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## Mucinous cyst adenocarcinoma of lung presented with recurrent pneumothorax

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Sir,

Mucinous cystadenocarcinoma of the lung (MCACL) is a very rare malignant neoplasm with unique morphological and clinical behavior. It is a tumor of non-smokers and in the early stage it remains silent and is incidentally diagnosed by chest imaging. MCACL produces copious mucin resembling tumors of the same name in the ovary, breast, and pancreas.<sup>[1]</sup> Patients usually present with symptoms due to airway irritation and obstruction. MCACL has a much more favorable prognosis than most other forms of adenocarcinoma and most other non-small cell lung carcinomas (NSCLCs).<sup>[2]</sup> Our patient presented with dyspnea, which was later diagnosed as mucinous cyst adenocarcinoma of the lung on Tru-cut lung biopsy.

A 37-year-old male non-smoker, with no known comorbidities, presented with productive cough and

progressively increasing shortness of breath since four months. The patient was initially admitted to a nearby hospital, where the patient was found to have a left-sided pneumothorax, which was managed by Intercostal tube drainage and then referred to us.

On examination he was found to be of average build, with bilateral crepitations on chest auscultation and subcutaneous emphysema, with an inter-costal drain (ICD) tube *in-situ* on the left side of the chest.

His routine investigations revealed normal complete blood count (CBC), Kidney function tests (KFTs), and liver function tests (LFTs). The patient's ICD was removed when the lung expanded completely and there was no air leak. The chest X-ray showed bilateral dense consolidation and left-sided subcutaneous emphysema. High-resolution computed tomography (HRCT) of the

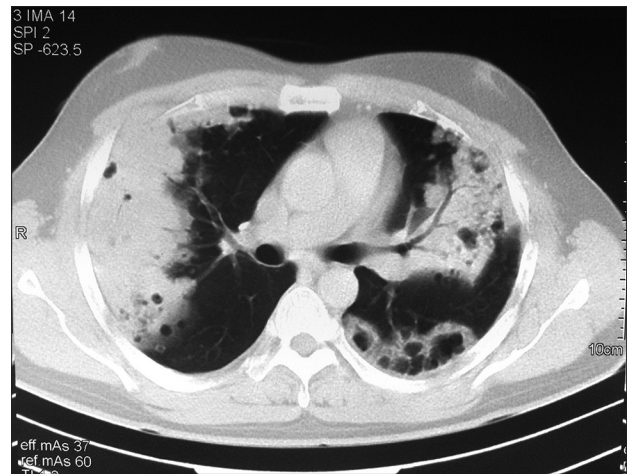
patient's chest [Figure 1] was done, which showed bilateral areas of dense consolidation and cavitating or cystic lesions in between the consolidation.

A bronchoscopy with lavage and transbronchial lung biopsy was done, which did not reveal any diagnosis. Subsequently video-assisted thoracoscopic surgery (VATS) was performed for a lung biopsy. Histopathology of the biopsy specimen confirmed the diagnosis of mucinous cyst adenocarcinoma of the lung [Figure 2]. The patient was referred to the Oncology department where platinum-based chemotherapy was started after complete evaluation and a positron emission tomography (PET) scan (no distant metastasis) was performed. The patient was discharged in stable condition, but after a few weeks he again reported to us with increased shortness of breath. An urgent chest x-ray (CXR) was done, which showed left-sided subcutaneous emphysema and bilateral consolidation. A CT-scan was done, which revealed a left-sided pneumothorax [Figure 3]. The pneumothorax was managed by intercostal tube drainage and talc pleurodesis. The ICD was removed and the patient was discharged in a stable condition.

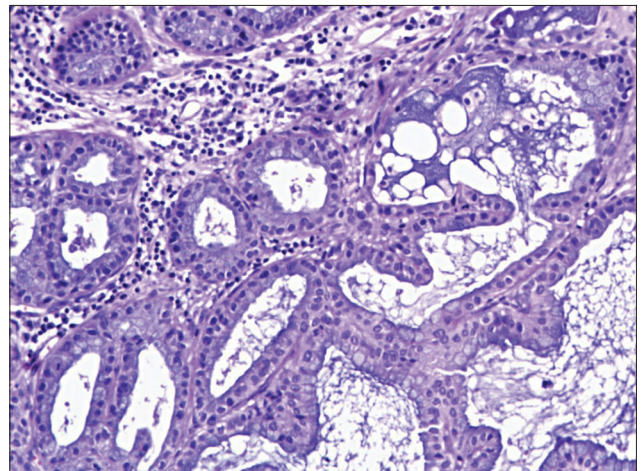
A pulmonary mucinous cyst adenocarcinoma is an extremely rare subtype of pulmonary adenocarcinoma that was first described by Gowar, in 1978.<sup>[2]</sup> According to the World Health Organization (WHO) classification, pulmonary mucinous cyst adenocarcinoma is a distinct variant of adenocarcinoma of the lung.<sup>[3]</sup> However, the 2010 International Association for the Study of Lung Cancer or the American Thoracic Classification System for lung tumors includes MCACL as part of the colloid adenocarcinomas.<sup>[3]</sup> Histologically they are well-circumscribed neoplasms, with a fibrous tissue capsule and central cystic changes with a mucin feeling, due to uncontrolled mucus production by neoplastic transformation of the mucin producing alveolar epithelial cells. The majority of MCACL patients remains asymptomatic and present late or are incidentally diagnosed. Some patients present with symptoms due to airway irritation or obstruction, such as, cough, dyspnea, wheezing, and chest pain, and non-specific manifestations, such as, pneumothorax, recurrent bronchitis, hemoptysis, weight loss, and fever.<sup>[3]</sup>

The differential diagnosis of MCACL includes mucous gland adenoma, mucoepidermoid carcinoma, mucinous bronchioalveolar carcinoma, metastatic carcinoma, and mucinous-looking non-neoplasms (mucocele and bronchogenic cyst), as well as lymphangiomas.<sup>[4]</sup> Pleural mesothelioma can also be included as differential of peripherally located pulmonary mucinous cyst adenocarcinoma.<sup>[5]</sup>

The chest X-ray findings are non-specific in case of MCACL, which include dense lobar consolidation, ill-defined nodules or rarely cystic lesions. The typical CT-scan features



**Figure 1:** HRCT-chest showing bilateral areas of dense consolidation and cavitating or cystic lesions



**Figure 2:** Tumor glands under high magnification with few mucin-filled columnar cells lining the gland, with basally located hyperchromatic nuclei with mild variation in the cytoplasm



**Figure 3:** CT-chest showing left-sided pneumothorax with bilateral consolidation

include well-defined, partly-lobulated, homogeneous, and persistently-low attenuation lesions, with focal, mural, or septal enhancement.<sup>[2]</sup> Sometimes unilocular cystic or low-attenuation lesions are also observed.

Positron Emission Tomography scanning can be of assistance in diagnosing MCACL. Fluoro-deoxy-glucose (FDG) PET might show little FDG uptake, because of low tumor cellularity and abundant mucin.

For treatment purposes, MCACL has been traditionally considered as a non-small cell lung carcinoma (NSCLC). Complete radical surgical resection is the treatment of choice. Other treatment options are radiotherapy, chemotherapy, and immunotherapy.

MCACL has a much more favorable prognosis than most other forms of adenocarcinoma and most other NSCLCs. Cases have been documented of continued growth of these lesions over a period of 10 years without symptoms or metastasis.

Our case was presented with cough and dyspnea, pneumothorax was found on left side which was managed by ICD tube but conclusive diagnosis was still uncertain, so VATS guided lung biopsy was done which showed MCACL. We assume that rupture of peripherally or subpleurally located cysts lead to recurrent pneumothorax in our case. This was our first experience of MCACL and we searched the literature thoroughly which revealed that it is a very rare tumor, with only a few dozen cases reported in the literature to date.

Mucinous cystadenocarcinoma of the lung is a very rarely reported malignancy of the lung and our case is also among the few reported cases. Our case of MCACL was unique because it developed recurrent pneumothorax during the clinical course. There was no previously reported case of MCACL with pneumothorax. We assumed that pneumothorax developed due to necrosis or rupture of the peripheral cysts. Therefore, if a patient with bilateral areas of dense consolidation or cavitation develops pneumothorax, MCACL should be a differential diagnosis.

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#### Conflicts of interest

There are no conflicts of interest.

#### REFERENCES

1. Gao ZH, Urbanski SJ. The spectrum of pulmonary mucinous cystic neoplasia: A clinicopathologic and immunohistochemical study of ten cases and review of literature. *Am J ClinPathol* 2005;124:62-70.
2. Chhieng DC. Fine-needle aspiration cytology of pulmonary mucinous cystadenocarcinoma. *Diagn Cytopathol* 2008;36:581-5.
3. Beasley MB, Brambilla E, Travis WD. The 2004 world health organization classification of lung tumors. *Semin Roentgenol* 2005;40:90-7.
4. Efsthathiou A, Asteriou C, Barbetakis N, Miliaras D, Kleontas A, Karvelas C, et al. Primary pulmonary mucinous cystadenocarcinoma: A case report. *Case Rep Med* 2011;2011:562026.
5. Kassir R, Forest F, Kaczmarek D. Pulmonary mucinous cystadenocarcinoma presenting as a pleural mesothelioma. *Int J Surg Case Rep* 2013;4:942-4.

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