

## Elevated Carbohydrate Antigen 19-9 Level in a Patient with Horseshoe Type Pulmonary Sequestration

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Elevated carbohydrate antigen (CA) 19-9 can indicate malignancies of the gastrointestinal, pancreatic, and biliary tracts, and be found in a pulmonary sequestration. A 30-year-old man visited Seoul National University Bundang Hospital due to elevated CA 19-9 levels, representing pulmonary sequestration of the bilateral lower lobes, which were connected with each other. We performed left lower lobectomy and division of the systemic arteries. After operation, CA 19-9 levels decreased to normal range, even though a small amount of sequestered lung remained in the right lower lobe. It is not uncommon that presence of pulmonary sequestration might elevate serum CA 19-9 levels; however, horseshoe type bilateral pulmonary sequestration is very rare.

**Key words:** 1. Bronchopulmonary sequestration  
2. Carbohydrate antigen 19-9  
3. Horseshoe lung

### Case report

Elevated levels of the serum carbohydrate antigen (CA) 19-9 can indicate malignancies of the gastrointestinal, pancreatic, and biliary tracts. However, CA 19-9 levels can also be elevated in a pulmonary sequestration. We report the case of a patient with pulmonary sequestration of the bilateral lower lobes, which were connected with each other and associated with an elevated CA 19-9 level. The patient was treated successfully with thoracoscopic left lower lobectomy and division of systemic arteries.

A 30-year-old man was referred to our gastroenterology department for elevated serum CA 19-9 and carcinoembryonic antigen (CEA) levels. The patient had no abdominal pain and no abnormal findings on

physical examination. The patient had never smoked and was a social drinker. Routine labs, which included evaluation of tumor markers, esophagogastroduodenoscopy, pancreatobiliary computed tomography (CT), and whole body positron emission tomography/CT (PET/CT), were performed to rule out malignancies of the gastrointestinal, pancreatic, and biliary tracts. The serum amylase, lipase, and liver enzymes lab profiles showed no abnormalities. The CEA level was low at 3.5 ng/mL (within normal range), but the CA 19-9 level was elevated at 2,400 U/mL. Pancreatobiliary CT was normal and the PET/CT scan demonstrated no abnormal hypermetabolic lesion, suggesting malignancy. However, the abdominal CT scan from the previous admitting hospital showed multiple, variable sized, cystic lung lesions, suggesting pul-

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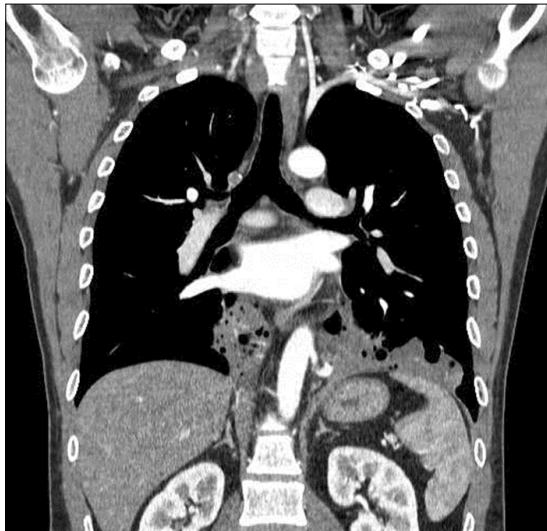


Fig. 1. Computed tomography findings of each abnormal systemic artery from the thoracic aorta.



Fig. 2. Computed tomography findings of pulmonary sequestration in the horseshoe lung.

monary sequestration at the left lower lobe. The patient was then referred to the thoracic surgery department. A CT angiography of the thoracic aorta confirmed intralobar pulmonary sequestration by the presence of a systemic vascular supply from the descending thoracic aorta (Fig. 1).

The patient refused to undergo surgery and visited our gastroenterology department 32 months later, presenting with cough and sputum. The serum CA 19-9 level was 4,200 U/mL. Chest CT showed pneu-

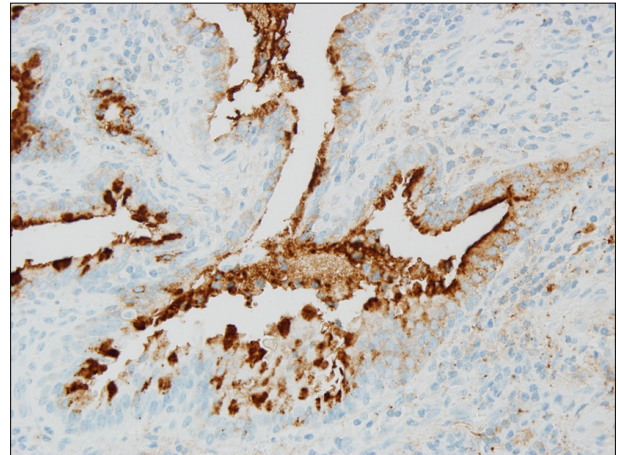


Fig. 3. Immunohistochemistry staining against human carbohydrate antigen 19-9 ( $\times 400$ ).

monia combined with pulmonary sequestration. The pulmonary sequestration was located in bilateral lower lobes, although mainly in the left lower lobe, and connected to each other (Fig. 2). Left lower lobectomy by video-assisted thoracic surgery and division of the systemic feeding arteries were performed. Intraoperatively, the sequestered lung of the left lower lobe parenchyma was connected to the right side, passing between the aorta and the pericardium. Two abnormal systemic arteries were ligated from the thoracic aorta. Resection was not performed because the pulmonary sequestration of the right lower lobe was too small. The chest tube was removed on postoperative day 4 and the patient was discharged on postoperative day 6 without any postoperative complications. Eight months later, the CA 19-9 level decreased from 4,600 to 15.8 U/mL, even though the pulmonary sequestration of the right lower lobe was still present. Intralobar type pulmonary sequestration was confirmed on pathologic examination. Immunohistochemistry staining against human CA 19-9 showed an obvious positive result in the respiratory epithelium and alveolus, and was especially condensed in the mucous cyst (Fig. 3).

## Discussion

Intralobar pulmonary sequestration is a congenital malformation that is characterized by a nonfunctional lung lesion that does not communicate normally with the bronchial tree [1]. In this case, intralobar pulmo-

nary sequestration had not been diagnosed until the patient visited the hospital due to an elevated CA 19-9 level. CA 19-9 is a useful serum marker for detecting the pancreatobiliary or gastrointestinal cancers. However, serum CA 19-9 level can rise when there are inflammation or proliferation of normal tissues, such as pancreatitis, cholangitis, bronchiectasis and ovarian cysts. CA 19-9 level is also elevated when the pathway discharging CA 19-9 is obstructed by duct stenosis or stones. Additionally, chronic hepatitis and chronic kidney disease can increase serum CA 19-9 level by hindring CA 19-9 metabolism [2]. CA 19-9 secreted by the bronchiolar epithelia may appear in the serum as a result of airway damage in the lower respiratory tract, such as pulmonary sequestration. Bronchial mucus contains large amounts of CA 19-9, which appears to be produced in the columnar epithelia of respiratory glands, even if the serum CA 19-9 levels are normal [3]. The result of CA 19-9 immunohistochemistry staining is consistent with these previous studies. Therefore, our findings can explain the elevated CA 19-9 in pulmonary sequestration. The marked decrease of serum CA 19-9 level after pulmonary resection confirms that the patient's increased level of serum CA 19-9 was due to pulmonary sequestration. The right lower lobe lesion was connected to the left lower lobe, which was diagnosed as horseshoe lung. Horseshoe lung is a very rare congenital anomaly in which the bilateral lung parenchyma conjoin in the posterior mediastinum, usually anterior to the aorta. There have been dozens of articles in English journals, but most of the case reports are correlated with other congenital anomalies like Scimitar syndrome and were identified in infancy or early childhood. Horseshoe lung can be misapprehended with mediastinal lung herniation in terms of the presence of pulmonary parenchyma in the midline of the posterior mediastinum. Mediastinal lung herniation, which is commonly associated with pulmonary sequestration,

have 4 layers of the pleura in the midline connecting region of the lung parenchyma. On the other hand, horseshoe lung is the synthesis of bilateral lung segments through an abnormal parietal pleura gap. The lung parenchyme isthmus are generally part of more hypoplastic side of the bilateral lung segments. The pleural layers are hard to be distinguished on high resolution CT [4].

In conclusion, this case shows that pulmonary sequestration can be a cause of elevated CA 19-9 levels. Chest CT can be considered as a diagnostic option for unexplained elevated CA 19-9 levels. This case is also interesting because of the accompanying diagnosis of horseshoe lung. We will follow up with monitoring the patient's CA 19-9 levels. Contralateral thoracic surgery can be considered if the patient presents with respiratory infections associated with the remaining lung lesion in the future.

### Conflict of interest

No potential conflict of interest relevant to this article was reported.

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