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Short Communication

# Glutinous and Perimetric-Mucinous Cystadenoma Lung

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Mucinous cystadenoma emerges as a benign, localized, cystic tumefaction permeated with mucin and enmeshed within a fibrous tissue wall. Neoplasm is layered with well differentiated, mucinous columnar epithelium and is commonly confined to pancreas, ovaries or appendix. Additionally, mucinous cystadenoma may incriminate hepatic parenchyma, mammary glands, ovaries, urinary bladder or retroperitoneal area. The benign mucinous cystadenoma exceptionally incriminates pulmonary parenchyma.

Initially scripted by Eck in 1969, mucinous cystadenoma manifests as a unilocular cyst permeated with mucus and encompassed within a fibrous capsule. Occasionally, multilocular cysts may be enunciated. The essentially benign neoplasm may demonstrate minimal malignant potential [1].

Notwithstanding, Sam Brooke Gowar in 1978 denominated 'primary pulmonary mucinous cystadenoma neoplasia' as an umbrella term which represents neoplasia such as mucinous cyst tumour, mucinous cyst, mucinous cyst tumour of low malignancy or mucinous cystadenocarcinoma [2].

Preoperative determination of infrequently discerned mucinous cystadenoma may be challenging, especially as the neoplasm is predominantly (90%) comprised of mucin. However, lesion may be appropriately discerned intraoperatively.

Antecedent and comprehensive surgical resection of pulmonary mucinous cystadenoma is recommended in order to circumvent possible malignant metamorphosis or exterminate borderline lesions.

Of obscure aetiology, neoplasm is posited to arise from non alveolar epithelium. The preponderantly cystic mucinous cystadenoma pervaded with gelatinous mucin emerges as a well defined lesion confined to peripheral pulmonary parenchyma [3,4].

KRAS genetic mutations may emerge as a precise mechanism for occurrence of mucinous cystadenocarcinoma engendered from pulmonary mucinous cystadenoma.

Median age of disease emergence is 61 years although pulmonary mucinous cystadenoma may appear between 32 years to 75 years. A mild female predilection is observed [3,4].

Frequently confined within peripheral pulmonary lobes, right lung is frequently implicated. Median tumour diameter appears at 5 centimetres although neoplasm ranges from 0.8 centimetres to 15 centimetres.

Mucinous cystadenoma is predominantly (~68%) asymptomatic. Nevertheless, cogent clinical symptoms as cough, haemoptysis or repetitive bouts of pneumonia may be exemplified.

Incriminated subjects may represent with haemoptysis or demonstrate asthenia. History of cigarette smoking may be elicited. Commonly, right lung or lower lobes are implicated [3,4].

Grossly, a solitary, well circumscribed, cystic lesion of variable magnitude may be encompassed with or appear devoid of a distinct fibrous tissue wall and is confined to pulmonary parenchyma. Adjoining bronchial mucosa appears devoid of evident foci of carcinoma in situ. Tumefaction is preponderantly (> 90%) composed of mucin. Cystic wall appears attenuated, transparent and smooth. Few dilated bronchi appear to circumscribe the neoplasm [5,6].

Upon microscopy, the preponderantly cystic tumefaction is permeated with mucous contents. Tumour wall is layered by mucus secreting, cylindrical or cuboidal epithelium accompanied by focal giant cell reaction. Cylindrical or cuboidal epithelial cells are pervaded with basophilic cytoplasm and hyperchromatic nuclei. Cellular or nuclear atypia is absent. Neoplastic epithelial cells configure

as mucin producing cells which appear disseminated within pools of mucin or layer the fibrous cystic wall [5,6].

Characteristically, the localized, cystic tumefaction represents with benign proliferation of mucus secreting epithelial cells.

Adjoining pulmonary tissue exhibits altered architecture and benign cellular proliferation. Alternatively, adjacent pulmonary parenchyma may represent with organizing pneumonia or dilated bronchi with thickened walls and layering ulcerated epithelium.

Incriminated, enlarged regional lymph nodes manifest with follicular hyperplasia, anthracosis or sinus histiocytosis [5,6].

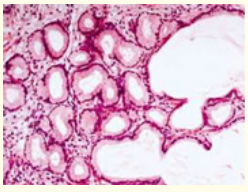

**Figure 1:** Mucinous cystadenoma demonstrating a cystic lesion layered by columnar, mucus secreting epithelial cells imbued with basophilic cytoplasm and hyperchromatic nuclei devoid of cellular and nuclear atypia. Surrounding pulmonary parenchyma is inflamed [8].

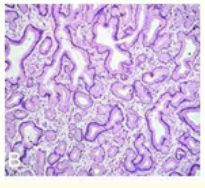

**Figure 2:** Mucinous cystadenoma delineating cystic lesions layered by columnar, mucus secreting epithelium incorporated with basophilic cytoplasm and hyperchromatic nuclei devoid of cellular and nuclear atypia. Circumscribing pulmonary parenchyma appears inflamed [9].

Mucinous cystadenoma appears intensely immune reactive to CK7, CK 20 or CDX-2 and minimally immune reactive to AE1/AE3. Tumour cells appear immune reactive to pan-cytokeratin (CK) or surfactant-associated protein A.

Tumefaction appears immune non reactive to CA125, carcinoembryonic antigen (CEA), thyroid transcription factor-1 (TTF-1), D2-40 or WT1. Multifocal primary pulmonary mucinous cystic neoplasia encompassed with ovarian subtype of stroma appear immune reactive to AE1/AE3, epithelial membrane antigen (EMA) and CK7. Tumefaction appears immune non reactive to CK19 or CK20.

Mucinous cystadenoma exhibits minimal expression of proliferating cell nuclear antigen (PCNA) or MIB1 gene [6,7].

Mucinous cystadenoma requires segregation from non neoplastic conditions such as congenital adenomatoid malformation, bronchogenic cyst or post-infectious bronchogenic cyst and neoplasms such as bronchial cystadenoma, mucinous cystadenoma, pulmonary mucinous tumour of low malignancy, pulmonary mucinous cystadenocarcinoma, mucoepidermoid carcinoma, mucinous bronchoalveolar carcinoma, diverse metastatic mucinous ovarian or pancreatic neoplasms or metastatic pulmonary cystic mucoid lesions [6,7].

Upon radiography, a well defined, homogeneous tumefaction is observed. Plain radiographs display macro-nodular opacities confined to various lobes of pulmonary parenchyma.

Computerized tomography (CT) exhibits a well circumscribed, homogeneous neoplasm. Cyst wall appears attenuated.

Inflammation and atelectasis of adjacent pulmonary parenchyma may ensue following enlargement of lesion along with compression and distortion of surrounding tissue [6,7].

| Subtype | Epidemiology | Immunohistochemistry | Molecular Alterations |
|---|---|---|---|
| PEAC | M>F, smokers (0.5%) | CK7, CDX-2, villin | KRASG12V, ERBB2, EGFR-del ex19, L8S8R |
| Foetal adenocarcinoma | Young female, smokers (0.5%) | Synaptophysin, vimentin | WNT signal |
| Colloid adenocarcinoma | F>M, smokers (0.1%) | CK7, CK20, CDX-2, MUC2 | KRAS codon 12/13 |
| NOS carcinoma | M>F(0.1%) | TTF-1, p40, none | EGFR, ALK-EML4 |

**Table 1:** Characteristics of Pulmonary Adenocarcinoma [5].

PEAC: Pulmonary enteric adenocarcinoma, NOS: Not otherwise specified, TTF-1: thyroid transcription factor-1.

Computerized tomography (CT) of thoracic cavity or superior abdomen along with administration of contrast exhibits a dense, homogenous tumefaction with an irregular perimeter. Regional lymph nodes as hilar, mediastinal or subcarinal nodes appear enlarged [6,7].

Positron emission tomography with 2-deoxy-2-(fluorine-18) fluoro- D-glucose (PET-CT 18F-FDG) may enunciate minimal and diffuse image capture of the pulmonary lesion whereas hilar and paratracheal lymph nodes delineate an intense image capture [6,7].

Pulmonary function tests depict minimally diminished forced expiratory volume (FEV) and forced vital capacity (FVC). Mild anaemia is observed. Mucinous cystadenoma can be appropriately subjected to antero-lateral thoracotomy. Surgical extermination of elastic, resistant tumefaction in the absence of eradication of visceral pleura appears as an optimal therapeutic strategy [6,7].

Additionally, manoeuvers such as pneumotomy or dissection of the encapsulated, gelatinous, cystic pulmonary nodule of variable magnitude may be adopted.

Intraoperative discernment of mucinous cystadenoma along with mediastinal assessment may demonstrate enlarged para-tracheal, hilar or para-oesophageal lymph nodes wherein procedures such as lobectomy may be combined with hilar and mediastinal lymphadenectomy.

The benign pulmonary mucinous cystadenoma may possibly demonstrate malignant potential wherein preliminary and comprehensive surgical resection with removal of tumour free surgical margin is recommended and optimal. Localized lesions may be optimally subjected to minimally invasive surgical excision procedures. Additional monitoring is contingent to cogent pathological features [6,7].

Prognostic outcomes are superior wherein neoplastic reappearance or distant metastases emerging from malignant lesions is exceptional. Tumour reoccurrence may ensue in up to 20 years following initial detection, especially within instances lacking cogent anatomical resection. Thus, extended monitoring is recommended [6,7].

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8. Image 1 Courtesy: Journal of Pathology and Translational medicine.

9. Image 2 Courtesy: Hindawi.com.