Mucoepidermoid carcinoma of the airways in a young adult male

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Radiology Case. 2017 Feb; 11(2):8-15 :: DOI: 10.3941/jrcr.v11i2.3061

ABSTRACT

Mucoepidermoid carcinoma (MEC) of the airways is a rare entity most often found in young patients. We present a case of a 23 year old patient with symptoms of pneumonia, which progresses to a pulmonary abscess within a week. Diagnostic work-up reveals an endobronchial obstruction by a pedunculated low grade MEC. A literature review is provided and radiological appearances are described.

CASE REPORT

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A 23-year-old male presented with a fever and cough at his general physician. A chest radiograph showed a left lower lobe consolidation consistent with pneumonia (Figure 1). Antibiotics (amoxicillin/clavulanate 625 mg TID) were not sufficient and his symptoms progressed with hemoptysis. A second chest X-ray was performed six days later. The consolidation progressed in size and cavitation with gas-fluid levels developed. Due to mass effect of the lower lobe, bulging of the major fissure was present (Figure 2). The differential diagnosis of the conventional imaging was not specific and included an infection, abscess and neoplasia. A contrast enhanced computed tomography chest (CECT) scan was performed a day later showing a large lesion in the left lower lobe. Multiple air-fluid levels and fluid collections with capsular enhancement were present surrounded by consolidation (Figures 3 and 4). There were no enlarged mediastinal or perihilar lymph nodes. These findings were consistent with a pulmonary abscess, however, the etiology remained unclear.

Our differential diagnoses included bacterial infection, tuberculosis, fungal infection and airway obstruction due to a tumor. The latter seemed less likely given the age of the patient and the lack of risk factors. Following the CT scan our patient underwent a bronchoscopy, which revealed a pedunculated endobronchial tumor in the apex of the left lower lobe (Figure 5). Tissue of this mass was sampled and histopathologic analyses showed characteristics of an MEC. There was no necrosis and very low mitotic activity suggesting this was a low grade tumor. A positron emission tomography (PET)-CT scan was performed for oncologic staging eight days after the chest CT. There was no metabolic activity indicative of lymph node metastases or distant metastases. Only the consolidated lung parenchyma with central abscess showed fluorodeoxyglucose (F18-FDG) uptake without evidence for tumor mass (Figure 6). Based on the work-up, histopathology and the advanced irreversible damage to the left lower lobe, a lobectomy was performed (Figure 7). Analyses of the resected lung tissue showed tumor with glandular and solid components as well as lymphangio-invasion of tumor (Figure 8). Based on these findings the tumor was classified as a pT2aN0, stage 1B tumor. This is a low grade type of MEC tumor therefore no additional chemotherapy was necessary and follow up was considered to be enough.

Initially the patient suffered from post-operative chest pain and fatigue. After five months he was fully recovered without any residual constraint. A follow-up chest X-ray after five months shows normal post-lobectomy changes without complications (Figure 9).

DISCUSSION

Background:

MEC was first described by Smetana in 1952 [1]. The World Health Organization defined MEC as a salivary gland-type tumor characterized by a combination of mucus-secreting, squamous, and intermediate cell types [2]. MEC in general is not an uncommon malignancy. Usually it arises from the parotid and submandibular salivary glands [3]. MEC with primary origin in the airways however is rare with a reported incidence of about 0,1%-0,2% of primary lung tumors [4]. Available literature is limited and consists mainly of case reports and small series.

Clinical Features:

MEC is seen in patients from a wide age range; from 3-78 years. Young adults are mostly affected, with more than 50% of patients under the age of 30. Males and females are affected equally [2, 5-8]. MEC usually arises from minor submucosal salivary glands of the large airways [3]. Therefore signs and symptoms are consistent with large airway obstruction i.e. cough, hemoptysis, bronchitis, wheezing, fever due to superimposing pneumonia and chest pain. Asymptomatic disease occurs in the minority of patients.

Histopathological Findings:

MEC usually presents as an exophytic intraluminal mass, which can be either sessile, polypoid with a broad bronchial wall or pedunculated with a well-formed stalk. Size varies from several millimeters up to 6 centimeters [3]. Histologically, MEC is usually classified as low-grade or high-grade. High-grade features of a MEC tumor are necrosis, nuclear polymorphism, hyperchromasia and active mitosis (>4 per 10 high-powered fields). High-grade tumors have a tendency for mural invasion and lymph node metastasis. Low-grade tumors are mostly confined to the bronchus [9].

Radiological Findings:

Common findings on chest radiographs are those associated with obstruction, i.e. post obstructive pneumonia / pulmonary abscess and atelectasis. Chest radiographs can also reveal a pulmonary nodule or mass. CECT imaging is the modality of choice when it comes to pulmonary neoplasms. Xinchun Li et al [10] performed a retrospective analysis of the CT-findings of 16 patients with MEC. In their study several CT findings were found, of which we will describe two in this case report. Firstly a well-circumscribed mass within the bronchus or a peripheral pulmonary nodule. Secondly a thick-walled cavity seen in only one patient. Thirdly, in a single patient, the tumor appeared as a mass involving lung parenchyma, the trachea and main bronchial wall. Calcification was found in four cases but other authors describe a higher incidence of calcification up to 50% [11]. Calcification seems to be an important finding being a sign of tumor necrosis and therefore possibly high-grade malignancy. However, the results from different studies are not consistent. For example Wang et al [12] they described an enhancing homogenous endobronchial mass on CT suggestive of low-grade MEC. High-grade MEC tends to be more heterogeneous and lobulated with poorly defined margins. These show less enhancement and tend to be located more peripherally. In the population of Wang et al calcifications were found in only two of 17 patients and both were low-grade MEC. This indicates that calcification is not a sign of high grade malignancy.

PET-CT results are dependent on malignancy grade and size. Lymph node metastases can often be detected.

Differential Diagnoses:

When an endobronchial mass or nodule is present on CT or when there are signs of solitary bronchial obstruction on chest CT or X-ray MEC should always be in the differential diagnoses, especially when it concerns a young patient without any risk factors for other pulmonary neoplasms. Other conditions that should be considered are (non) small cell lung cancer (NSCLC), carcinoid, adenoid cystic carcinoma and endobronchial hamartoma. (N)SCLC and MEC can have similar imaging characteristics. Advanced disease at the time of presentation, however, is more common, e.g. lymph node metastases, lymphangitic carcinomatosis. Endobronchial carcinoid enhance on CECT. They are located endobronchial, peribronchial or perihilar and usually sharp defined round or oval masses or nodules. Calcifications occur in 30% of carcinoid cases. Post obstructive findings are also common in carcinoid [13]. Adenoid cystic carcinoma formerly was called cylindroma because of its tendency to manifest as a circumferential mass surrounding trachea or main bronchus. Diffuse wall thickening of the trachea, pedunculated or broad based lesions also occur as well as lymph node metastases. Calcifications are rarely encountered [14]. When an endobronchial mass or nodule is well defined and contains fat or fat and calcifications, endobronchial hamartoma is the most probable diagnosis [15]. Hamartoma and carcinoid usually do not show any FDG uptake.

Treatment & Prognosis:

There are several surgical options for the treatment of MEC (e.g. lobectomy, sleeve resection, local resection and endoscopic removal) [16]. Given the limited available literature there is little known about risk factors and prognoses. In a recently published study by Takefumi et al [17] tried to identify risk factors. Their multivariate analysis showed that age, distance metastasis and malignancy grade were significant determinants of the 5-year disease-specific survival for patients with primary lung MEC [17]. Jun-Jie Xi et al.[18] analyzed 21 cases of patients with MEC. Only one of the 17 patients with low-grade tumor passed away, but none of the four patients with high-grade malignancy survived. Their analysis showed that age, differentiation grade, lymph node metastasis and TNM stage influenced overall survival and progression free survival [18]. There is insufficient evidence for the efficiency of chemotherapy or radiation [19-21]. Available data does suggest that long-term follow-up is indicated, certainly for patients with lymph node metastasis, which is present in about 10% of patients.

TEACHING POINT

In case of an endobronchial nodule or mass, with or without signs of bronchial obstruction, mucoepidermoid carcinoma should always be in the differential diagnoses. If it concerns a patient under the age of thirty without risk factors for pulmonary malignancy suspicion should be even higher.

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FIGURES

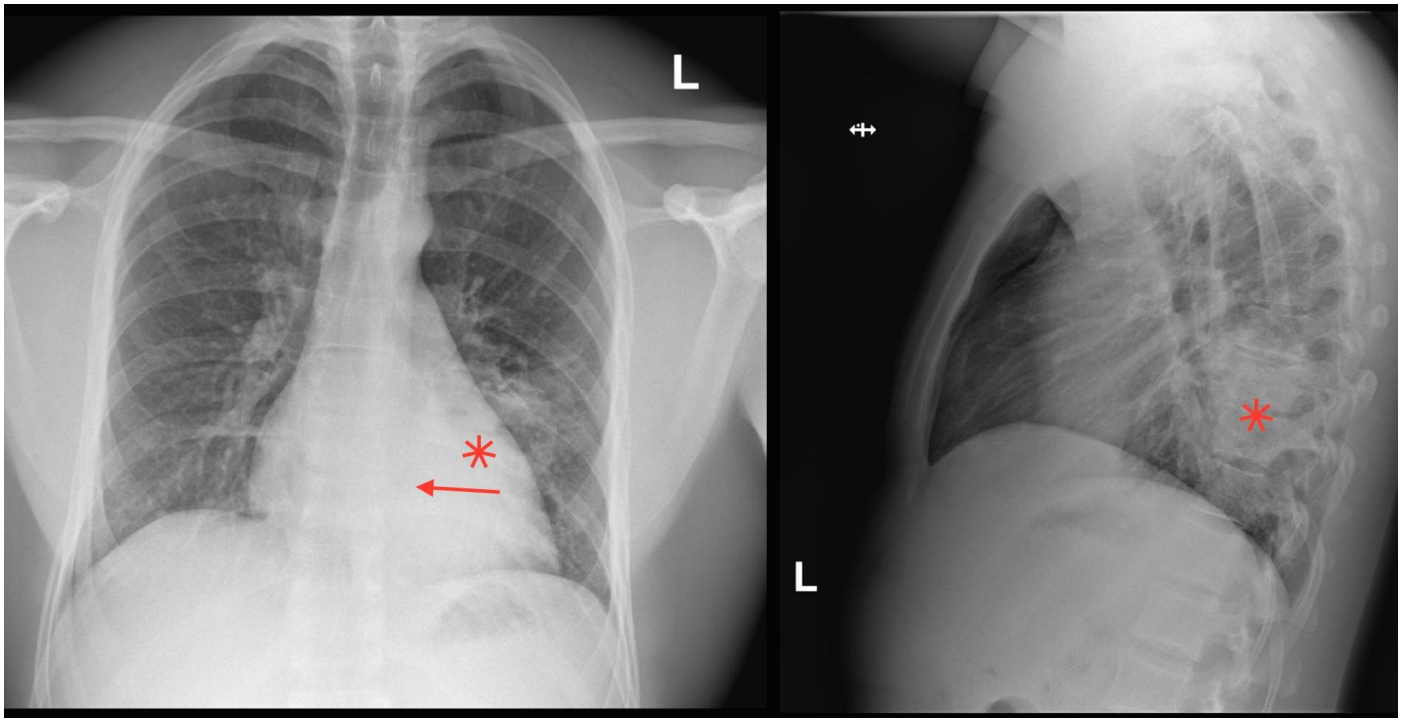

Figure 1: 23 year old male with post obstruction pneumonia due to mucoepidermoid carcinoma of the airways.
Technique: plain PA and lateral radiograph (Siemens. 0,2 mm CU filter. PA:125 kV; 2,7 mAs. Lateral: 125 kV; 16,5 mAs).
Findings: Consolidation in the left lower lobe (red asterisks) with silhouette sign of the descending thoracic aorta (red arrow).

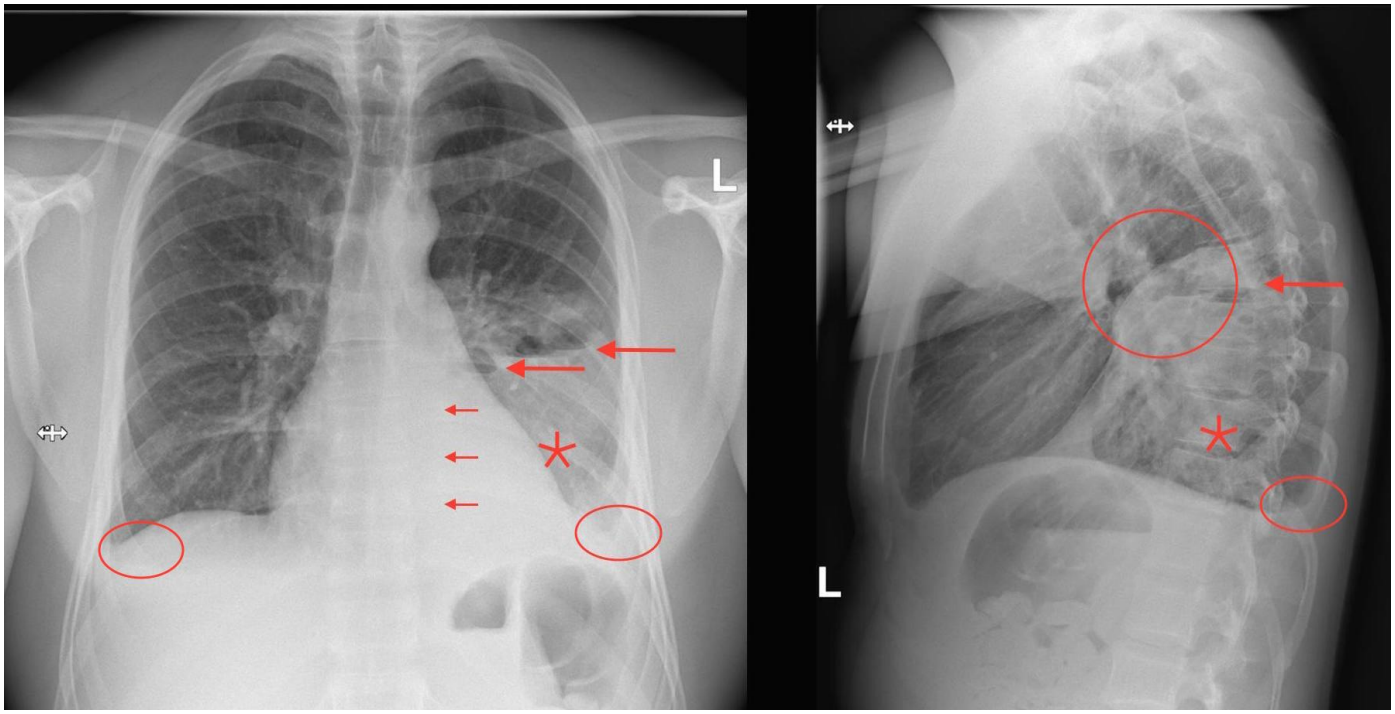

Figure 2: 23 year old male with post obstruction pneumonia and pulmonary abscess due to mucoepidermoid carcinoma of the airways.
Technique: plain PA and lateral radiograph (Siemens. 0,2 mm CU filter. PA:125 kV; 2,6 mAs. Lateral: 125 kV; 24,2 mAs).
Findings: Progression of consolidation in the left lower lobe (red asterisks) en silhouette sign of the descending thoracic aorta (small red arrows). Multiple gas pockets with fluid levels (large red arrows). Bilateral pleural effusion, predominantly on the left (red ovals). Anterior bulging of the major fissure due to mass effect in the left lower lobe (red circle).

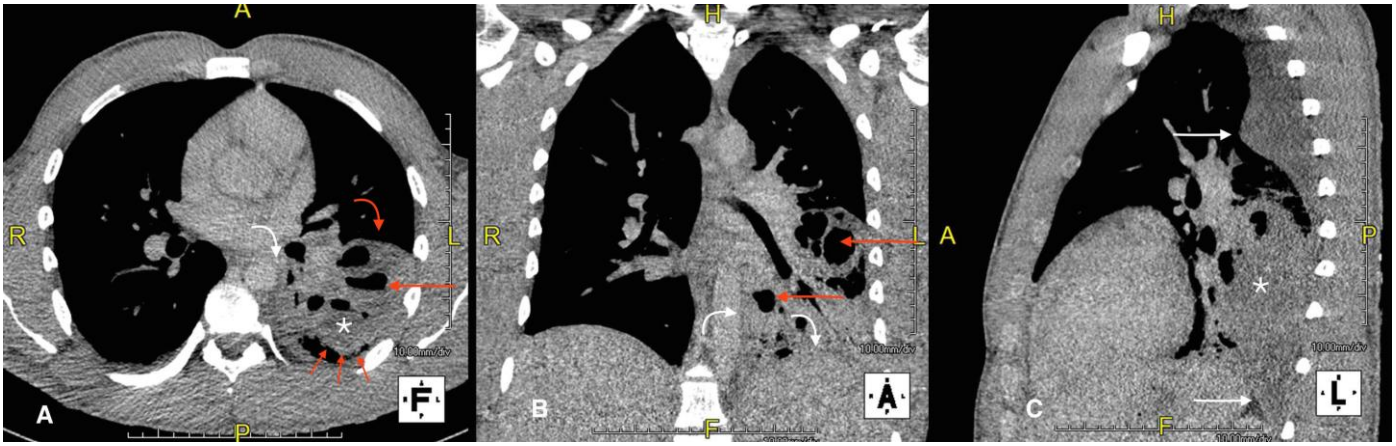

Figure 3: 23 year old male with post obstruction pneumonia and pulmonary abscess due to mucoepidermoid carcinoma of the airways.
Technique: chest CT scan after intravenous administration of 70 ml Xenetix© 300 (Siemens. 100kV; 188 mAs; 0,6 mm; 352 mGycm). Mediastinum window.
Findings: Consolidation in the left lower lobe (white asterisks on A and C). This is adjacent to the left hemi-diaphragm and the descending thoracic aorta (curved white arrows on A and B) explaining the silhouette sign on plain radiograph. Cavitation with gas-fluid levels (large red arrows on A and B) and subtle capsular enhancement surrounding some of the fluid filled pockets (small red arrows on A) consistent with abscess. Due to mass effect there is anterior displacement of the major fissure (curved red arrow on A). Pleural effusion (white arrows on C). The obstructing tumor is not identifiable.

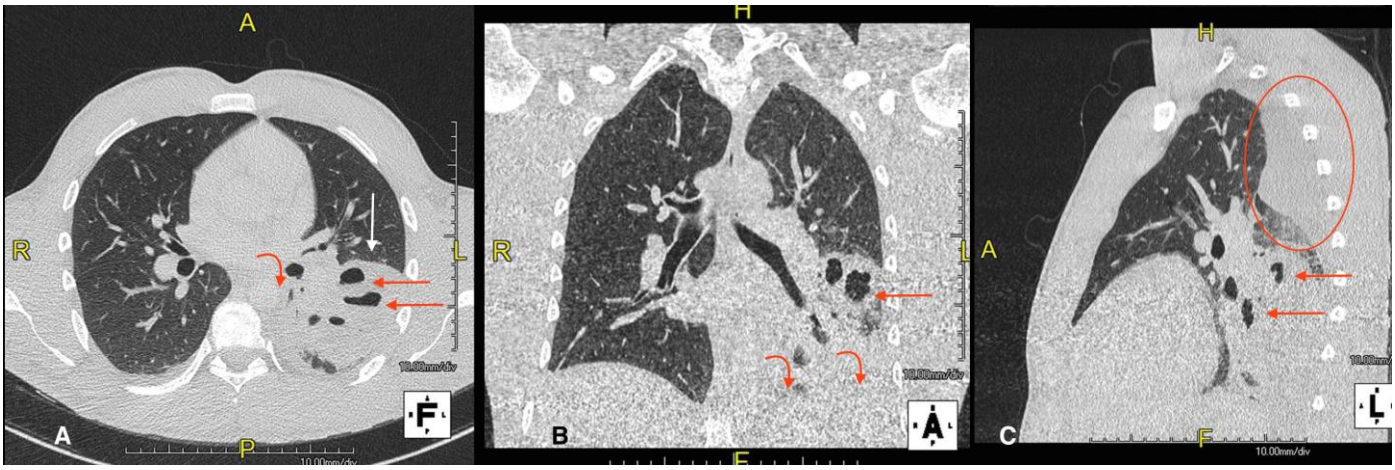

Figure 4: 23 year old male with post obstruction pneumonia and pulmonary abscess due to mucoepidermoid carcinoma of the airways.
Technique: chest CT scan after intravenous administration of 70 ml Xenetix© 300 (Siemens. 100kV; 188 mAs; 0,6 mm; 352 mGycm). Lung parenchyma window.
Findings: Consolidation in the left lower lobe adjacent to the left hemi-diaphragm and the descending thoracic aorta (curved red arrows on A and B). Cavitation with gas-fluid levels (large red arrows). Due to mass effect there is anterior displacement of the major fissure (white arrow on A). Pleural effusion (red oval on C). The obstructing tumor is not identifiable.

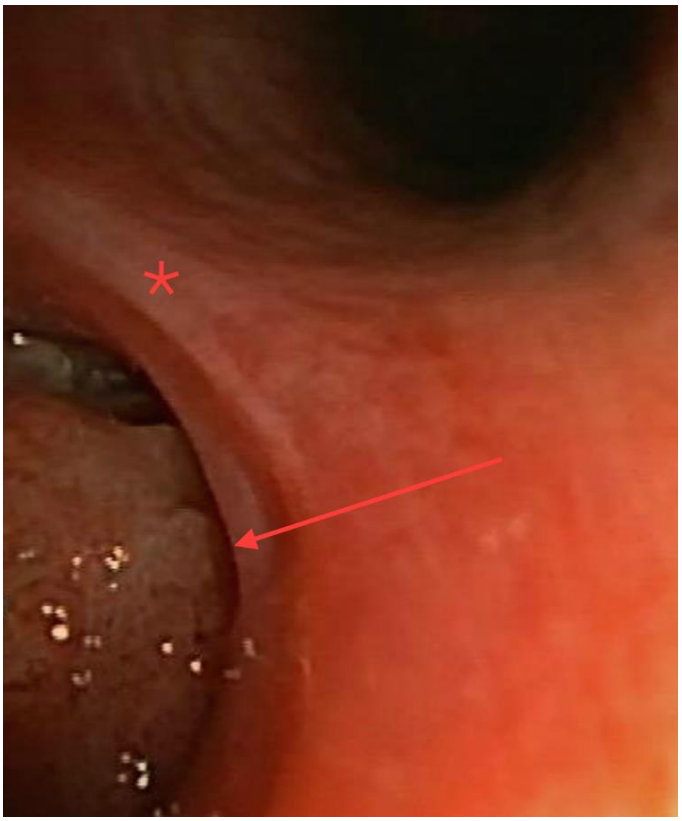

Figure 5: 23 year old male with post obstruction pneumonia and pulmonary abscess due to mucoepidermoid carcinoma of the airways.
Technique: standard bronchoscopy image.
Findings: A mobile endobronchial pedunculated tumor is visible in one of the bronchi to the apex of the left lower lobe (red arrow). The tumor is positioned proximally in the bronchus at the level of a carina (red asterisk).

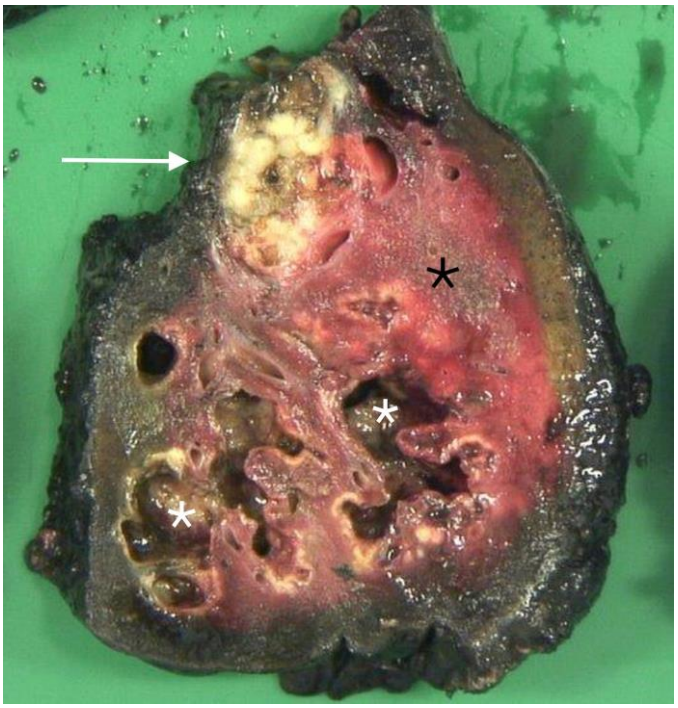

Figure 7: Macroscopic pathology. 23 year old male with mucoepidermoid carcinoma of the airways.
Technique: standard marcoscopy image after lobectomy.
Findings: Section of the mucoepidermoid carcinoma (white arrow) with underlying cavitation (white asterisks) of the lung parenchyma and inflamed lung tissue (black asterisks).
With courtesy of dr. Maria Tebar.

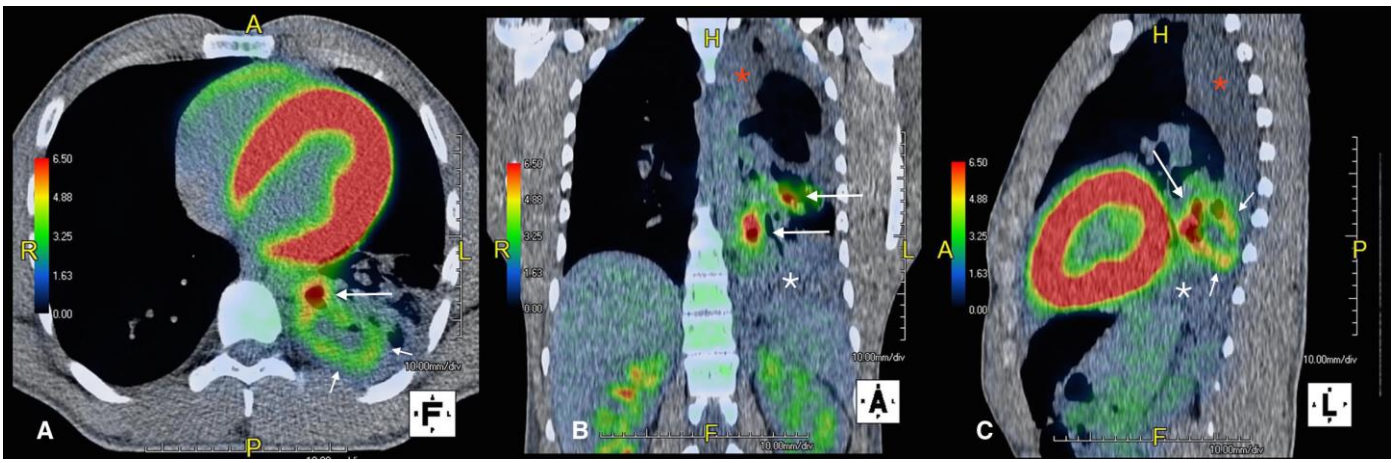

Figure 6: 23 year old male with post obstruction pneumonia and pulmonary abscess due to mucoepidermoid carcinoma of the airways.
Technique: Siemens. PET CT scan attained 1 hour after intravenous administration of 348 mBq (6,5 mGy) F18-FDG. Low dose CT scan, 3 mGy.
Findings: Consolidation in the left lower lobe with heterogeneous FDG uptake. The standardized uptake value maximum (SUVmax) is 8,5. There are multiple focal hot spots (large white arrows) but also areas were there is no measurable FGD uptake (white asterisk on B and C). Circular areas of medium FDG uptake possibly corresponding with the enhancement on the CECT and representing abscess walls (small white arrows on A and B). The obstructing tumor is not identifiable. Pleural effusion without measurable FDG uptake dorsal and paravertebral (red asterisk on B and C).

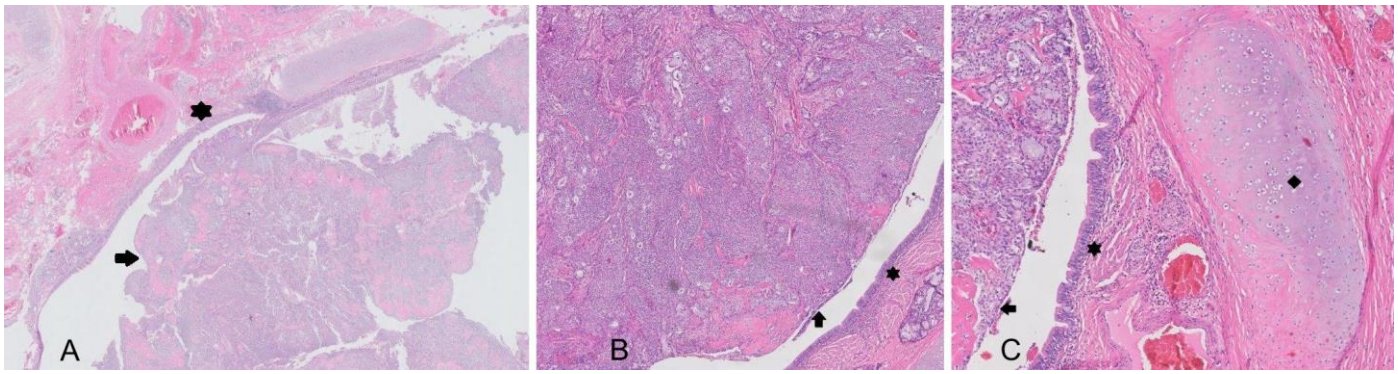

Figure 8: Microscopic pathology. 23 year old male with mucoepidermoid carcinoma of the airways.
Technique: Standard Hemotoxylin and Eosin stain. Magnification of 1 x, 5 x and 10 x on image A, B and C respectively.
Findings: Endobronchial mucoepidermoid carcinoma (black arrow) and bronchial epithelium (black asterisk) on image A, B and C. Bronchial cartilage (black diamond) on image C. Lymphangio-invasion of tumor was present but not visible here.
With courtesy of dr. Maria Tebar.

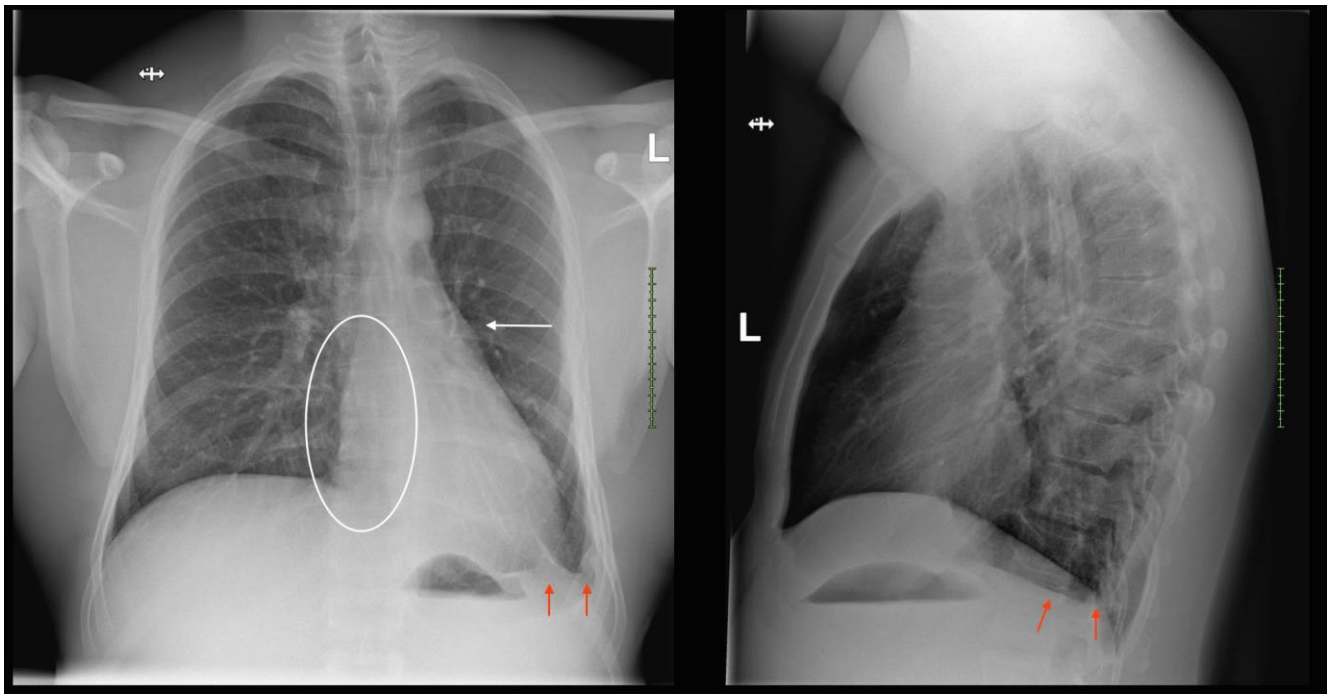

Figure 9: 23 year old male post lobectomy of the left lower lobe because of mucoepidermoid carcinoma of the airways.
Technique: plain PA and lateral radiograph (Siemens. 0,2 mm CU filter. PA:125 kV; 2,3 mAs. Lateral: 125 kV; 14,2 mAs).
Findings: Due to lobectomy of the left lower lobe there is caudal displacement of the left hilum (white arrow) and mediastinal shift to the left. This causes projection of the right atrium directly over the spine (white oval) where it used to project over the right lung. There are some adhesive pleural effects at the left hemi diaphragm and pleural sinus (small red arrows). The pleural effusion has resorbed completely and there are no metastases.

Etiology	Salivary gland-type tumor arising from airways. Characterized by a combination of mucus-secreting, squamous, and intermediate cell types.
Incidence	0,1%-0,2% of primary lung tumors
Gender ratio	Male = female
Age predilection	50% of patients is 30 years of age or younger
Risk factors	No association with cigarette smoking No other risk factors
Treatment	Surgical resection. There is insufficient evidence for chemo /-radiation therapy
Prognosis	Grade dependent. Distant or lymph-node metastases reduce survival rate
Findings on imaging	Chest x-rays: mass, post obstructive pneumonia/atelectasis or pulmonary abscess CECT: endoluminal mass with enhancement characteristics dependent on grade.

Table 1: Summary table for mucoepidermoid carcinoma of the airways.

	X-ray	CT	FDG-PET
Mucoepidermoid carcinoma	• Post obstructive pneumonia/atelectasis • Nodule/mass	• Endobronchial lesion • Enhancement dependent on tumor grade • Post obstructive pneumonia/atelectasis/ pulmonary abscess • Calcifications • Lymph-node or distant metastases	• Uptake dependent on grade • Metastases
(non) Small cell lung cancer	• Nodule/mass • Consolidation • Linear densities	• Nodule(s)/mass(es) • Consolidation • Lymph node metastases • Mediastinal/chest wall invasion • Lymphangitic carcinomatosis	• Uptake dependent on size and grade • Metastases
Carcinoid	• Nodule/mass • Post obstructive consolidation/ atelectasis/bronchiectasis	• Enhancing nodule/mass • (Partially) endobronchial/peribronchial/ perihilar • Post obstructive atelectasis/pneumonia/ abscess/bronchiectasis • Calcification (30%)	• Usually negative
Endobronchial hamartoma	• Post obstructive consolidation/atelectasis/ bronchiectasis • Endobronchial nodule	• Endobronchial nodule in central airway • Fat/calcification • Post obstructive consolidation/atelectasis/ bronchiectasis	• Minimal to no uptake
Adenoid cystic carcinoma	• Nodule/mass in trachea/bronchus • Stenosis of trachea or bronchus • Post obstructive consolidation/ atelectasis/bronchiectasis	• Pedunculated/broad based nodule/mass • Circumferential mass surrounding trachea or main bronchus • Variable margins • Calcification rare • Post obstructive • Consolidation/atelectasis/bronchiectasis • Lymph node metastases	• Uptake dependent on grade • Lymph-node metastases

Table 2: Differential diagnosis table for mucoepidermoid carcinoma of the airways.

ABBREVIATIONS

CECT = contrast enhanced computed tomography
MEC = mucoepidermoid carcinoma

KEYWORDS

Mucoepidermoid carcinoma; MEC; primary salivary gland-type lung cancer; Endobronchial mass; lung tumor