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CT and MRI findings of mucin-containing tumors and pseudotumors of the thorax: pictorial review

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Abstract Bronchial mucus has tomodensitometric features and MR signal intensity similar to that of water. However, chronic entrapped mucus collections, due to water reabsorption and higher protein content, can have CT attenuation values higher than 20 and reaching even 130 HU. Higher protein concentration also causes a sensible reduction in T1 relaxation time. The demonstration of mucus within a mediastinal, bronchial or pulmonary lesion is an important diagnostic clue permitting remarkable shortening of the list of differential diagnoses. This article illustrates the CT and MR findings allowing correct characterization of the mucus-containing lesions of mediastinum, bronchi, and lung.

 $\begin{array}{ll} \textbf{Keywords} & Lung \cdot Mucin \cdot MR \\ imaging \cdot CT \end{array}$

Introduction

Mucinous tumors and pseudo-tumors of mediastinum, bronchial tree, and lung are a heterogeneous group of space-occupying lesions characterized by high mucin content. Although their origin is variable, all these lesions are constituted exclusively or chiefly by mucus.

The normal bronchial mucus is 95–98% water and 2–5% solids, and the majority of the solid component (60%) is formed by glycoproteins [1]. Due to its high water content, mucus has CT attenuation and MRI sig-

nal similar to that of water and does not enhance after injection of intravenous contrast medium; however, a chronic entrapped mucus collection can undergo a gradual transformation. The increased production of mucus protein is accompanied by a gradual reabsorption of water with a decrease in free water content and an increase in protein concentration. On CT, concentrated mucus may have attenuation values above 20 HU, and even as high as 130 HU. In addition, protein concentration influences the T1 relaxation time and an elevation in protein concentration may cause impressive shortening of the T1 relaxation time. Mucus with a pro-

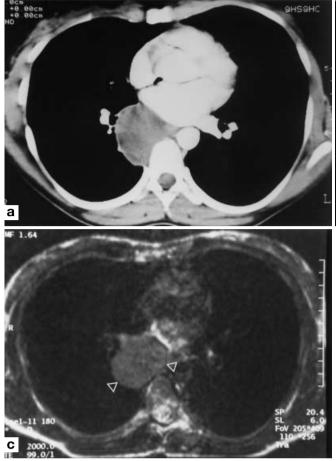




Fig.1a-c A 30-year-old woman with mediastinal bronchogenic cyst. **a** Enhanced helical CT shows a 4-cm mediastinal mass of 50-HU attenuation. The mass showed no enhancement in comparison with unenhanced scan (not shown). **b** Coronal T1-weighted MR image shows a hyperintense subcarinal mass. *C* carina; *A* aortic arch. **c** On T2-weighted axial MR image, the mass (*arrowheads*) shows a marked loss of signal. At surgery, a bronchogenic cyst with brownish, mucoid content was found. Protein concentration was 16 mg/dl

tein level > 9000 mg/dl has high signal intensity on T1-weighted images [2]. The detection of mucus within a mediastinal or pulmonary lesion is important because it permits shortening of the list of differential diagnoses [3, 4].

The purpose of this article is to illustrate the CT and MRI findings of mucinous tumors and pseudo-tumors of mediastinum, bronchial tree, and lung.

Mediastinum

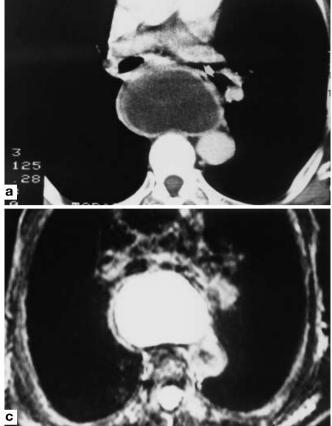
Congenital cysts

Congenital cysts encompass a spectrum of bronchopulmonary malformations that includes bronchogenic cysts, esophageal duplications, and neurenteric cyst. All of these entities probably share a common embryogenesis as a result of anomalous budding from the primitive foregut and early tracheo-bronchial tree. These cysts contain serous or mucinous fluid. Mucinous cysts are lined by mucus-secretive epithelium, and their contents range from milky white to brown mucoid material, with variable viscosities. Consequently, the CT densities of the mucinous cysts can vary from near-water to substantially higher than muscle (Fig. 1).

Most mediastinal mucinous cysts are low in signal intensity on T1-weighted MR images and very high in signal intensity on T2-weighted images reflecting the long T1 and T2 of mucus; however, proteinaceous cysts may have high signal intensity on T1-weighted images (Figs. 1, 2) [2]. High protein concentrations of over 15,000 mg/dl in cysts produce a low signal intensity on T2-weighted images (Fig. 1) [5].

Bronchogenic cysts appear as round or oval, thin-walled, smoothly marginated, homogeneous mass; approximately 90% are located within the middle mediastinum, under the carina, or along the right paratracheal wall (Fig. 1), and 10% are within the lung parenchyma, usually in the lower lobes (Fig. 3).

Esophageal duplication cysts result from failure of the originally solid esophagus to vacuolate completely to produce a hollow tube. They are usually located in the posterior mediastinum and connected to the esophageal wall (Fig. 2).



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Fig. 2a-c A 67-year-old man with dysphagia due to esophageal duplication. a A CT scan reveals a cystic mass into the posterior mediastinum. The esophagus (arrow) is compressed and strictly adherent to the cyst wall. b A sagittal T1-weighted MR image shows a mass with high signal intensity due to high protein content (10 mg/dl). T trachea; A ascending aorta. c On axial T2-weighted MR image, the mass is isointense with respect to cerebrospinal fluid in the thoracic subarachnoid space

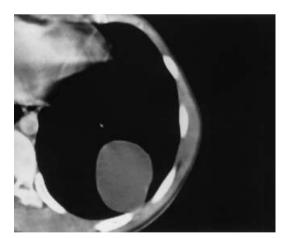


Fig. 3 Asymptomatic 25-year-old patient with pulmonary bronchogenic cyst. Contrast-enhanced CT scan shows a well-defined, water density mass in the left lower lobe

Neurenteric cysts arise from a gut anlage that has herniated through a split in the notochord. They occur in the posterior mediastinum and may be distinguished by their usual association with spinal anomalies.

Central bronchi

Mucous plugs

Mucus can accumulate within the bronchial lumen for two reasons: increased production and impaired mucociliary clearance. In the trachea and major bronchi, smaller mucous plugs appear as low-attenuation, polypoid-like lesions of the endobronchial surface with no involvement of the bronchial wall (Fig. 4). Rescanning after coughing may demonstrate the disappearance of these lesions. Larger mucous plugs may occlude the central bronchi and cause atelectasis (Fig. 5). Bronchoaspiration could be required in such cases.

Mucous gland adenoma

Mucous gland adenoma [6] is a rare, benign, multicystic tumor arising from the submucosal seromucous glands of the lobar or segmental bronchi. On CT, it presents as a low-attenuation hilar nodule growing as an intrabronchial polypoid mass which causes stenosis of the involved bronchus [6]. Usually, the residual bronchial lumen appears as an air meniscus surrounding the tumor

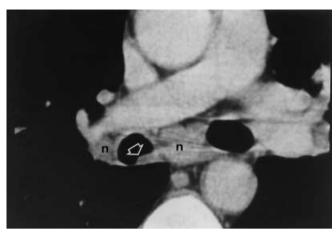


Fig. 4 A 35-year-old-man with sarcoidosis and mucous pseudotumor. A contrast-enhanced, 4-mm-thick CT scan shows a small, low-attenuation polypoid pseudotumor (*arrow*) in the intermediate bronchus. There are no findings of bronchial wall involvement. Note lymphadenopathies (*n*) due to sarcoidosis



Fig. 5 A 72-year-old man with recent abdominal surgery and obstructive mucous plug. At lectasis of the middle and lower lobes of the right lung is due to a large mucous plug (arrows) obstructing the intermediate bronchus. Asterisks indicate left main bronchus

(so-called air-meniscus sign). Histologically, mucous gland adenoma is constituted by cystic spaces rich in mucin, lined by well-differentiated mucin-secreting cells.

Peripheral bronchi and lung

Bronchocele

Bronchial atresia is a rare congenital abnormality that consists of atresia or stenosis of a segmental bronchus [7]. Mucus secreted within the patent airways distal to

the point of atresia accumulates in the form of a plug or bronchocele which appears as a pulmonary nodule or mass. Because of collateral ventilation, the bronchial obstruction is not responsible for atelectasis but for air trapping leading to regional hyperinflation (Fig. 6). Bronchocele has been identified in association with a variety of lesions that cause segmental bronchial occlusion, including cicatricial stenosis (Fig. 7), foreign-body, and bronchial neoplasms [7]. A transient bronchocele due to mucoid impaction of bronchiectasis in allergic bronchopulmonary aspergillosis and cystic fibrosis may occur without bronchial stenosis.

Mucinous adenocarcinoma

Histologically and biologically, adenocarcinoma of the lung constitutes a heterogeneous group of tumors. A defining feature of adenocarcinoma of the lung is its ability to produce mucinous substances. The overall frequency of mucin production is not known, but approximately half of adenocarcinomas of the lung are mucin positive. However, with the exception of the mucinous bronchioloalveolar carcinoma (BAC), mucin production is seldom a truly prominent characteristic of adenocarcinoma. The term "mucinous" is restricted to adenocarcinomas that are composed of at least 60% extracellular mucin [8].

Rarely, mucinous adenocarcinoma presents as a solitary pulmonary nodule with low or no uptake of contrast medium (Fig. 8). Such a lesion may simulate a benign nodule.

Mucinous BAC usually has one of two radiologic patterns: consolidation or diffuse disease. The consolidation may be segmental or may involve an entire lobe ("lobar" BAC). On CT, the low attenuation allows vessels to be seen after administration of intravenous contrast material [9]. Similarly, intratumoral vessels may be detected on T1-weighted gadolinium-enhanced MR scans (Fig. 9).

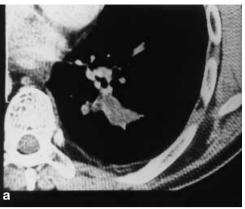
This finding, described as the angiogram sign, is suggestive but not specific for bronchioloalveolar carcinoma and may be identified in consolidations due to various causes, namely pneumonia or pneumonitis with central obstruction, atelectasis, lymphoma, lipoid pneumonia, and metastasis [10].

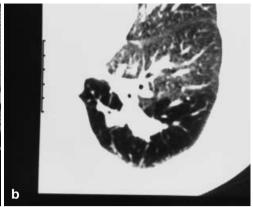
Mucinous neoplastic consolidations are isointense with respect to cerebrospinal fluid in the thoracic spine on heavily T2-weighted MR images. This finding has been named "white lung sign" (Fig. 10) [11].

Diffuse mucinous BAC usually appears as multiple low-attenuation consolidations and/or ground-glass opacities on CT scans and as multiple hyperintense lesions on T2-weighted MR images (Fig. 11).

Mucinous BAC has the tendency to disseminate into the lungs by aerogenous spread [12]. Rapid develop-

Fig. 6a, b A 44-year-old patient with bronchocele due to bronchial atresia. a A 4-mmthick, contrast-enhanced CT scan through the left lower lobe shows a water-density, branching nodule surrounded by subsegmentary pulmonary arteries. b At lung window setting the hyperlucent pulmonary parenchyma distal to the bronchial atresia is seen. Bronchoscopy confirmed bronchial atresia





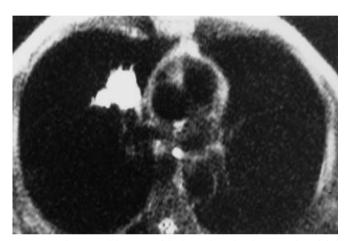


Fig. 7 A 63-year-old man with bronchocele due to cicatricial bronchial stenosis. A 4-mm-thick, heavily T2-weighted half Fourier acquisition single-shot turbo spin-echo (HASTE) image shows a sharply marginated, bifurcating, hyperintense mass in the middle lobe. Bronchoscopy demonstrated the cicatricial stenosis of the medial subsegmental bronchus of the middle lobe

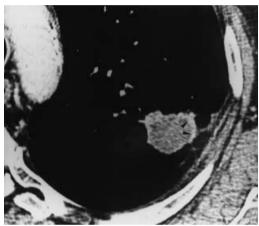


Fig. 8 A 61-year-old man with nodular mucinous adenocarcinoma. A 2-mm-thick contrast-enhanced CT scan through the left upper lobe shows a 2.5-cm nodule with irregular margins. The nodule has a low-attenuation (15 HU) and does not enhance. A contrast uptake is visible only at the periphery of the nodule (*arrowheads*). Transthoracic needle biopsy yielded mucinous neoplastic cells. The presence of irregular margins is the only CT finding of malignancy in this nodule

ment of centrilobular ground-glass nodules is the radiologic hallmark of aerogenous neoplastic dissemination (Fig. 12).

Mucinous cystoadenocarcinoma

Mucinous cystoadenocarcinoma is a rare cystic tumor composed of abundant extracellular mucin lined by mucinous epithelium. This tumor appears to have a remarkably favorable prognosis and some authors believe that it may be considered the counterpart of mucinous cystoadenoma of the ovary. On CT, tumor appears as a cystic mass with thickened wall and intratumoral enhancing areas (Fig. 13) [13].

Mucinous metastasis

Adenocarcinomas of the gastrointestinal tract are the most common source of mucinous lung metastasis. On CT, the presence of multiple low-attenuation lung nodules should raise the suspicion of mucinous metastases. Rarely, mucinous lung metastases may calcify in a manner analogous to that of mucinous hepatic metastases (Fig. 14).

Less frequently, metastases from adenocarcinoma may be spread into the lung along the intact alveolar wall (lepidic growth) in a fashion similar to BAC, so-called air-space metastasis (Fig. 15) [14, 15].



Fig. 9 A 70-year-old man with mucinous "lobar" bronchioloalveolar carcinoma. Gadolinium-enhanced T1-weighted MR image shows a non-segmental pulmonary consolidation of the right lower lobe. Consolidation has low signal intensity and contains enhanced vessels (angiogram sign) and patent bronchi

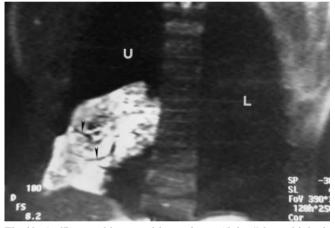


Fig. 10 A 67-year-old man with mucinous "lobar" bronchioloal-veolar carcinoma. A 4-mm-thick coronal HASTE MR image shows a hyperintense consolidation of the right lower lobe. Patent bronchi can be seen within the consolidation as tubular areas of signal void (arrowheads). No signal from the normal left lung (L) and right upper lobe (U)

Drowned lung

Obstructive pneumonia occurs for retention of epithelial secretions distal to an obstructing airway tumor. The most obvious retained secretions is mucus, which accumulates within bronchi and bronchioles and occasionally extends into alveoli [16]. In addition, alveoli may be filled with a proteinaceous material resembling edema fluid. If the amount of retained secretions is large, there is no or mild atelectasis of the lung. The result is termed

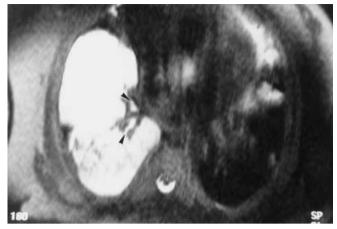


Fig. 11 A 70-year-old man with bronchorrhea due to diffuse mucinous bronchioloalveolar carcinoma. A 4-mm-thick HASTE MR image shows a high-signal-intensity mucinous consolidation involving the middle and the lower lobe of the right lung. Other mucinous lesions are visible in the left lung. The involved lung is isointense with respect to cerebrospinal fluid in the thoracic subarachnoid space. Note hyperintense mucous plugs into the middle and right lower lobe bronchi (*arrowheads*)

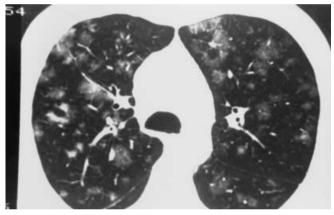
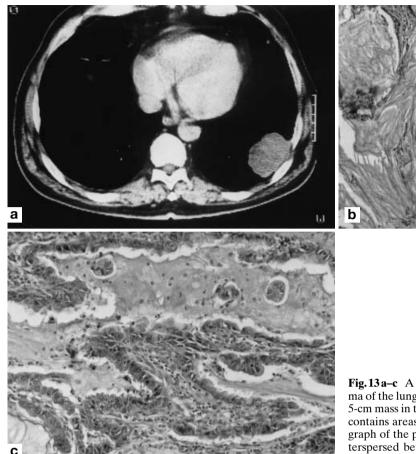


Fig. 12 A 58-year-old man with aerogenous spread of bronchioloalveolar carcinoma. A 2-mm-thick high-resolution CT scan through the upper lobes shows multiple centrilobular ground-glass nodules. The patient had a right lower lobe neoplastic consolidation due to mucinous bronchioloalveolar carcinoma (not shown). Transbronchial biopsy and bronchoalveolar lavage of the upper lobes yielded mucinous cancer cells

drowned lung. On CT and MRI scans, drowned lung may mimic mucinous BAC (Fig. 16). The demonstration of a central obstructing tumor permits diagnossis of drowned lung (Fig. 16).

Sequestration

Lung sequestration is a congenital anomaly in which a mass of pulmonary parenchyma is separated from the



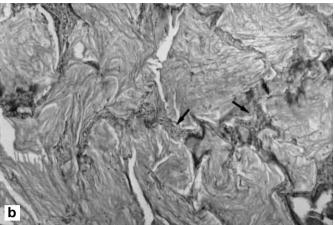


Fig. 13 a–c A 45-year-old man with mucinous cystoadenocarcinoma of the lung. **a** A 4-mm-thick contrast-enhanced CT scan shows a 5-cm mass in the left lower lobe. The mass has low-attenuation and contains areas with mild enhancement. **b** Low-power photomicrograph of the pathologic specimen reveals cancer cells (*arrows*) interspersed between abundant mucus. **c** High-power photomicrograph clearly shows mucin-producing cancer cells



Fig. 14 An 83-year-old woman with calcified mucinous lung metastases from colorectal cancer. A 4-mm-thick CT scan shows two densely mineralized lung masses in the lower lobes

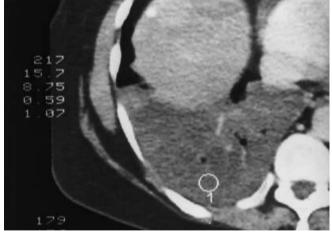
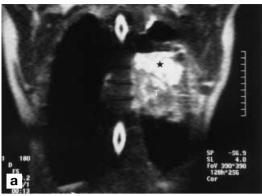


Fig. 15 A 74-year-old woman with mucinous air-space metastasis from pancreatic adenocarcinoma. Two years after resection of pancreatic mucinous adenocarcinoma, enhanced CT shows a low-attenuation consolidation (15 HU) of the right lower lobe. Enhancing vessels (angiogram sign) and patent bronchi is seen within the consolidation. Sputum levels of amylase and CA 19–9 were elevated. Biopsy yielded mucinous cancer cells. This type of metastasis simulates bronchioloalveolar carcinoma. (From [13])

Fig. 16a, b A 62-year old man with drowned lung due to central small-cell carcinoma. a A 4-mm-thick coronal HASTE MR image reveals a central tumor (T) and a high-signal-intensity segmental consolidation (drowned lung; asterisk). b Source image from MR pulmonary angiography reveals vessels (arrow) within the segmental consolidation (angiogram sign) and the hypovascular tumor (T)



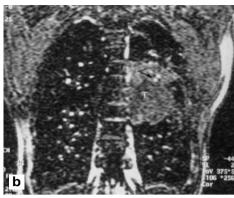




Fig. 17 A 70-year-old patient with cystic pulmonary sequestration. Curved transverse reformation from helical CT angiography of aorta demonstrates a cystic lung mass in the left lower lobe. A left arterial branch originates from the aorta to supply the mass (*small arrow*). A right aberrant branch (*large arrow*) supply to otherwise normal right lower lobe parenchyma. (From [17])

normal lung. It can be classified into two types: intralobar and extralobar. In the intralobar variety the arterial supply is usually from the thoracic or abdominal aorta, and the venous drainage terminates in the pulmonary veins. In the extralobar variety, the sequestered lung is covered by its own visceral pleura. The arterial supply and venous drainage are provided by systemic vessels. Macroscopically, pulmonary sequestration may show a wide spectrum of appearances. The cystic pattern (with a solitary cyst or polycysts containing mucus) is frequent (Fig. 17). Helical CT or MR angiography, demonstrating systemic arterial supply, permit diagnosis [17].

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