



# Surgical Treatment of a Neuromucoepidermoid Carcinoma of the Left Main Bronchus via Posterolateral Thoracotomy in a 4-Year-Old Boy

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## Abstract

### Keywords

- ▶ neuro-mucoepidermoid carcinoma
- ▶ bronchus resection
- ▶ child

We report the case of a 4-year-old boy with a neuromucoepidermoid carcinoma of the left main bronchus. Complete resection of the carcinoma and reconstruction of the carina between lower and upper lobe by means of an end-to-end anastomosis was performed via a left-sided thoracotomy.

## Introduction

Primary malignant bronchial neuroendocrine neoplasms are rarely found in childhood. Mucoepidermoid carcinoma of the lung, however, is very rare with a reported frequency of 0.01 to 0.2% of primary lung tumors.<sup>1</sup> Typical symptoms are of a respiratory nature and include wheezing, coughing, dyspnea, and recurrent obstructive pneumonia.<sup>2</sup> Owing to the critical anatomical location of the tumors, their surgical management presents a considerable challenge and, in all described cases, sternotomies were performed for surgical access.<sup>3,4</sup>

## Case Description

A 4-year-old boy was referred to our clinic for further evaluation of wheezing, coughing, increasing nightly pauses of breath of up to 30 seconds in duration, and progressive dyspnea observed in a sleep laboratory over a 2-month period. The relevant medical history of the child included an adenoidectomy and tonsillectomy 8 months prior, as well

as an otorhinolaryngological examination 2 weeks earlier. The otorhinolaryngological examination reported no pathological findings except for pneumonia. On admission, the child was dyspnoeic with inspiratory and expiratory stridor. A polysomnography showed peripheral oxygen desaturation of up to 83% and a mixture of obstructive and central apnoeic episodes. A chest radiograph revealed emphysema of the left lung with a mediastinal shift. A cardiac malformation was ruled out using transthoracic echocardiography. A computed tomography (CT) scan of the chest demonstrated an endoluminal broad-based tumor with incomplete obstruction of the left main bronchus. A rigid bronchoscopy under general anesthesia was performed to obtain a biopsy prior to surgery (▶ Fig. 1A). After receiving the immunohistological diagnosis of neuromucoepidermoid carcinoma, surgery was planned with cardiopulmonary bypass standby facility. Bronchoscopically guided selective intubation of the right main-stem bronchus was performed for single-lung ventilation. The child was placed in a right lateral decubitus position and posterolateral thoracotomy was performed.

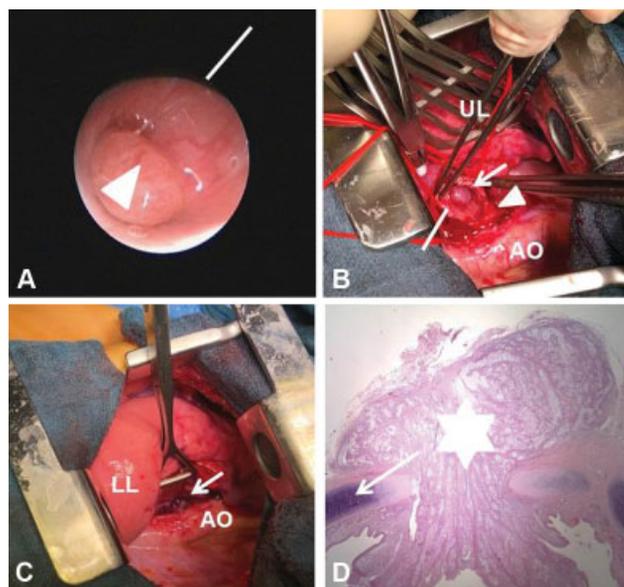
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**Fig. 1** (A) Preoperative rigid bronchoscopy demonstrating a large red sessile tumor (indicated by triangle). (B) Intraoperative image showing the endoluminal tumor after opening of the left main bronchus (solid line = left main bronchus; arrow head = esophagus; arrow = tumor, AO = descending aorta; UL = left upper lobe of the lung). (C) Intraoperative image after bronchus resection and completed end-to-end anastomosis (arrow head = end-to-end anastomosis; LL = left lower lobe of the lung, AO = descending aorta). (D) Histomorphological examination using haematoxylin and eosin staining (star = tumor; arrow = tracheobronchial ring).

Locoregional lymph nodes were dissected and excised for histomorphological examination. After identification of the left main bronchus, the vagus nerve and phrenic nerve as well as the left main pulmonary artery, were dissected and protected. Following mobilization of the left main bronchus from the esophagus, the left main bronchus was opened by longitudinal incision toward the carina, and the tumor was detected with involvement of the left main stem bronchus and spread up to the carina between upper and lower lobe. En bloc resection of two rings of the main bronchus was performed after meticulous dissection of segmental arteries of the upper and lower lobes (► Fig. 1B). The resection margins were checked intraoperatively by frozen section to verify complete tumor removal. End-to-end anastomosis with reconstruction of the carina between upper and lower lobe was performed by using 4-0 polydioxanone sutures. Running sutures were made on the paries membranaceous and interrupted sutures on the cartilaginous part of the bronchus (► Fig. 1C). The left pleural space was drained using a 16-French chest tube, the thoracotomy was closed in standard fashion, and the child was transferred to the pediatric intensive care unit. The child was extubated 4 hours after surgery. The postoperative course was uncomplicated. The chest drain was removed on the third day after surgery, and 10 days after the operation the child was discharged from the hospital in excellent general condition. The final pathological investigation demonstrated negative locoregional lymph node histology, which excluded meta-

static spread and confirmed the diagnosis of a neuromucoepidermoid carcinoma (► Fig. 1D). At 6-month follow-up, the child was asymptomatic and tumor free.

## Discussion

Neuromucoepidermoid neoplasms are rare tumors of the bronchial tree arising from the mucous and serous glands of the epithelium and have an age-adjusted incidence rate ranging from 0.2 to 2/100,000 population/year in both the United States and Europe.<sup>1</sup> Due to the fact that these neoplasms have a wide spectrum of biological activity and often show malignant behavior, they are now commonly referred to as “bronchial gland tumors,” rather than “bronchial adenomas,” as they were in the past. “Bronchial adenomas” include the following three different types of pathological entities: (1) carcinoid cystic carcinoma, (2) adenoid cystic carcinoma, and (3) mucoepidermoid tumor. Since 1986, only 15 cases of bronchial mucoepidermoid tumors in children, under the age of 14 years, have been reported.<sup>2</sup> These slow-growing neoplasms usually manifest with obstruction of the trachea-bronchial tree, and symptoms are related to their location in the trachea and the main bronchii. As in the case reported, they mainly cause upper respiratory tract occlusion with coughing, wheezing, and dyspnea, and only rarely produce symptoms like atelectasis and hemoptysis. In this young child, respiratory distress and relapsing pneumonia were undiagnosed until a bronchoscopy demonstrated the tracheal obstruction which was then confirmed by CT scan. Complete surgical resection via a range of different surgical access routes is the treatment of choice for this subtype of neuro-mucoepidermoid carcinoma. The key principles of surgery are complete locoregional lymph node removal, a tumor-free surgical margin at frozen section, and maximum sparing of lung parenchyma using tracheobronchial resection and reconstructive techniques. Endoscopic resection has been attempted, but the incidence of local relapse was unacceptably high. Papiashvilli et al have performed excellent and successful operations via sternotomy, which represents a difficult operative access to the distal trachea.<sup>3</sup> As stated by the authors, their decision to choose a sternotomy was based on “the difficulty of ventilating the child” and “the existence of immediate cardiopulmonary bypass facilities.” We performed a similar operation with a more comfortable surgical exposure of the lower trachea through a left-sided thoracotomy. We would have employed extracorporeal circulation via peripheral cannulation of the femoral vessels, if required. Yu and colleagues from Boston presented their own results with 40 patients between 3 month and 19 years within the past 90 years. Only 34 patients received surgical treatment, and most of them had major pulmonary resection, none were treated with isolated bronchial resection, although eight patients were suffering from a carcinoid and five patients from a neuromucoepidermoid tumor.<sup>5</sup> Therefore, such patients should be treated curatively, whenever possible, with parenchyma-sparing techniques. In this case report, we demonstrated and discussed the applied surgical technique the removal of a tracheal neuromucoepidermoid carcinoma in 4-year-old boy who was successfully treated with a

tracheal resection and reconstruction by means of a left-sided thoracotomy.

## Conclusion

In conclusion, these patients should be treated by an interdisciplinary team consisting of pediatric, thoracic, and congenital cardiac surgeons by applying a curative therapeutic approach including parenchyma-sparing techniques.

### Conflict of Interest

None declared.

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