

## Case Report

# Bronchial carcinoid in children

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### ABSTRACT

Bronchial carcinoids (BCs) are uncommon, slow growing, low-grade malignant neoplasm comprising 0.5-2.5% of all primary lung cancers. Although BC's in childhood often have an endobronchial location causing airway obstruction, they are frequently misdiagnosed as benign conditions, resulting in a delay in definitive diagnosis and treatment. Lung sparing surgery should be done whenever possible, more so in children, for better quality of life and minimizing skeletal abnormality which follows pneumonectomy. We present two cases of bronchial carcinoid in young children.

**Keywords:** Bronchial carcinoid, Lung sparing surgery, Pneumonectomy

### INTRODUCTION

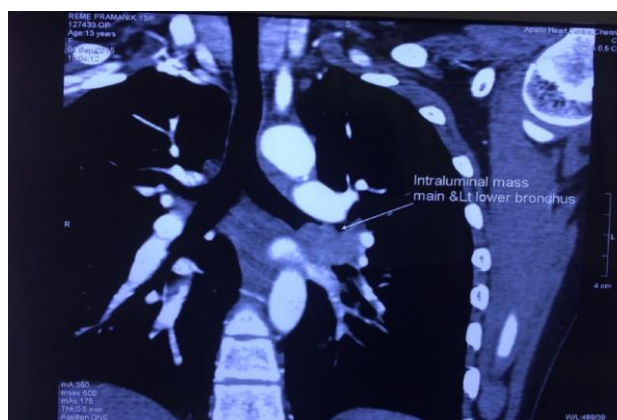
Bronchial carcinoid tumors (BCTs) are an uncommon group of lung tumors first reported by Laennec in 1831. They arise from the kulchistky cells or enterochromaffin cells of the Bronchial mucosa which forms part of the neuroendocrine system. Carcinoid tumors account for 0.5-2.5% of all primary lung tumors and are generally considered malignant.<sup>1</sup> BCTs though a rare entity in the pediatric population, make up 80-90% of the group of tumors that were formerly classified as bronchial adenomas and included adenoid cystic carcinomas and mucoepidermoid carcinomas.<sup>2</sup>

### CASE REPORT

#### Case 1

A thirteen-year-old girl presented with complaints of recurrent episodes of chest pain, cough and hemoptysis of 3 months duration. Initially she was evaluated elsewhere for TB and other infections. But since her symptoms persisted she came to our Institute for further evaluation. Routine blood investigations including renal and liver function tests were unremarkable. Echocardiogram was

normal. Sputum for AFB smear, bacteriology and fungal stain were negative. USG Abdomen was normal. Chest X-ray looked apparently normal. However, CT Thorax showed a tumor mass in the Left main bronchus (LMB) extending into the Left Lower Lobe (LLL) bronchus (Figure 1).

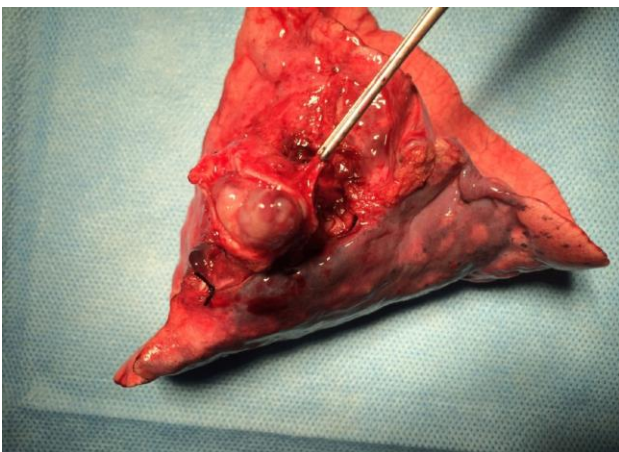


**Figure 1:** CT thorax showing intraluminal mass in the left main bronchus(LMB) extending into the left lower lobe (LLL) bronchus.

The features were s/o Carcinoid tumor. Bronchoscopy confirmed a polypoid shiny mass with overlying normal epithelium almost completely occluding the Left main bronchus (Figure 2).



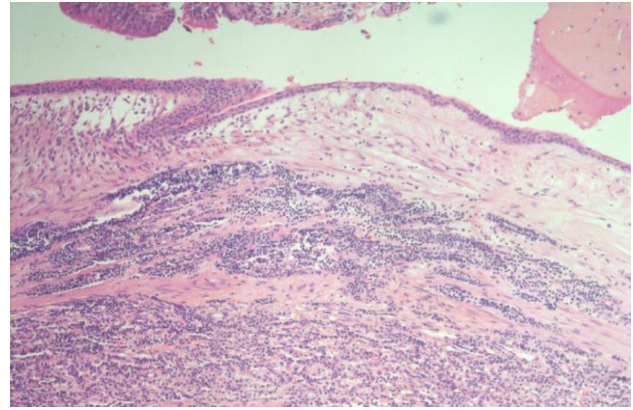
**Figure 2: Bronchoscopy picture showing a polypoid shiny mass with overlying normal epithelium almost completely occluding the Left main bronchus.**



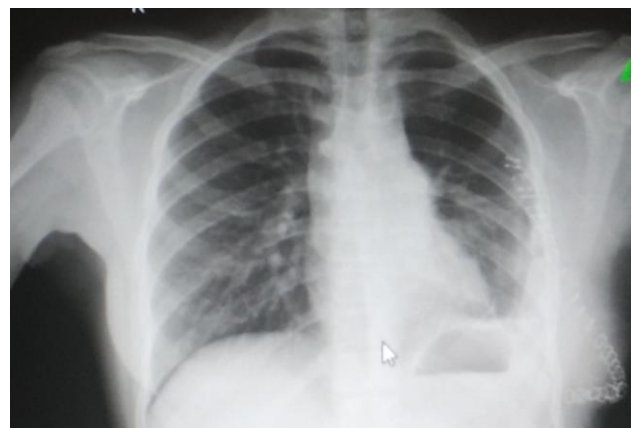
**Figure 3: Post-operative specimen showing the LLL with Tumor mass within it.**

Patient underwent Sleeve Left Lower Lobectomy (Figure 3) with anastomosis of the Left main bronchus to the Left upper lobe bronchus. HPE showed well differentiated carcinoid tumor (Figure 4). (mitosis  $<2/10$  HPF, no necrosis was present). The mediastinal nodes were free of tumor cells. Resection margins were clear. Post-operative period was uneventful. Repeat (post-operative) chest X-ray showed completely expanded Left upper lobe (LUL) (Figure 5).

Follow up CT Thorax was done after 6 months which showed well expanded LUL. Bronchoscopy showed no residual tumor with patent segmental bronchi (Figure 6).



**Figure 4: HPE showing well differentiated carcinoid tumor.**



**Figure 5: Post-operative Chest X-ray showing completely expanded Left upper lobe(LUL).**



**Figure 6: Follow up bronchoscopy after 6 months showing no residual tumor with patent segmental bronchi.**

## CASE 2

An eight-year-old male child presented with fever, cough and streaky hemoptysis of one-month duration.

Routine blood investigations including renal and liver functions were normal. Sputum examination was negative for TB and bacterial infections. CT scan of chest showed a central mass in the left lower lobe with bronchial obstruction s/o Carcinoid tumor (Figure 7). 24hr urine serotonin levels were normal.

Bronchoscopy showed a shiny polypoid mass lesion almost completely obstructing the LLL, s/o Carcinoid tumor. Biopsy was not attempted due to the risk of bleeding. He underwent left lower lobectomy. Histopathology confirmed carcinoid tumor with involvement of Left sided hilar (N2) node. The

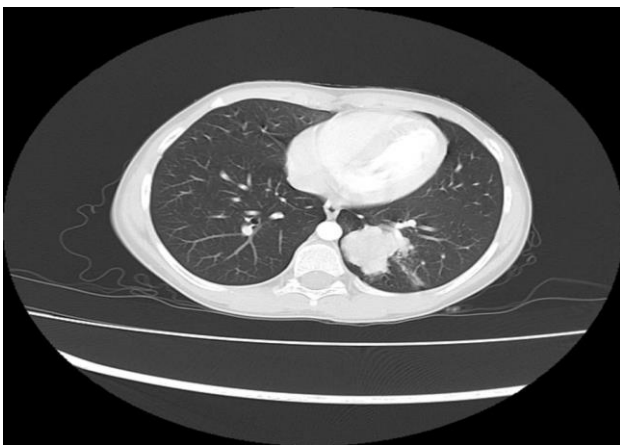
postoperative X-ray showed well preserved LUL (Figure 8).

**DISCUSSION**

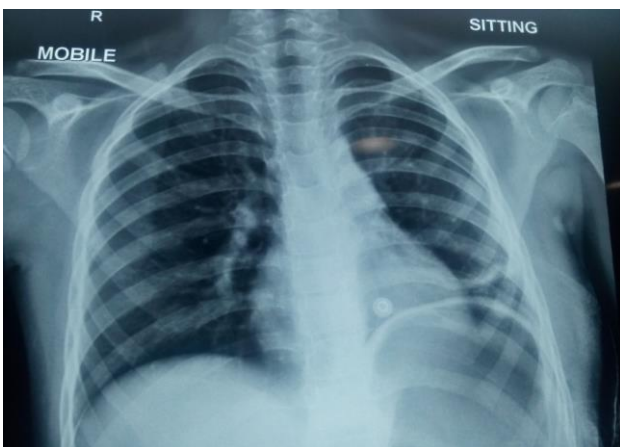
Lung is considered the second most common site of occurrence of carcinoid tumor. Right side is the most commonly involved site in most of the reported series.<sup>3,4,5</sup> Both the patients in our case had tumor in the left bronchus. Moreover, involvement of the main bronchus is rare as seen in Case 1. The characteristic features in both the cases are shown in Table1.

**Table 1: Shows the characteristic features and difference in both the cases.**

Case	Age	Sex	Site	Clinical presentation	Histology	Surgery	Nodal status
Case1	13yrs	F	LMB	Cough, chest pain, hemoptysis	Typical	Sleeve left lower lobectomy	Clear
Case2	8yrs	M	LLL	Fever, cough, hemoptysis	Typical	Left lower lobectomy	Left hilar (n2)



**Figure 7: CT scan of chest (case II) showing a central mass in the left lower lobe with bronchial obstruction s/o Carcinoid tumor.**



**Figure 8: Postoperative X-ray showing well preserved LUL.**

The 2004 WHO classification recognizes two types of carcinoid tumor-typical and atypical.<sup>6</sup> Carcinoid syndrome is rare (2-5% of cases) and is not produced by bronchial carcinoids unless liver metastases are present.<sup>7</sup> Apart from symptoms like chest pain, pleural effusion, cough, wheeze, hoarse voice, or atelectasis, common pulmonary manifestations are hemoptysis (18%), post obstructive pneumonitis (17%), and dyspnea in 2% of patients.<sup>8</sup> But, given the rarity of Bronchial carcinoids in children the diagnosis always remains a challenge. The first patient in our study had recurrent symptoms for almost 3months and she was being evaluated for TB and other infections.

Moreover, the CXR also looked normal. Recurrent pneumonia, atelectasis affecting the same lobe with persistent symptoms should alert the physician and more extensive and invasive investigations like CT scan and bronchoscopy should be done to arrive at a diagnosis. Nevertheless, TB should always remain as a differential diagnosis especially in endemic countries like India.

Tracheobronchial sleeve resection is the treatment of choice for BC tumours in adult population as it gives the best short- and long-term outcomes.<sup>9</sup> Gaissert et al proposed the procedure in the paediatric population and presented a series of 12 patients (aged 8-19years old), including four with BC.<sup>10</sup> Later, Rizzarda et al reported case series of 15 children who underwent 10 parenchyma-saving procedures (five sleeve lobectomies, three sleeve resections of the main bronchus and two bronchoplasties).<sup>11</sup>

In young patients, when technically possible, lung sparing resections should be performed. In these operations oncological results are similar to pneumonectomy but with a better quality of life and

without skeletal problems related to growth.<sup>11</sup> Pneumonectomy is a high-risk procedure correlated with a poor quality of life and non-harmonic growth of the chest.<sup>12</sup>

Wilkins and colleagues recognized the impact of bronchoplastic procedures (sleeve resection) in the management of bronchial carcinoid tumors.<sup>13</sup> Because a minimal margin is all that is required for curing these tumor bronchoplastic procedures allow removal of the tumor and preservation of sufficient portion of lung. Compared to the right bronchial sleeve resection, the second carina reconstruction on the left side is much more complex and difficult.<sup>14</sup>

First of all, because of the aortic arch and thoracic aorta around the hilum, the relative positions are deeper when not only free of the left main bronchus and pulmonary artery. Also, the acute angulation of the LUL bronchus with the LMB makes the anastomosis more difficult and technically challenging. Secondly, compared to the right bronchus located at the rear side of the pulmonary artery, left pulmonary artery is through the top of the left main bronchus, forming the pulmonary arch.

So, when reconstructing the second carina on the left side, the anastomosis of upper lobe bronchus and the left main bronchus is often hidden in the rear of the left pulmonary artery. The first patient in our series had the tumor almost completely occluding the LMB extending into the LLL. Had it been not for the sleeve procedure and the surgical expertise, the patient would have parted with the whole Left lung crippling her for the rest of her life. The second patient had tumor in the LLL which only required a Lobectomy. 5 to 20 percent of typical bronchial Neuroendocrine tumors and 30 to 70 percent of atypical tumors metastasize to lymph nodes.<sup>15,16</sup>

A complete mediastinal lymph node sampling or dissection at the time of initial treatment is indicated with complete resection of nodal metastasis if at all possible.<sup>17,18</sup> Metastatic involvement of mediastinal lymph nodes does not preclude a complete (R0) surgical resection or long-term cure. Though there is difference of opinion regarding postoperative adjuvant chemotherapy or radiotherapy in patients with mediastinal node involvement, a lack of benefit from adjuvant therapy was seen in node positive typical carcinoid tumors, in a retrospective analysis of 629 patients from the National Cancer Database.

The use of chemotherapy was not only associated with poor survival advantage, but also the outcomes in the treated group were worse compared to those who did not receive chemotherapy.<sup>19</sup> The second patient in our series had left hilar node involvement (N2). After discussion with the oncologist it was decided to manage with only surgical dissection and follow up. The rarity of bronchial carcinoid in children, technical challenges and surgical

expertise required for a sleeve resection surgery are the reasons for reporting our cases.

## CONCLUSION

BCTs though rare, are the most common primary lung tumor in children. Lung sparing surgery should always be attempted in young children for better quality of life. Bronchoplastic procedures (sleeve resection) is a difficult and technically demanding procedure which can be undertaken through proper patient selection and with surgical expertise.

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