Bronchial Carcinoid in a Child: A Case Report

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Bronchial carcinoid (BC) make up 80-90% of a group of lung tumors formerly called “bronchial adenomas,” which also included adenoid cystic carcinoma and mucoepidermoid carcinoma; hence, their actual incidence was not accurately known. It is a rare entity in the pediatric population where metastatic lung tumors greatly outnumber primary lung tumors. BC account for 2-5% of all lung neoplasms in adults but are the most common primary lung neoplasm of childhood, accounting for 80% of malignant bronchopulmonary neoplasms in children. We have presented a case of left BC in an 11-year-old boy, with a short review of literature, and discussed the management. We have discussed the surgical procedure in detail, pointing out the possible pitfalls and their management.

Keywords: Carcinoid, Bronchial carcinoid, Bronchial sleeve resection

INTRODUCTION

Carcinoid tumors are rare, malignant, neuroendocrine neoplasm first described in 1888 in the ileum and called “Karzenoide” by Oberndorfer in 1907. Neuroendocrine cells were earlier called “clear cells” and later “amine precursor uptake and decarboxylation” system cells. The term “neuroendocrine” refers to integration between nervous and endocrine elements. The hypothalamus controls the endocrine cells which secrete bioactive amines for a purpose. Neuroendocrine tumors of the lung arise from bronchial mucosal cells known as “enterochromaffin cells” or “Kulchitsky cells.” These cells are a part of the diffuse neuroendocrine system. Bronchial carcinoids (BC) are classified as typical carcinoid (TC) and atypical carcinoids (AC). The two have distinct biological features, clinical behavior, and prognosis. The difference in histology was first described by Arrigoni et al. and later modified by Travis et al. The final description was stated by the World Health Organization in 1999.

TC has a low-grade malignancy profile. Histology shows <2 mitosis/10 high power field (HPF), nuclear pleomorphism, and absence of necrosis. They rarely metastasise. AC is an intermediate-grade malignancy. Its histology shows ≥2 but <10 mitosis/10 HPF and/or coagulative necrosis. Carcinoids are consistently associated with immune histochemical expression of neuroendocrine markers such as chromogranin A, neuron-specific enolase, synaptophysin, and Leu7. Genetic research on BC has revealed under-representation of 11q in the DNA quite frequently. In AC, there is also a loss of 10q and 13q suggesting its relation to a more aggressive behavior.

EPIDEMIOLOGY

BC make up 80-90% of a group of lung tumors formerly called “bronchial adenomas,” which also included adenoid cystic carcinoma and mucoepidermoid carcinoma; hence, their actual incidence was not accurately known. It is a rare entity in the pediatric population where metastatic lung tumors greatly outnumber primary lung tumors. Among primary lung tumors in childhood, malignant are three times more common than benign neoplasms (papillomas, leiomyomas, hemangiomas, inflammatory myofibroblastic tumors, and hamartomas).

BC account for 2-5% of all lung neoplasms in adults but are the most common primary lung neoplasm of childhood,
accounting for 80% of malignant bronchopulmonary neoplasms in children. Lung masses in children are 10 times more likely to be benign, developmental or reactive lesions than a neoplasm. Common congenital malformations are cystic adenomatoid malformation, bronchogenic cyst, segmental bronchial atresia, sequestration, and congenital lobar emphysema. Other lung masses are inflammatory, infectious or reactive processes which include abscess, pneumonia, septic embolus, infarction, hematoma and granuloma formation (fungal, mycobacterial, parasitic, sarcoidosis, and vasculitis). Foreign bodies are also fairly common.

**CASE REPORT**

Our patient was an 11-year-old male who had been suffering from repeated bouts of respiratory infection for the last 6 months. He presented with a cough, fever, and chest pain on the left side for the last 20 days. He also had loss of appetite and weight over 6 months. He was treated for pneumonitis and referred to us with multiple chest X-rays (CXR) and a recent contrast-enhanced computed tomography (CECT) of the chest. He had a cough with expectoration, fever (101°F), tachycardia (148/min), tachypnea (40/min), and SpO₂ 89%. He weighed 21 kg, while 3 months ago he weighed 25 kg. On examination, the air entry to the right lung was good throughout with occasional crepts at the base; the left lung had breath sounds in the upper third and apical region only with crepts. There was no air entry or breath sounds in the mid and lower third of the left chest, which was stony dull on percussion. The earlier CXRs showed a white out on the left side (Figure 1a) and in the later ones the upper lobe of the left lung was visualized. The CECT chest reported an intrabronchial, homogeneously enhancing mass in the left main bronchus, near tracheal bifurcation, with luminal occlusion and left lung collapse (Figure 1b). Bronchogram pictures also confirm the presence of a left bronchial space occupying lesions (Figure 2a and b).

He was put on broad spectrum antibiotics and supportive measures, oxygen by mask and chest physiotherapy. On oxygen, his SpO₂ rose to 96%. After 2 days of treatment, he stabilized and was taken up for bronchoscopy. The findings were as follows. The trachea and right bronchus were clear; the left bronchus was completely occluded by a glistening, smooth, soft tissue growth which would bleed on touch. Bronchial lavage was done for growth cultures and multiple punch biopsies taken from the mass with an optical forceps. There was sufficient bleeding to require a dose of tranexamic acid (200 mg). The biopsy reported a well differentiated neuroendocrine tumor, Grade 1 of 3, BC. Immune-histochemistry on the sections was positive for CK5, CD56, synaptophysin and chromogranin; negative for leukocyte common antigen. The relevant investigations are listed in Table 1. A whole body skeletal scintiscan did not show any areas of abnormal concentration of radiotracer in the skeletal system.

It was decided to proceed with a left bronchial sleeve resection of the tumor area, lung-sparing surgery, with the use of frozen section biopsy to get complete clearance of tumor.

**Operative Details**

Right lung ventilation was done by inserting a cuffed endotracheal tube into the right main bronchus. The patient underwent a left posterolateral thoracotomy through the 4th rib bed without rib resection. A left sided bronchial sleeve resection was done saving the left lung. Approach was transpleural, as for patent ductus arteriosus repair, retracting the lung downward and forward, incising the left pulmonary ligament inferiorly and the pleura over the root of the left lung. The window of approach was surrounded by the left pulmonary artery (LPA) anteriorly, the ascending.

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**Table 1: Specific investigation reports of the patient**

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Report</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum cortisol</td>
<td>13.5 μg/dl</td>
<td>5-25 μg/dl</td>
</tr>
<tr>
<td>LDH</td>
<td>529 U/L</td>
<td>230-400 U/L</td>
</tr>
<tr>
<td>Chromogranin A</td>
<td>9.86 ng/ml</td>
<td>&lt;100 ng/ml</td>
</tr>
<tr>
<td>ACTH</td>
<td>13.4 μg/ml</td>
<td>&lt;46 μg/ml</td>
</tr>
<tr>
<td>5HIAA (24 h urine)</td>
<td>0.02 mg/24 h</td>
<td>0.7-8.2 mg/24 h</td>
</tr>
</tbody>
</table>

LDH: Lactate dehydrogenase, ACTH: Adrenocorticotropic hormone, 5HIAA: 5-hydroxyindoleacetic acid
arch and descending aorta in the anterosuperior, superior and posterior aspect, the left vagus nerve was running down the left of the mediastinum posterior to the root of the left lung. Its branch the left recurrent laryngeal nerve, hooked round the aortic arch next to the ligamentum arteriosum and travelled upward. At the outset, after exposure of the contents of the left lung root and part of the adjoining mediastinum, we initially hooked up in vesiloops/vascular tape, the aorta, the left vagus nerve, the left recurrent laryngeal nerve, and the LPA. These loops were color coded and also used to retract the structures they were holding (Figure 3). The initial dissection exposed the left bronchus within the curve of the aorta and posterior to the left and main pulmonary arteries. The ligamentum arteriosum was transfixed and ligated. The proximal part of the left bronchus and part of trachea and right bronchus at the carina were exposed. The lowermost part of trachea was dissected and looped so that it could be pulled toward the surgeon when required.

We then proceeded to dissect the left main bronchus all round and could palpate the tumor about 2 cm from the carina. We incised over it superiorly and took a good 1-2 mm margin all around its attachment taking the bronchial sleeve out (Figure 3). This was sent for a frozen section study. The left lung was disconnected from the trachea with intact vascularity. While waiting we dissected hilar and mediastinal nodes for biopsy. We inflated the left lung once with sterile tubing and oxygen for confirmation.

Frozen section biopsy reported clear margins of the resected specimen; we proceeded to prepare for a left bronchial anastomosis to the short cuff of proximal bronchus at the carina. As the two ends of the bronchus were under tension, a circular incision was given inferiorly on the pericardium, around the left lung root vessels below the left inferior pulmonary vein and completed all round. The pre-tracheal dissection was done and the tracheal traction applied with vesiloops around the trachea. The lung was mobilized upward by placing sponge packs inferiorly. The two ends of the left bronchus now came together and repair was done with 4/0 polydioxanone (PDS from Ethicon, Johnson and Johnson Ltd., India) interrupted sutures. The anastomosis was not covered with a flap as it lay well within the mediastinum (Figure 3). Two chest tubes were placed connected to an underwater sealed drainage system and closure was done. The patient was kept ventilated and pain-free and was gradually weaned off the ventilator on the second postoperative day. In his immediate postoperative CXR, he had collapse-consolidation of the right upper lobe. This improved with regular chest physiotherapy. The chest drainage reduced significantly by the third postoperative day and the drains were removed (serial X-rays). He was given cefuroxime and amikacin for 1 week. Good expansion of left lung occurred before discharge. All nodes were negative for tumor cells, and the histopathology (Figure 4a) of the specimen reported BC with clear margins. The patient is now 2½ years in follow-up and doing well (Figure 4b).

**DISCUSSION**

Through this case report we wish to highlight the problems faced during a bronchial sleeve resection.7,8 The approach to the left bronchus is between the main and LPA anteriorly and the arch of aorta and descending aorta, superiorly and posteriorly. The ligamentum arteriosum with the recurrent left laryngeal nerve adjacent to it are to be watched for at all times. In our patient, the carcinoid was about 1-2 cm from the carina. If our incision had gone right up to the carina, we would probably not have been able to anastomose the left bronchus to the trachea. In that case, we had the alternatives of anastomosing to a higher level or a pneumonectomy; sacrificing a normal lung. Certain manoeuvres help in this situation allowing a tension free anastomosis. Dissection of the lower trachea, particularly in the anterior region, and holding it in multiple vesiloops with traction helped in the anastomosis. Pericardial lift and laryngeal release are other helpful manoeuvres. Pericardial section in “U” shape below and encircling the inferior pulmonary vein.

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**Figure 3:** (a) Resected sleeve of left bronchus with carcinoid in situ. (b) Left bronchial anastomosis after resection shown by arrow. The inlayed text box explains the color coding of the slings (LPA - Left pulmonary artery, anas - anastomosis)

**Figure 4:** (a) Nests of uniform round tumor cells infiltrating the smooth muscle in the bronchial wall (H and E, 200). (b) Chest X-ray of the patient 1 month after surgery
helps in lifting up the left bronchus by up to 2 cm or more depending on individual variation. Laryngeal release, mainly thyrohyoid release, and suprahypoid release are expected to give another 1 cm drop of the trachea increasing length; authors who have used this technique do not find it very useful for lower tracheal or carinal regions. Sleeve resection of the pulmonary artery is the final weapon in the armamentarium. This requires the heart to be put on bypass and the involvement of a cardiac surgeon. By shortening the LPA the trachea and left bronchus can be brought together.

CONCLUSION

Nonsecreting BC is the most common bronchopulmonary malignancy in the pediatric age group. It becomes necessary to do a bronchial sleeve resection instead of an endoscopic excision as the nature of the tumor is to spread through the wall of the bronchus and not laterally along it. The ipsilateral lung is usually normal and must be saved. Hence, a bronchial sleeve resection is the ideal procedure for this condition. Having said that, for undertaking a bronchial sleeve resection, especially on the left side, a good amount of surgical expertise is required. It also requires a good Intensive Care Unit backup, anesthetic expertise and the availability of cardiothoracic surgical backup. One should be prepared for all eventualities before undertaking such a procedure.

REFERENCES


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