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Case Report

Primary High-Grade Mucoepidermoid Carcinoma Lung Mimicking Endobronchial Papilloma: A Case Report and Review of Literature

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ABSTRACT

Primary mucoepidermoid carcinomas (MECs) of the lung are rare and present a diagnostic challenge. MEC lung comes under the broad classification of primary salivary gland type tumours of the lung. It usually involves large airways and presents clinically and radiologically with non-specific features. We report the case of a 26-year-old female patient wherein an initial clinicoradiological consideration of tuberculosis and biopsy report of endobronchial papilloma led to misdiagnosis and treatment delay. Flexible bronchoscopic biopsy confirmed the diagnosis of MEC following which patient underwent curative pulmonary sleeve resection with good post-operative outcome. Final histopathology revealed high-grade MEC, stage as pT2N0, and thus, patient is receiving cisplatin-based adjuvant chemotherapy. More studies and reporting of such cases are needed to formulate guidelines and avoid delay in management.

Keywords: Mucoepidermoid carcinoma, Endobronchial papilloma, Salivary gland tumour lung, Pulmonary sleeve resection

INTRODUCTION

Mucoepidermoid carcinoma (MEC) of the lung is a rare tumour of bronchial origin accounting for 0.1–0.2% of all pulmonary neoplasms.[1] In the lung, the tumour arises from submucosal glands of the tracheobronchial tree and proliferates in a polyp-like form in the central bronchial lumen up to the segmental bronchus level. It is slow-growing, affecting young individuals and usually presents with symptoms related to bronchial obstruction including cough, haemoptysis, wheezing and signs of post obstructive pneumonia.[2] The non-specific clinical and radiological findings of this tumour can lead to a diagnostic dilemma.[3] In tuberculosis (TB) endemic countries, such a clinicoradiological profile may lead to misdiagnosis and diagnostic delay. We present a case report of 26-year-old female in whom MEC right lung was initially misdiagnosed as endobronchial papilloma resulting in delay in diagnosis and management.

CASE REPORT

A 26-year-old female with no significant medical history presented with cough with mucoid expectoration and breathlessness (Modified Medical Research Council Grade II) of 4 months

duration in pulmonary medicine department. The patient denied history of any fever or weight loss. She is a homemaker, with no history of smoking and no significant history of any cancer or cancer-related deaths in the family.

On evaluation outside this centre at the onset, she was noted to have monophonic wheeze over right hemithorax with right upper zone collapse and consolidation on chest X-ray posteroanterior view [Figure 1a]. Contrast-enhanced computed tomography (CT) chest documented partial cutoff of the right main bronchus (RMB) with segmental collapse and consolidation in the right upper lobe (RUL) with mediastinal and right hilar lymphadenopathy causing invasion into the RMB. CT findings suggested an active infective aetiology with TB as differential diagnosis [Figures 1b and c]. Work-up for TB including sputum Ziehl–Neelsen stain and GeneXpert was negative. Bronchoscopy showed a polypoidal mass in RMB [Figure 2a] which could not be navigated past and bled on touch. 18-Fluoro-deoxy Glucose (18-FDG) positron emission tomography–computed tomography (PET/CT) was suggestive of localised disease with FDG avid uptake in RUL. Endobronchial biopsy showed evidence of mixed glandular and squamous papilloma with thyroid transcription factor-1 positive in columnar cells, p40 positive in squamous cells, synaptophysin negative and Ki67 of <5%. Review of the biopsy also revealed the same findings.

In view of near total occlusion of RMB, the patient initially underwent therapeutic endobronchial cauterisation and cryofulguration and extraction of RUL papilloma under general anaesthesia. Near-total resection of the lesion with complete opening of the middle and lower lobe bronchus and partial opening of the upper lobe bronchus was achieved.

The patient was symptomatically relieved and was kept on follow-up with reassessment after 4 weeks.

Mild dyspnoea and cough persisted. However, over time the severity increased. Reassessment CT and check bronchoscopy were suggestive of recurrence [Figure 2b] with collapse consolidation of RUL with volume loss and compensatory hyperinflation of the remaining lobes.

Due to aggressive nature of the disease and early recurrence, the patient was referred to surgical oncology for re-evaluation. Endobronchial biopsy at this centre revealed salivary gland tumour of the lung favouring adenoid cystic carcinoma. Review of the blocks and slides at All India Institute of Medical Sciences, New Delhi was suggestive of low-grade MEC lung. Repeat imaging with CT chest and 18-FDG PET/CT showed an ill-defined soft-tissue density lesion of size 2.5 × 2.2 × 1.9 cm (Standardised Uptake Value max-5.7) in right supra-hilar region. The lesion was encasing and infiltrating the right main and upper lobe bronchus causing its abrupt cutoff. Bronchoscopy showed endobronchial growth involving RMB starting 0.5–0.7 cm below the carina with complete obliteration of RUL bronchus. Right middle lobe and right lower lobe bronchus appeared normal.

Therefore, with a working diagnosis of MEC lung, the patient was planned for the right pulmonary sleeve resection after proper pulmonary prehabilitation and optimisation. She was counselled regarding the possibility of right pneumonectomy. Intraoperatively, there was a 2 × 2 cm nodal mass near the RMB with an endobronchial growth in the RMB and RUL bronchus [Figures 3a and b]. A RUL right upper lobectomy with excision of the endobronchial lesion with adequate margins including a small cuff of RMB and regional lymphadenectomy through a right posterolateral thoracotomy was achieved and bronchus intermedius was anastomosed to RMB reinforced with a parietal pleural flap [Figure 3c].

Gross examination of the specimen showed a right upper lobectomy specimen with 2 × 2 cm growth in the RMB which was extending into RUL bronchus with proximal and distal cut margins free of tumour [Figure 3d].

Histopathological examination revealed both solid and cystic areas within the tumour. The solid areas showed cells

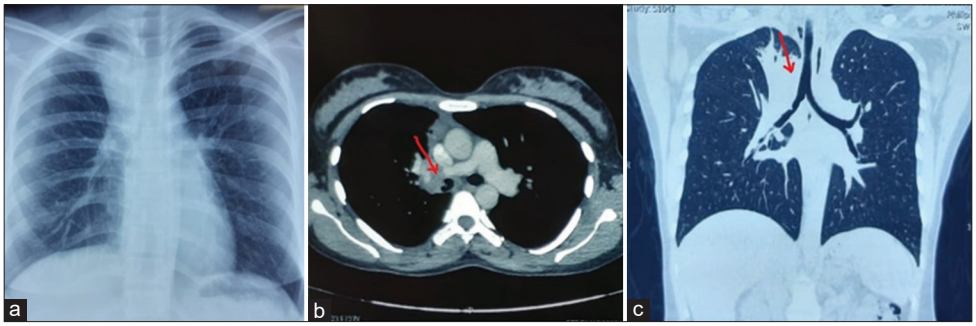

Figure 1: Pre-operative images. (a) Chest X-ray posteroanterior view showing right upper zone collapse and consolidation. (b and c) Contrast enhanced computed tomography chest axial and coronal sections showing partial cutoff of right main bronchus (red arrow) with segmental collapse and consolidation in the right upper lobe with mediastinal, right hilar lymphadenopathy and compensatory hyperinflation of the right middle lobe.

arranged in nests comprising of three type of cells – mucin secreting cells, intermediate cells and sheets of squamoid cells with columnar epithelium lined cystic areas. Final report was mucoepidermoid ca lung, Brandwein high grade with no lymphovascular invasion/perineural invasion (LVI/PNI) with 12 lymph nodes free of tumour and all margins negative, staged as pT2N0 [Figures 4a and b].

Post-operative period was complicated by pneumothorax on post-operative day (POD)#6 after removal of intercostal drainage tube [Figure 4c]. However, the same was resolved with conservative management and patient was discharged in stable condition on POD#9. On reassessment at 1 month postoperatively, the patient was asymptomatic with no fresh complaints.

Lung cancer 12 gene panel with programmed cell death ligand-1 (PD-L1) – VENTANA SP142 was performed to guide the adjuvant treatment decision for MEC following surgery. No gene mutation, gene fusion and copy number variation in MET gene was detected. PD-L1 expression was positive in 0% tumour cells and <1% tumour infiltrating immune cells. In view of high grade, adjuvant cisplatin-based chemotherapy was planned for the patient.

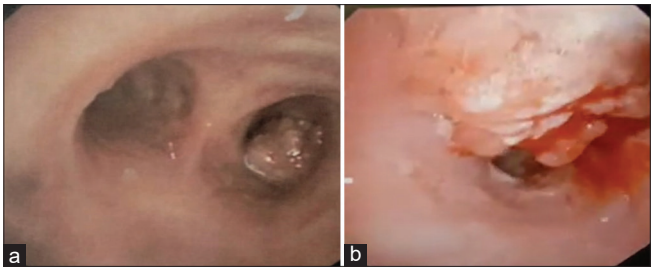

Figure 2: Pre-operative bronchoscopy images. (a) Pre-cryofulguration bronchoscopy showing endobronchial growth involving right main bronchus with complete obliteration of the right upper lobe (RUL) bronchus. (b) Post-cryofulguration images showing recurrence of tumour in RUL bronchus with near total occlusion.

DISCUSSION

The World Health Organisation classifies pulmonary MECs as 'salivary gland type' tumours along with pulmonary adenoid cystic carcinomas and epimyoepithelial lung carcinomas.[4] Histologically, MEC is comprised of a mixture of different cell types including mucin-secreting glandular cells, squamous cells and intermediate cells. Low-grade MEC is distinguished from high-grade MEC based on the lack of cytological atypia including nuclear pleomorphism and absence of significant mitotic activity and cellular necrosis. Histological grade is an important prognostic indicator (Brandwein grading), with high-grade MECs demonstrating a greater risk for metastases, tumour recurrence and death.[5]

PubMed and Google Scholar search using following terms: 'primary pulmonary mucoepidermoid carcinoma' OR 'primary lung mucoepidermoid carcinoma' OR 'primary mucoepidermoid carcinoma of lung' OR 'primary mucoepidermoid tumour of lung' OR 'primary pulmonary low-grade mucoepidermoid carcinoma' OR 'pulmonary mucoepidermoid carcinoma' OR 'primary bronchial mucoepidermoid carcinoma' OR 'bronchial mucoepidermoid carcinoma' OR 'primary mucoepidermoid carcinoma of bronchus' OR 'primary mucoepidermoid carcinoma of tracheobronchial tree' yielded 81 results. Most of them are case reports or case series with only 600 cases reported till date.[5,6]

Surgical resection remains the standard therapy for patients with pulmonary MEC. The goal of surgery is to obtain a complete resection with negative surgical margins. Our

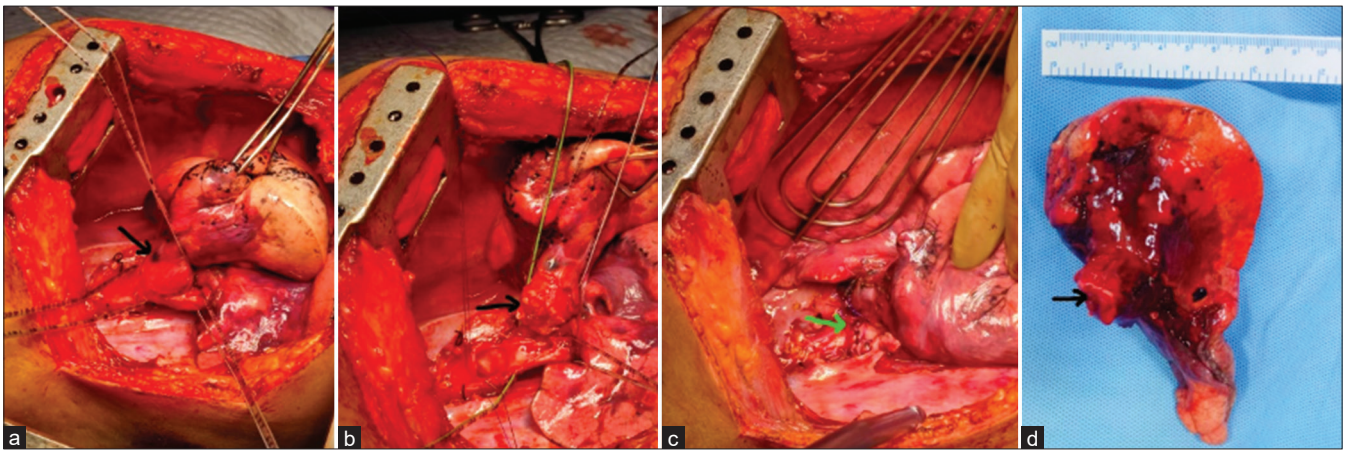

Figure 3: Intraoperative images. (a and b) 2 × 2 cm nodal mass near the right main bronchus (RMB) with an endobronchial growth (black arrow) in the RMB and right upper lobe Bronchus. (c) Bronchus intermedius was anastomosed to right main bronchus (green arrow) and reinforced with a parietal pleural flap. (d) Gross specimen showing right upper lobe with an endobronchial growth (black arrow) causing complete luminal obstruction.

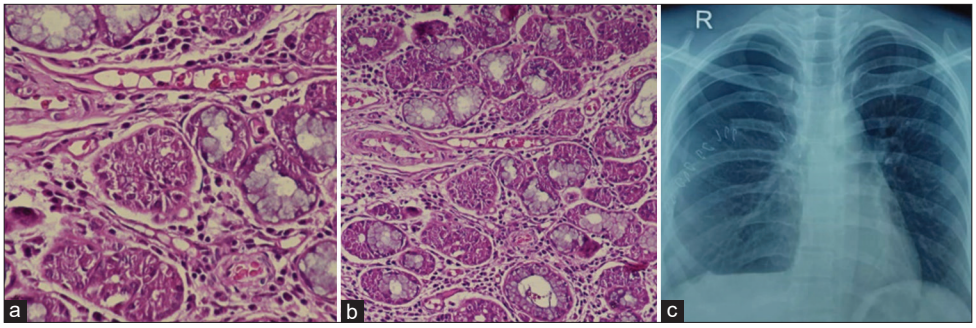

Figure 4: Post-operative histopathology images. (a and b) Haematoxylin and eosin stain, ×40 and ×20 views revealed nests of three types of cells in solid areas of the tumour-mucin secreting cells, intermediate cells and sheets of squamoid cells with columnar epithelium lined cystic areas with nuclear atypia favouring mucoepidermoid ca lung. (c) Post-operative day #7 follow-up chest X-ray – showing inflated right lung with pneumothorax with no tracheal deviation or costophrenic angle blunting.

patient had typical presentation for a low-grade MEC, a single centrally located well-circumscribed endobronchial tumour without evidence of locoregional or distant metastasis with 4 months history though the histopathological examination revealed differently. Patients with low-grade MEC generally have a good prognosis, with a 5-year survival rate of 95%, and adjuvant treatment is considered unnecessary. However, for high-grade tumours, adjuvant treatment in form of chemotherapy or radiotherapy is indicated, though its usefulness remains controversial and these patients have a poor prognosis. 12-gene lung cancer panel can help identify patients who are at high risk of recurrence and may benefit from adjuvant chemotherapy. PD-L1 assessment can help identify patients who may benefit from immune checkpoint inhibitors.

CONCLUSION

While there have been numerous case reports and case series on MEC, the presented case report adds to the limited literature on the diagnostic challenges and management of MEC in TB-endemic countries and emphasises the importance of histological grading in predicting prognosis. It also highlights the need for a multidisciplinary approach to patient management and the potential utility of a 12-gene lung cancer panel and PD-L1 assessment in identifying patients at high risk of recurrence and those who may benefit from adjuvant therapy.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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