Case Report

# Curious case of primary pulmonary mucoepidermoid carcinoma

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## Abstract

Pulmonary mucoepidermoid carcinoma (MEC) is an uncommon tumor constituting only 0.1% to 0.2% of all lung carcinoma. It is classified under "salivary gland type" tumors in the World Health Organization (WHO) classification of lung cancers. It generally carries a better prognosis than the more common small cell and nonsmall cell lung carcinomas. It is pathologically classified into high-grade and low-grade tumors. High-grade tumors are usually unresectable at presentation. However, surgery is the mainstay of treatment, which aims at surgically negative margins for a complete cure. In our patient, pulmonary MEC presented with intrathoracic mass with pericardial effusion. It came out to be low-grade neoplasm, which was treated with platinum-based doublet chemotherapy and responded well with near-total disappearance of tumor, like a vanishing lung tumor.

**Keywords:** CECT thorax, chemotherapy, intrathoracic mass, mucoepidermoid carcinoma

## Introduction

Pulmonary mucoepidermoid carcinoma (MEC) are rare primary neoplasms of the lung with a varied response to chemotherapy. Although surgery is the mainstay of therapy in localized disease, for patients who suffer from the advanced disease without any scope of surgical resection, chemotherapy is the only option. Chemotherapy has got a variable response, depending on the grade of the tumor and stage of the disease, so it is imperative to start the patient with appropriate chemotherapy.

## Case History

A 32-year-old nonsmoker male was admitted to the emergency department with complaints of breathlessness, chest pain, and pedal edema for 15 days with breathlessness, which was insidious in onset, gradually progressive, aggravated on exertion, and relieved on taking rest. Chest pain was a dull ache, nonradiating, and localized to the left side. On clinical examination, the jugular venous pressure was raised, heart sounds were muffled, and breath sounds were decreased over the left infrascapular region. There was no significant previous medical, social, and family history.

Initial routine blood investigations were within normal limits. A chest X-ray revealed a heterogeneously enhancing mass lesion with pleural effusion and cardiomegaly on the left hemithorax [Figure 1]. An electrocardiogram (ECG) showed low voltage complexes. Two-dimensional echocardiography showed mild circumferential pericardial effusion without evidence of tamponade physiology. A contrast-enhanced computed tomography (CECT) scan of the thorax revealed a heterogeneously enhancing intrathoracic mass infiltrating both pericardium and chest wall with bilateral pleural effusion [Figure 2]. A CT-guided

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transthoracic biopsy of the mass was done and was subjected to histopathological examination (HPE). The HPE revealed nests of epidermoid and intermediate cells with focal acinar structures lined by mucin-filled columnar cells [Figure 3]. Immunohistochemistry (IHC) showed p-63 positivity and TTF-1 negativity, consistent with a diagnosis of MEC.

Although initial differential diagnosis included only bronchogenic carcinoma and lymphoma, the biopsy confirmed the diagnosis of MEC.

After diagnosis, the staging of the tumor was done in accordance with the American Joint Committee on Cancer (AJCC) TNM system, 7th edition. The patient's designated stage was Stage IV-T4N3M1a. After a normal prechemotherapy evaluation, platinum-based doublet chemotherapy consisting of gemcitabine and cisplatin was initiated. After four cycles of chemotherapy, the chest X-ray and CECT thorax revealed near-total disappearance of primary tumor and resolution of pleural and pericardial effusion [Figure 4]. Response evaluation done with the response evaluation criteria in solid tumor (RECIST) yardstick revealed that he had a partial response.

## Discussion

Primary pulmonary MEC, though uncommon, deserves special attention as it has a better prognosis than other conventional lung carcinoma types.[1] Surgery is the mainstay of treatment in resectable cases, which are usually low-grade histological MEC. In a previous study by Hsieh *et al.*, surgical outcomes of surgery in MEC have been discussed and the outcome is dependent on age, tumor size, and pathological factors.[2] In surgically untreatable cases, chemotherapy is the only option for management. There is no universally accepted chemotherapy regimen for MEC and an individualized approved is advocated. For high-grade MEC, carboplatin and paclitaxel were used with a good response.[3] Docetaxel was studied by Hui-Ching Wang *et al.*, which was administered biweekly.[4] The patient survived for 6 months after which the disease progressed. In our case, the patient had tremendous improvement with combination chemotherapy and near-total disappearance of the primary lesion and symptom-free up to 7 months of chemotherapy (including 3 months of maintenance chemotherapy with gemcitabine). In some reports of MEC, epidermal growth factor receptor (EGFR) mutations were studied and tailored chemotherapy with tyrosine kinase inhibitor (TKI) was given, but the response was equivocal.[5] Some studies have highlighted a good response of the wild-type EGFR mutations to gefitinib.[6]

To conclude, low-grade intrathoracic MEC has a better prognosis, and a high index of suspicion is required for diagnosis.[7] Variable responses to chemotherapy advocates

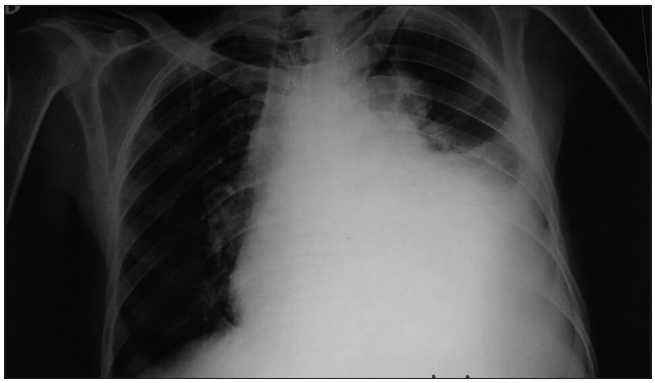

**Figure 1:** Chest X-ray posteroanterior view showing a heterogeneously enhancing mass lesion in the left lower zone, pleural effusion, and cardiomegaly

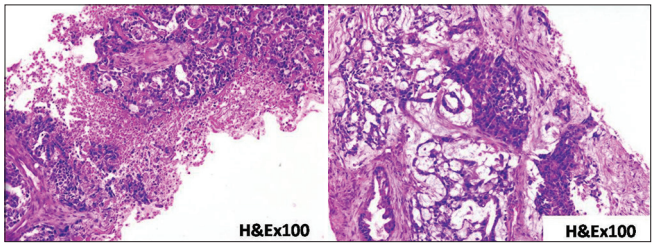


**Figure 3:** Histopathological examination revealed nests of epidermoid and intermediate cells with focal acinar structures lined by mucin-filled columnar cells (HXE100)

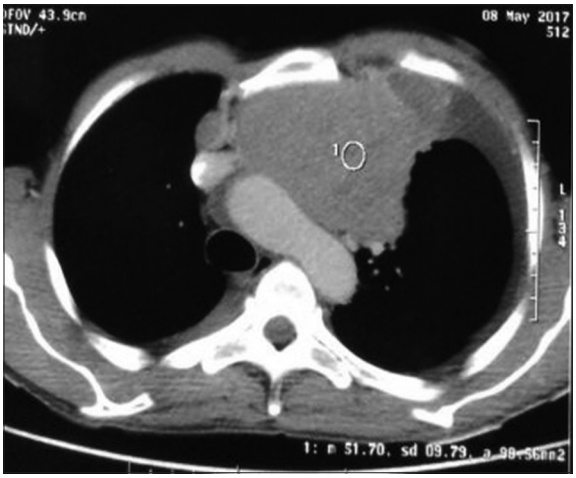


**Figure 2:** Contrast-enhanced computed tomography scan of thorax revealed a heterogeneously enhancing intrathoracic mass with bilateral mild pleural effusion

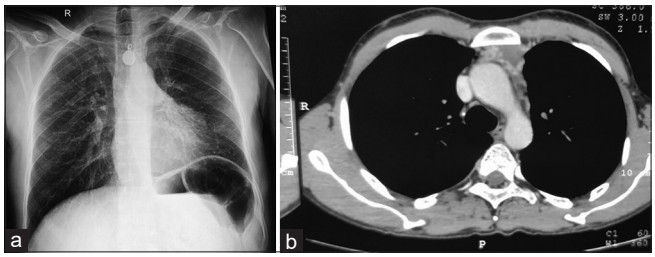


**Figure 4:** (a) Chest X-ray revealed after four cycles of chemotherapy a near total disappearance of primary tumor and resolution of pleural and pericardial effusion (b) Contrast-enhanced computed tomography scan thorax revealed after four cycles of chemotherapy near-total disappearance of primary tumor and resolution of pleural and pericardial effusion

careful selection of drugs and vigilant follow-up. Surgical resection is the option in resectable tumors while chemotherapy with platinum-based chemotherapy remains the next best option.[8]

## Implications for general practice

Lung cancer is a disease that is prevalent in our country. Primary pulmonary MEC represents a rare type of lung cancer. The clinical symptoms are similar to that of asthma, COPD or pneumonia. Hence one should keep in mind of this rare entity in patients who are unresponsive to regular treatment. Low-grade MEC has a much better prognosis than high-grade tumour, the latter being similar to non-small-cell lung carcinoma. Hence, physicians in primary care need to rely on HPE and IHC findings when lung cancer is suspected rather than history or radiological findings.

## 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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Nil.

## Conflicts of interest

There are no conflicts of interest.

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