

## Case Report

# Left middle lobe resection for typical carcinoid in a patient with complete situs inversus

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**Abstract:** We present the first report of a rare case of typical carcinoid tumor of the lung in a patient with complete situs inversus. The patient was a 65-year-old woman with the chief complaint of productive and mild chest pain. Chest X-ray and computed tomography (CT) scans of the thorax showed a mirror image of the organs and vessels and revealed a tumor 2.5 cm in diameter in the left middle lobe. The patient underwent resection of the left middle lobe through posterolateral incision. Postoperative pathological examination returned a diagnosis of typical carcinoid tumor. The patient made an uneventful recovery. This case highlights the importance of considering malignant tumors in pulmonary nodules in patients with complete situs inversus.

**Keywords:** Complete situs inversus, carcinoid tumor, lung, left lobectomy

## Introduction

Complete situs inversus is a full mirror image of the normal arrangement of the thoracic and abdominal viscera. The syndrome results from abnormal rotation of the cardiac tube during embryogenesis. It is rare syndrome with an estimated prevalence of 1/10,000 births and has an unknown mechanism of development. Patients with situs inversus have a life expectancy similar to that of the general population. Diagnosis of complete situs inversus is usually based on clinical manifestations and thoracic radiography and electrocardiogram.

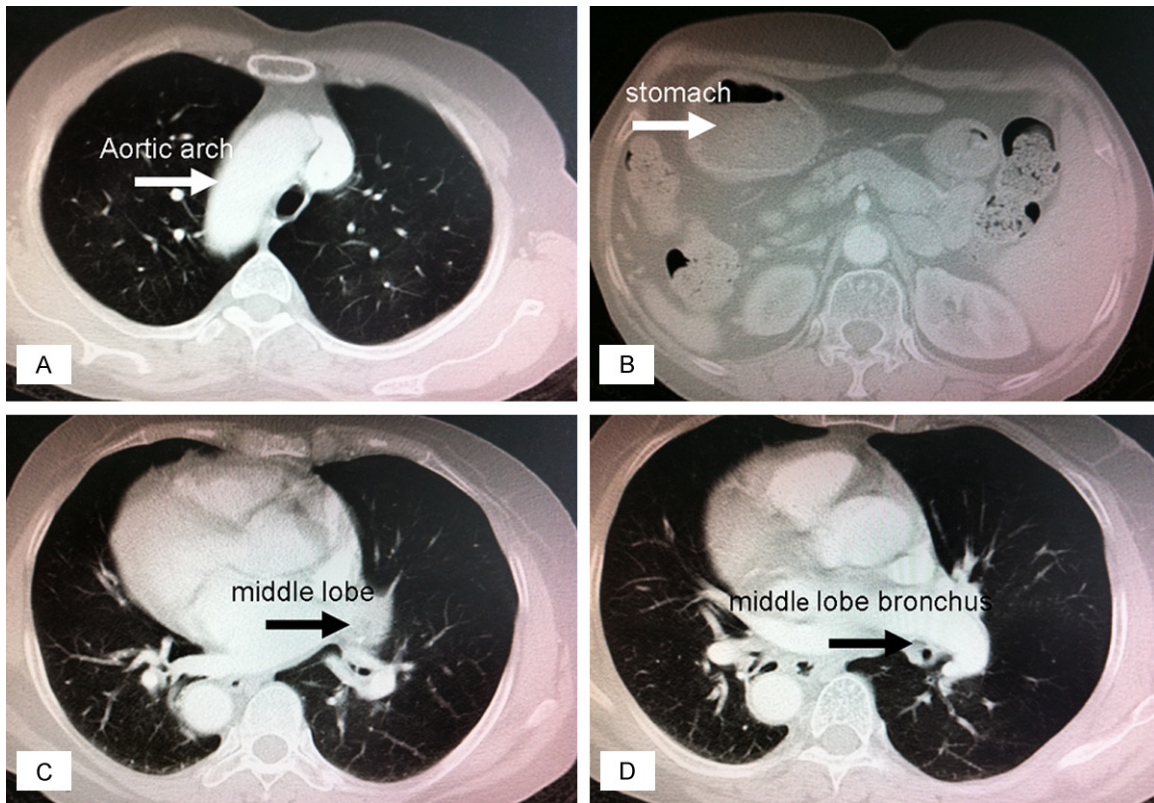
A carcinoid tumor, which is a rare type of neuroendocrine tumor, is derived from enterochromaffin cells and occurs mainly in the gastrointestinal tract [1]. It may also occur in the bronchopulmonary system and manifests as recurrent episodes of pneumonia, cough, hemoptysis, and chest pain [2, 3]. Pulmonary carcinoid tumors are low-grade malignancies. They belong to the neuroendocrine neoplasms, which account for 1%-2% of all lung malignancies and approximately one fifth of neuroendocrine neoplasms. To our knowledge, there has been no report of pulmonary carcinoid tumor in patients with complete situs inversus. Here,

present the first report of a rare case of typical carcinoid tumor of the lung in a patient with complete situs inversus who was successfully managed by left lobectomy.

## Case report

A 65-year-old female patient was admitted to our hospital because of productive cough and mild chest pain on the left side for five years. The patient was conscious and cooperative during the examination, and was found to be in a good general state of health and nutrition. The results of hematological tests and urinalysis on admission showed no abnormalities. All tumor markers were within their normal range. Spirometry revealed normal respiratory function (VC, 2.1 L; %VC, 90.3%; FEV1.0, 1.62 L; FEV1.0%, 70.0%). Arterial gas analysis fell within their normal ranges (PaCO<sub>2</sub>: 36.5 mmHg; PaO<sub>2</sub>: 85.0 mmHg; SaO<sub>2</sub>: 96%).

Computed tomography (CT) scan of the thorax showed a mirror-image of the usual arrangement of organs and vessels in which the aortic arch (**Figure 1A**) and the stomach (**Figure 1B**) were seen located on the right. A shadow was also observed on the CT scan (**Figure 1C**) in the left middle lobar location and a blurred shadow



**Figure 1.** Preoperative chest CT scan of a 65-year-old female patient showing a mirror image of intrathoracic organs and vessels in their usual arrangement. A: The ascending aorta (indicated by arrow) on the right side; B: The stomach (indicated by arrow) on the right side; C: A shadow (indicated by arrow) in the middle lobe; D: A blurred shadow (indicated by arrow) in the left middle lobe bronchus. AA, ascending aorta; DA, descending aorta; PA, pulmonary artery; SVC, superior vena cava.

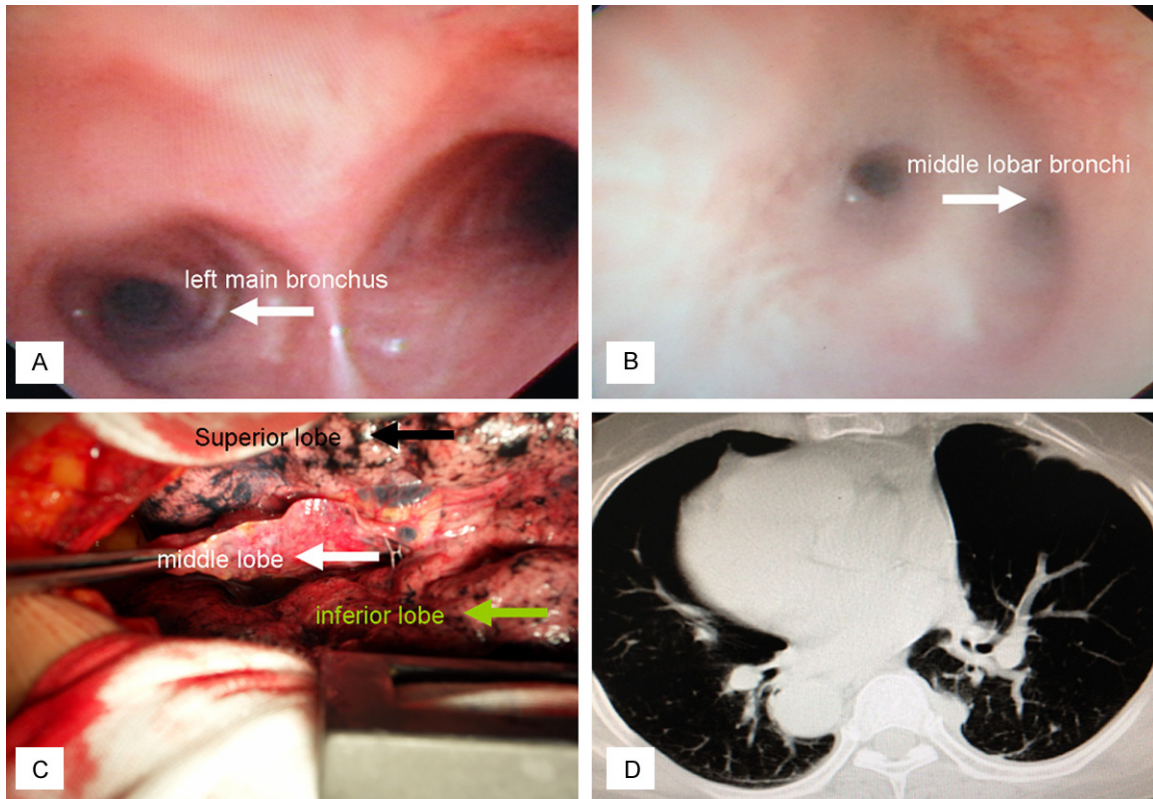
(**Figure 1D**) inside the left middle lobe bronchus was observed. The blurred shadow was considered pulmonary consolidation with atelectasis and sputum, respectively. Bronchoscopy revealed that the first lobar bronchus from the left main stem bronchus was located 1.5 cm from the carina (**Figure 2A**), which corresponded to the right side pattern. The left middle lobe bronchus was blocked by sputum, and when sputum was cleaned by suction, the middle lobe bronchus was observed on the left side (**Figure 2B**). There were no local lesions in other areas of the lungs during the CT scan. Malignancy cannot be established by brush cytology, which revealed only benign columnar cells, epithelioid cells, and lymphoid cells; therefore, a preliminary diagnosis of middle lobe syndrome was made.

The patient underwent resection of the left middle lobe through posterolateral incision. Upon examination, the left lung was composed of three lobes separated by well-defined fis-

tures (**Figure 2C**), which are consistent with typical features of the normal right side. The gross appearance of the left lung and the arrangement of the pulmonary vessels and the bronchi corresponded to those normally found on the right side. The left middle lobe showed pulmonary consolidation with hard texture and atelectasis.

Histological analysis revealed typical carcinoid. Hematoxylin and eosin (H&E) staining showed uniform tumor cells with oval nuclei and moderate cytoplasm. The cells were arranged in an acinar-like or trabecular pattern. Immunohistochemistry further showed that the tumor cells were positive for the epithelial marker CAM 5.2 and neuroendocrine markers, chromogranin A and synaptophysin.

The patient made an uneventful postoperative recovery and the patient was discharged 2 weeks after surgery. CT at 2 years of follow up revealed no recurrence (**Figure 2D**).



**Figure 2.** Intraoperative findings and postoperative imaging studies. A: The first lobar bronchus (indicated by the white arrow) is seen in the left lung; the black arrow indicates the carina. B: The middle lobe bronchus is patent. C: The three lobes of the left lung. D: No recurrence was seen during subsequent follow up visits.

## Discussion

A carcinoid tumor is a rare type of neuroendocrine tumor and when it occurs in the bronchopulmonary system, the disease manifests as recurrent episodes of pneumonia, cough, hemoptysis, and chest pain [2, 3]. Our patient had a protracted course of repeated episodes of productive cough and chest pain over 5 years with unexplained causes. CT scan unexpectedly revealed complete situs inversus in the patient. The CT scan also showed a shadow in the left middle lobe of the patient. Pulmonary shadows on CT scans are not infrequent, which often require further evaluation because of the concern for lung cancer. Gross intraoperative observation of the shadow region strongly suggested atelectasis of the left middle lobe; furthermore, brush cytology also failed to reveal the presence of malignant cells. By contrast, postoperative pathologic examination demonstrated features of typical carcinoid tumors, which were further confirmed by immunohistochemistry findings. These results suggest that unsuspected lung shadows and middle lobe

syndrome should be investigated for possible presence of malignancy.

Complete situs inversus is a rare syndrome and is seen only roughly in one out of 10,000 births. There has been no previous report of carcinoid tumor in patients with complete situs inversus. Here, we have provided the first documented case of carcinoid tumor in a patient with complete situs inversus.

## Disclosure of conflict of interest

None.

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## References

- [1] Pinchot SN, Holen K, Sippel RS and Chen H. Carcinoid tumors. *Oncologist* 2008; 13: 1255-1269.

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- [2] McCaughan BC, Martini N and Bains MS. Bronchial carcinoids: Review of 124 cases. J Thorac Cardiovasc Surg 1985; 89: 8-17.
- [3] Asamura H, Kameya T, Matsuno Y, Noguchi M, Tada H, Ishikawa Y, Yokose T, Jiang SX, Inoue T, Nakagawa K, Tajima K and Nagai K. Neuroendocrine neoplasms of the lung: a prognostic spectrum. J Clin Oncol 2006; 24: 70-76.