Case Report

Bronchial Carcinoid Tumor in an Adolescent Female: Diagnosis and Management by a Multi-Disciplinary Team

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Bronchial carcinoid is the most common primary malignant lung tumor in children; however, it remains a very rare diagnosis due to the overall low incidence of childhood lung malignancies. We report a case of a 17-year-old girl with respiratory symptoms who was initially misdiagnosed as a case of COVID pneumonia. She was later detected to have a right mainstem bronchial carcinoid which was managed successfully by a multi-disciplinary team.

Keywords: Bronchial, carcinoid, pediatric, pneumonectomy, surgery

INTRODUCTION

Primary malignant lung tumors in children are extremely rare and account for only 0.2% of all cases of childhood cancer.[1] Among these, bronchial carcinoids (BC) are the most common, comprising between 63% and 80% of all cases.[2,3] BC in childhood often has an endobronchial location causing airway obstruction and is frequently misdiagnosed as benign conditions, resulting in a delay in definitive diagnosis and treatment. Surgery represents the treatment of choice for BCs, and if promptly diagnosed and radically treated, BC in children has an excellent prognosis.[4]

We report a case of a 17-year-old girl with persistent respiratory symptoms who was initially referred to us as a case of COVID pneumonia. She was later diagnosed to have a right main bronchus carcinoid which was managed successfully by a multi-disciplinary team.

CASE REPORT

A 17-year-old girl was referred to us as a suspected case of COVID pneumonia with complaints of cough, dyspnea, chest pain, low-grade fever, and intermittent hemoptysis for 2 weeks. She reported multiple similar episodes in the past 2 years. On physical examination, she was tachypneic with absent air entry on the right side with a rightward shift of the trachea. Oxygen saturation on room air was 97%. COVID reverse transcriptase-polymerase chain reaction test was false positive from the referring hospital and was confirmed to be negative at our institute. Chest X-ray showed ground-glass opacity of the entire right lung field. Computed tomography (CT) scan of the chest revealed a peri-hilar soft-tissue density lesion, approx. 5.4 cm × 4.2 cm × 3.1 cm in size, completely obliterating the lumen of the right main bronchus, associated with the complete collapse of the right lung with mediastinal lymphadenopathy [Figure 1a]. Pulmonary function test parameters were approximately 50% of their predicted value.

The patient underwent a diagnostic flexible bronchoscopy which showed a fleshy mass completely covering the bronchial opening.

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obliterating the take-off of the right main bronchus at the carina. Suspecting an endobronchial carcinoid, tumor-specific laboratory tests were done which revealed a raised serum chromogranin-A level (214.3 ng/ml) and a normal 24 h urinary excretion of 5-hydroxyindolacetic acid. 68-GA-DOTANOC positron emission tomography-CT (PET-CT) whole-body scan showed tracer uptake in the peri-hilar soft-tissue lesion and mediastinal nodes suggesting a somatostatin receptor expressing tumor consistent with BC [Figure 1b].

After written informed consent, the patient was taken to the operating room. Left main bronchus was intubated with 35 Fr left-sided DLT (double-lumen tube) and position confirmed with fiber-optic bronchoscopy. On posterolateral thoracotomy, the right lung was found completely solidified and full of pus with extensive pleural adhesions. A large hilar mass was palpable which was completely obstructing the right main bronchus and reaching up to the carina. Due to dense peri-hilar adhesions, the pericardium was opened and right pulmonary vessels were dissected by an intra-pericardial approach. Right pulmonary artery, superior and inferior pulmonary veins were dissected, ligated, and divided. The right main bronchus was divided flush with trachea at the carina and the pneumonectomy specimen with endobronchial tumor was removed. The right bronchial stump was repaired with nonabsorbable sutures and reinforced with a muscle flap. All grossly enlarged lymph nodes (para-tracheal and sub-carinal) were excised.

Postoperatively, the patient was ventilated for 24 h and required oxygen support for another 48 h. She was discharged after 2 weeks in healthy condition. At 6 months follow up, she is asymptomatic with no evidence of residual or recurrent disease on 68-GA-DOTANOC PET-CT imaging with well-expanded left lung on x-ray [Figure 1c] with normal serum chromogranin-A level. On histopathology, the sections from the mass [Figure 2a] showed a tumor disposed of in nests and lobules separated by fibrous septae. The tumor cells showed mild anisonucleosis, with round-to-oval nuclei, stippled chromatin, small nucleoli, and a moderate amount of eosinophilic cytoplasm [Figure 2b]. On immunohistochemistry, the tumor cells were positive for synaptophysin and chromogranin [Figure 2c]. The mitotic figures were scarce (<2/10 HPFs) and the Ki-67 proliferation index was <2% [Figure 2d]. The tumor was seen infiltrating the wall of the right main bronchus; however, the resection margin was free of tumor. Metastasis was noted in 2 out of 10 lymph nodes. The findings were consistent with a typical carcinoid tumor.

**DISCUSSION**

In recent times, any respiratory symptoms are considered to be due to COVID pneumonia until proven otherwise.
As in the case of our patient, the presence of a tumor may only become apparent after investigations such as CT chest or bronchoscopy for recurrent episodes. Other presentations like classic carcinoid syndrome are very rare in BC and is generally associated with metastatic disease.[4] The most common site of metastasis is the intrathoracic lymph nodes, although liver and skeletal metastases have been described in children.[5] Our patient did not have any symptoms of carcinoid syndrome, although the tumor had metastasized to the mediastinal lymph nodes.

Somatostatin receptor scintigraphy, also called an octreotide scan, used to have a role in the diagnosis and follow-up of BC in addition to CT. In recent times, this has largely been replaced with 68-GA-DOTANOC PET-CT imaging. In a recent meta-analysis, the authors reported that both 68Ga-DOTA-peptide and 18F-FDG are highly sensitive in detecting pulmonary carcinoid, while 68Ga-DOTAPeptide is more sensitive than 18F-FDG (90.0% vs. 71.0%).[6] In our patient, the bronchoscopic biopsy was not taken to avoid the risk of hemorrhage into the single functioning left lung. However, the diagnosis was confirmed by the uptake of tracer by the primary tumor and mediastinal lymph nodes on 68-GA-DOTANOC PET-CT. This was also further supported by the raised serum chromogranin-A level (214 ng/ml).

Surgical resection represents the treatment of choice for BC, achieving long-term survival in cases of radical resection in both adults and children.[4] Pneumonectomy has been the most commonly reported procedure for BC in children, but in more recent years, the use of pneumonectomy has declined.[3,4] It is now recommended that in young patients, lung-sparing resections should be performed when technically feasible; provided basic oncologic goals and principles are not compromised. Lung-sparing resections comprise bronchoplasty, wedge or sleeve resections of the trachea or main bronchi or lobectomies associated with bronchial wedge or sleeve resection.[6] The indications for pneumonectomy are now limited to cases like ours, where a large tumor involves the main stem bronchus with irreversible destruction of the ipsilateral lung.

The relevance of lymph node sampling or radical lymph node dissection is not well established in children; however, most authors recommend systematic lymphadenectomy due to the risk of lymph node metastasis (approximately 20%) and the possibility of recurrence many years later.[4] At this time, there is no curative treatment for metastatic disease. Somatostatin analogs, interferon-alpha, and Temozolomide monotherapy have been used in advanced disease in adults without significant benefit.[3] Long-term follow-up is strongly recommended in BC. Despite low-grade malignancies, recurrences have been reported and their prompt diagnosis and treatment is important for long-term survival.

In conclusion, pediatric BC is very rare tumors, usually presenting with nonspecific symptoms which often lead to misdiagnosis and treatment as benign disease for long periods before definitive diagnosis. Early detection and prompt treatment by complete surgical resection are the key to a successful outcome. Most children with BC tumors have a good prognosis and achieve long-term survival; however, a careful and prolonged follow-up is strongly recommended.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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