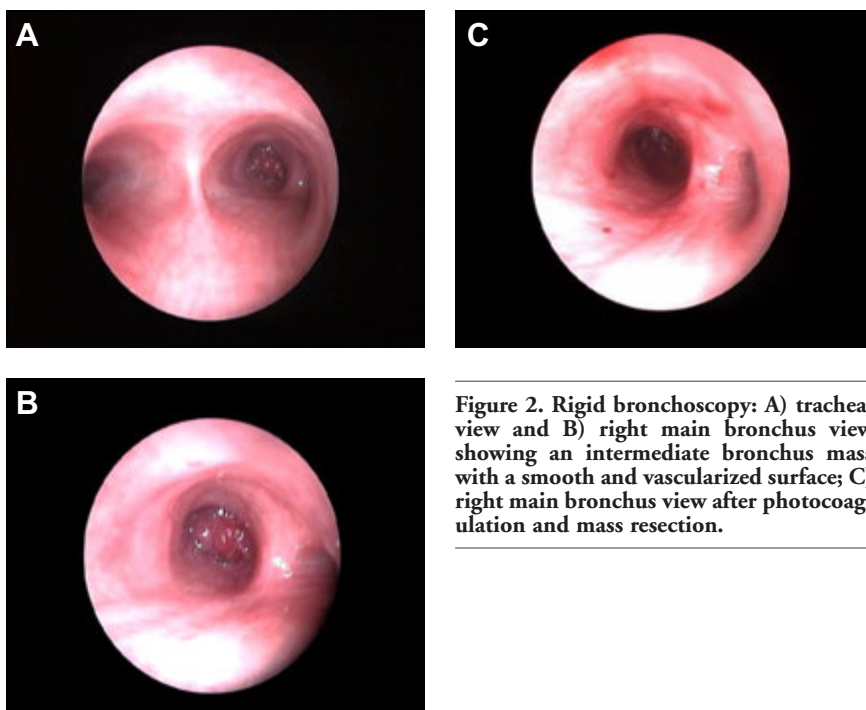


Figure 2. Rigid bronchoscopy: A) tracheal view and B) right main bronchus view showing an intermediate bronchus mass with a smooth and vascularized surface; C) right main bronchus view after photocoagulation and mass resection.

but it's not generally recommended. Although the tumor may be well-visualized endoscopically and a complete luminal resection feasible, intramural and extra-bronchial tumor components results in incomplete resection with residual tumor, as this case illustrated. That can explain the potential for tumor recurrence after endoscopic treatment. Endoluminal bronchoscopic therapy should be reserved for patients who are considered unacceptably high risk for surgery or occasionally as a possible bridge to surgery.¹³ Typical carcinoid has a nodal metastasis rate from 11.5% to 19%.^{14,15}

After treatment the recurrence rate is estimated at 8.2%, with an overall survival of 89% and 82% at 5 years and 10 years, respectively.¹⁶ Granberg *et al.*⁵ showed that some patients with typical bronchial carcinoids die from their disease and that a positive staining for Bcl-2 and p53 correlates with increased risk for distant metastasis as well as decreased survival. The same authors also showed that the expression of CD44 correlates with a decreased risk of metastasis and mortality.⁵

Although our patient's histology is associated with a good prognosis, the presence of mediastinal lymph node involvement may worsen the outcome. The typical carcinoid follow up after surgery consists in conventional imaging at 3 and 6 months and then every 12 months together with chromogranin A measurement for the first 2 years. After the first 2 years, annual chest X-ray and biochemistry profile are recommended as well as a CT-scan every 3 years. Somatostatin receptor scintigraphy should be carried out at 12 months and then only on suspicion of recurrence. Bronchoscopy should be performed if clinical symptoms or radiological findings suggest tumor recurrence and on routine basis every 5-10 years.¹³

Conclusions

Bronchial carcinoid tumor is rare in children and adolescents and the presentation can be non specific leading to a late diagnosis. Bronchoscopy is the diagnostic procedure of choice, having also a role in the treatment of obstructive symptoms. The preferable treatment consists in a surgical approach and the prognosis is good with an

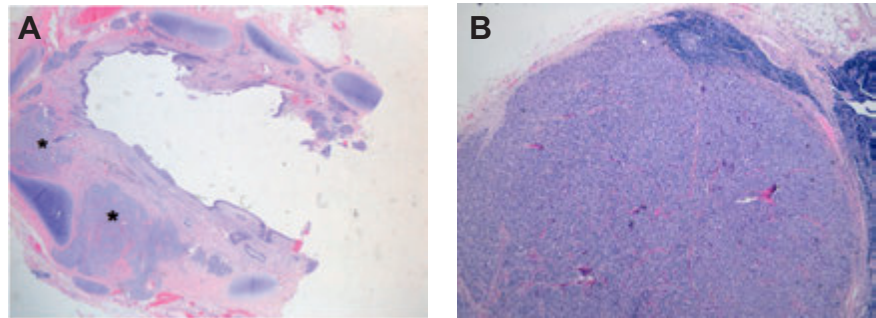


Figure 3. Surgical specimen examination: A) Bronchial wall (Hematoxylin & Eosin with magnifying glass) showing erupted epithelium corresponding to the former implantation local of the endobronchial mass and foci of carcinoid tumor within the bronchial wall (*); B) lymph node structure (Hematoxylin & Eosin 4×10) completely replaced by tumor infiltration.

overall survival of 82% at 10 years. This case report highlights the importance of bronchoscopy as a diagnostic procedure in atelectasis, even in pediatrics.

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