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Uniportal left middle lobectomy in a patient with situs inversus totalis: a case report

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Abstract

Background Situs inversus totalis (SIT), a rare recessive autosomal disease, involves the complete transposition of the thoracic and abdominal viscera in the left-right axis. Patients with SIT combined with lung cancer are extremely uncommon.

Case presentation We present a case of a 57-year-old woman with SIT who underwent uniportal video-assisted thoracoscopic left middle lobectomy for adenocarcinoma of the lung. The procedure was performed safely with adequate anatomical identification and careful intraoperative manipulation based on the preoperative three-dimensional-computed tomography bronchography and angiography (3D-CTBA). The patient's perioperative period was uneventful, and no recurrence was observed 2 year postoperatively.

Conclusion With the preoperative planning of the 3D-CTBA, uniportal video-assisted thoracoscopic lobectomy in lung cancer patients with sit can be performed safely and effectively.

Keywords Situs inversus totalis (SIT), Lung cancer, Thoracic surgery, UVATS

Background

Situs inversus totalis (SIT) is a rare congenital autosomal recessive genetic condition, which is probably associated with an X-chromosome defect [1, 2]. Although the incidence of SIT varies, the majority of the available literature describes it as affecting 1:8000 to 1:20000 newborns [3–5]. SIT patients present with inversed but regular lung anatomy, which means a symmetrical and complete transposition in the right-left axis of the major visceral organs within the thorax and abdomen.Vascular and bronchial anomalies are more frequent in patients with SIT than in the general population. Therefore, it requires particular attention to prevent complications in SIT patients [5]. Interventions in patients with uncomfortable reversal anatomy are a challenge for thoracic surgeons. Three-dimensional (3D) reconstruction software is a powerful tool for preoperative anatomic evaluation, which ensures a personalized and safe anatomic thoracoscopic lobectomy. This is the first report to describe a case of concurrent primary lung adenocarcinoma with SIT treated successfully by uniportal video-assisted thoracoscopic left middle lobectomy.

Case presentation

A 57-year-old non-smoking woman visited The First Affiliated Hospital of Soochow University in May 2022 with a persistent cough for 3 months. The patient was diagnosed with SIT in early youth (Fig. 1a). Chest

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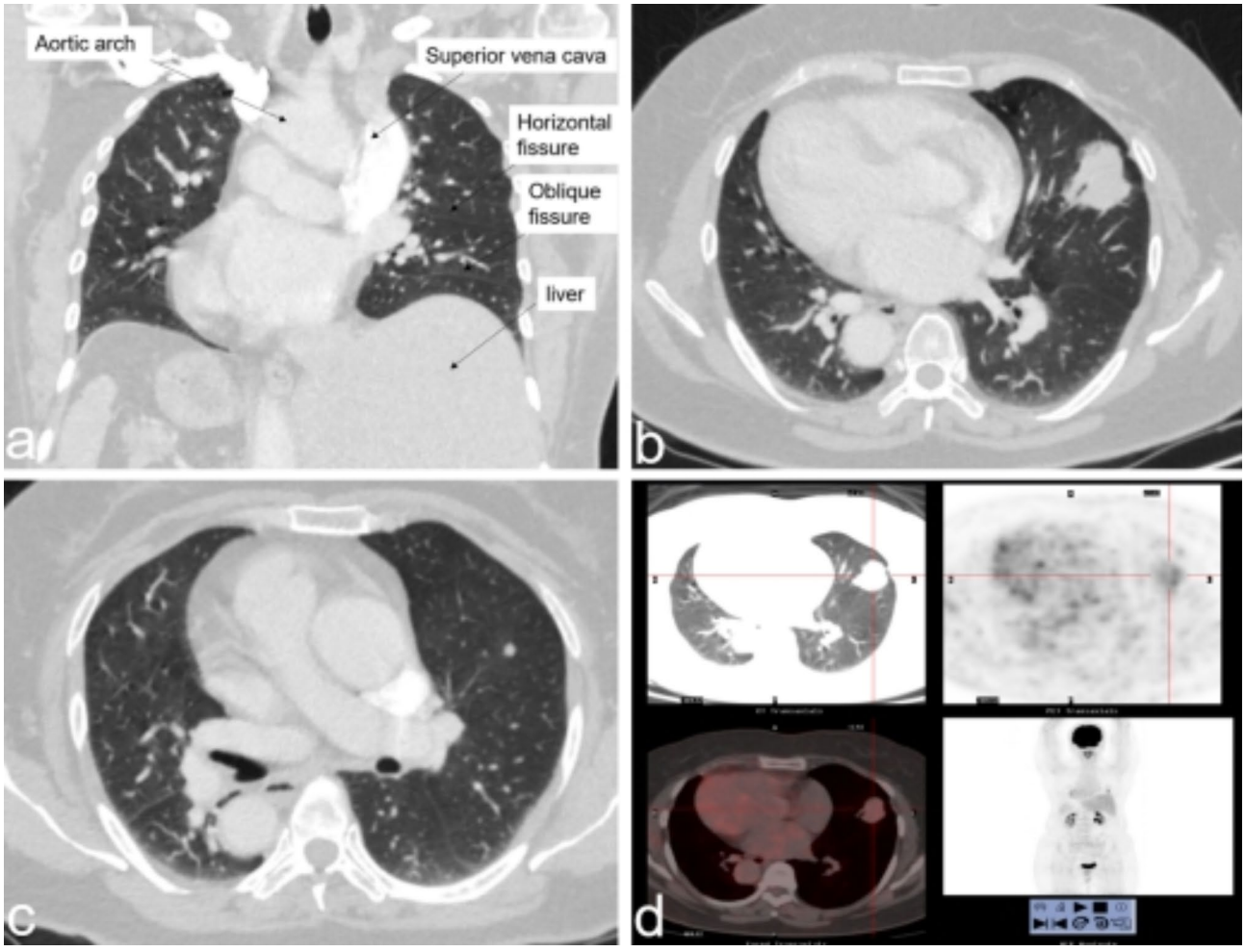

Fig. 1 Radiological findings of the pulmonary tumor and thoracic anatomy of the patient (**a**) CT image in the coronal view reveals situs inversus totalis. (**b**) Chest CT shows an irregularly shaped solid tumor shadow (measuring 35 × 25 mm in diameter) in the peripheral pleural area of the left middle lobe. (**c**) Chest CT shows a 5-mm solid nodule in the left upper lobe attached to the interlobar pleura. (**d**) 18 F-FDG-PET shows a hot spot in the tumor. (CT, computed tomography; 18 F-FDG-PET, fluorine 18 fluorodeoxyglucose-positron emission tomography)

computed tomography (CT) revealed an irregularly shaped solid tumor shadow, measuring 35×25 mm in diameter, in the peripheral pleural area of the left middle lobe (Fig. 1b) and a 5-mm solid nodule in the left upper lobe attached to the interlobar pleura (Fig. 1c). The diagnosis of adenocarcinoma was confirmed based on the CT-guided percutaneous lung biopsy results (cT2aN0M0, stage IB). On admission, the patient's vital signs were normal. The routine workup was conducted methodically, which revealed no swollen mediastinum and hilar lymph nodes and/or distant metastatic lesions. Positron emission tomography computed tomography (PET/CT) showed fluorodeoxyglucose accumulations in the tumor of the left middle lobe with standard uptake values of 3.70 (Fig. 1d). Accordingly, left middle lobe lobectomy and left upper lobe partial resection via uniportal video-assisted thoracoscopic surgery (UVATS) was planned.

To assess the possibility of anatomical variations of the pulmonary vessels, other than the transposition observed in SIT, and to make reasonable preoperative decisions, we reconstructed images using 3D-CT bronchography and angiography (3D-CTBA) preoperatively (Fig. 2). The surgical procedures are described below. The patient was placed in a right lateral decubitus position, and general anesthesia was induced. One-lung ventilation was performed using a double-lumen endotracheal tube (DLT) designed for the right side. The UVATS approach was planned through a single 3-cm incision in the fourth intercostal space of the anterior axillary line. In the immediate intraoperative period, the anatomy of the left thoracic cavity was consistent with the preoperative expectation. It was established that the left lung was lobulated into three lobes by a well-defined oblique fissure and a horizontal fissure. Moreover, the location of the azygos vein and superior vena cava exactly corresponded

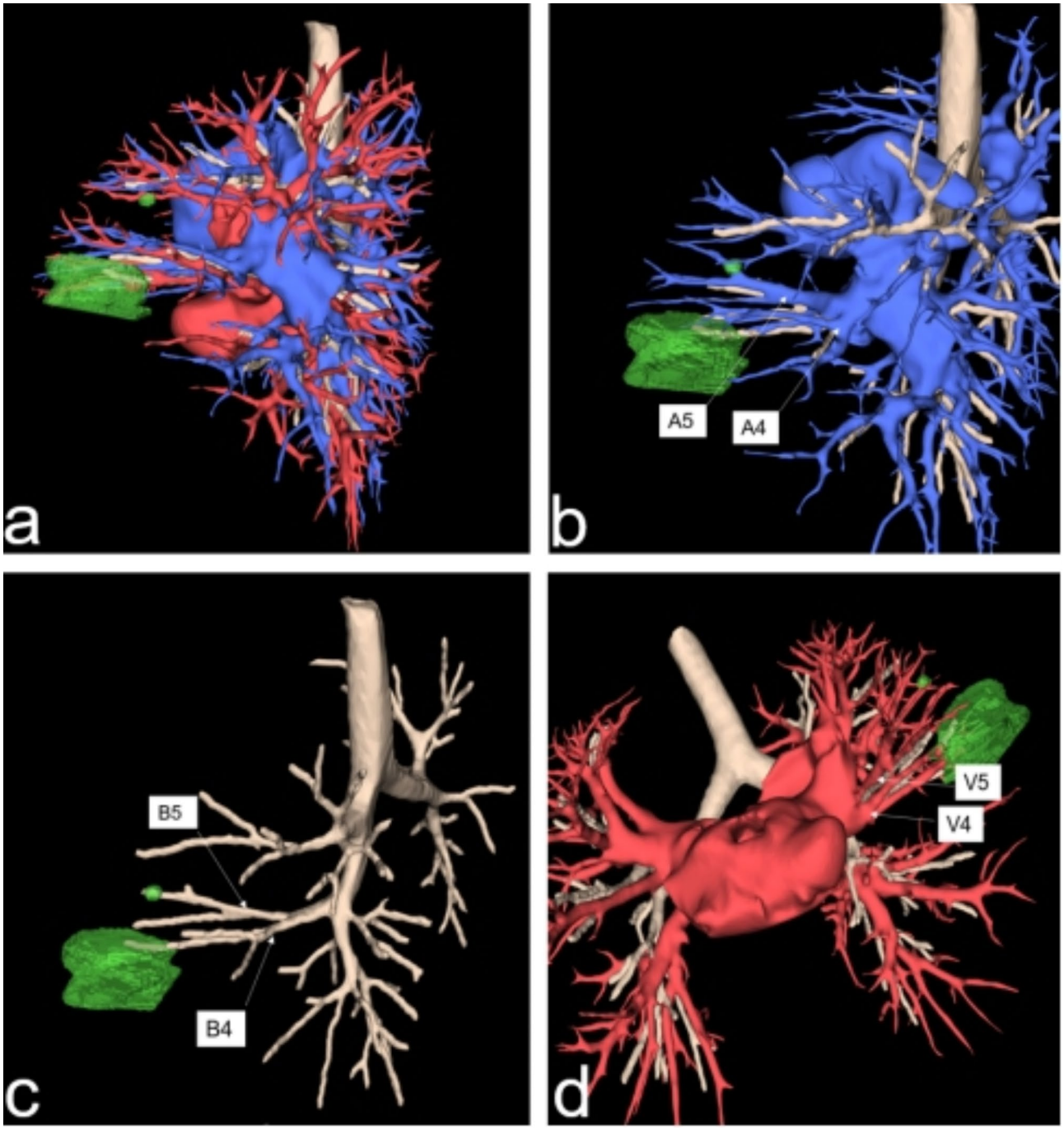

Fig. 2 A preoperative 3D reconstruction of the left pulmonary anatomic structure. (3D, three-dimensional)

to the arrangement in the right thoracic cavity, and the aortic arch was absent in the left thoracic cavity. A tumor of diameter 3.0 cm with pleural depression was detected in the middle lobe. A yellow nodule of approximately 7 mm in diameter was found in the middle horizontal fissure of the upper lobe. Before resection of the left middle lobe, the three middle lobe branches of the pulmonary veins, middle lobe bronchus, and middle pulmonary artery were excised sequentially (Fig. 3). Subsequently, the pulmonary nodule in the upper lobe was removed by wedge resection, and the two lesions were sent for pathological examination. The 2–4 and 11 thoracic lymph nodes were sampled during the procedure.

The total surgical time was 125 min, with approximately 50 mL of blood loss. A 28-F chest tube was placed through the incision and removed on the third day postoperatively. The patient was discharged on the fourth postoperative day without any complications.

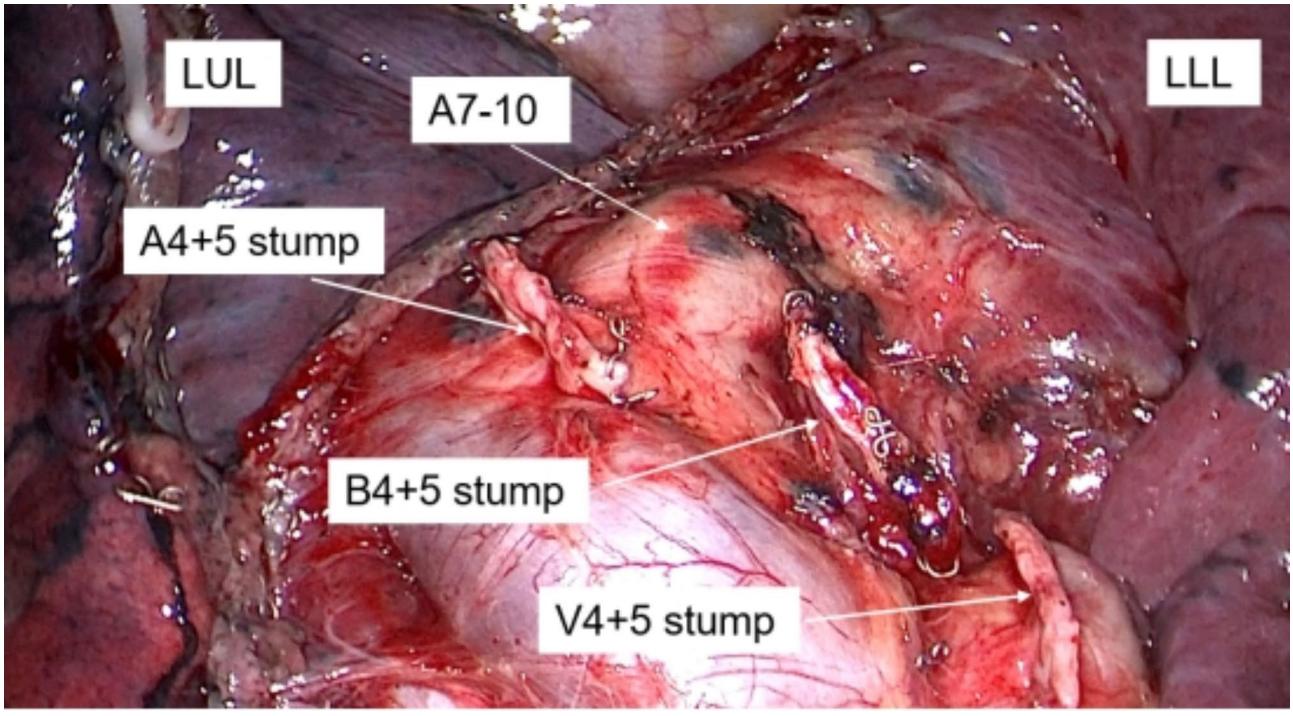

Fig. 3 Intraoperative views. (LUL, left upper lobe; LLL, left lower lobe)

The final pathological examination revealed that the tumor in the left middle lobe was an adenocarcinoma (4.0×3.0×1.8 cm). Additionally, the surgical margin, visceral pleura, and lymph nodes were negative for carcinoma. The pathological diagnosis of the nodule in the upper lobe was chronic inflammation with collagenization. No recurrence or complication was observed on chest CT during the two years postoperatively.

Discussion

SIT is a rare congenital deformity [6], in which the thoracic and abdominal viscera are arranged as a complete mirror image of the normal configuration in the right-left axis in the patient. This anomaly is usually found in conjunction with Kartagener syndrome in approximately 20–25% of patients, which is characterized by primary ciliary dyskinesia [7]. Our patient did not have Kartagener syndrome. However, if present, the surgical procedure could be more challenging due to strong adhesions in the connective tissue and increased and irregular vascularity due to chronic and recurrent infections of the bronchopulmonary tree [8]. Radiologic images of bronchial dilation and the presence of Kartagener syndrome-related symptoms, such as hemoptysis and pneumonia, could help surgeons in diagnosis.

Compared to normal anatomy, the risks and challenges during surgery are high in SIT patients. Previous studies suggest that patients with SIT present with inversed but regular lung and vessel anatomy, which enables the use of the VATS technique [9, 10]. However, the research on thoracoscopic lobectomy is limited, and it is mostly performed by the multiportal approach. Due to the relative fixation of the camera in the uniportal approach and reduction in the operating space, UVATS requires advanced surgical skills and proficiency of both the operator and assistant, especially in cases with difficulty in diagnosis and orientation and dissection during surgery due to the anatomical variations in SIT. In SIT, the organs are sagittally reversed, and the courses of the pulmonary vessels and bronchi also show some variations. Accurate identification and appropriate handling of the vascular and bronchial branches in the hilum of the lung is crucial. Therefore, individual preoperative anatomical simulations and accurate preoperative evaluation are conducive to the safe and efficient performance of UVATS of anatomical lobectomy in SIT patients. The 3D-CTBA could meticulously display the mirror-imaged anatomy and vascular patterns for further analysis of the regional anatomy and reasonable preoperative planning, which finally helped us avoid blind dissection of the vessels and unexpected complications during the procedure. Preoperative imaging can also help to select the appropriate surgical incision for different vascular variations [11].

Based on our experience and relevant literature, the optimal option is inverting the tube, if a DLT is required during surgery in SIT patients [12]. In this case, the left bronchial anatomy was situated on the right, and a conventional right-sided DLT was inserted in the left main

bronchus. This method allowed us to perform one-lung ventilation safely without a specially designed instrument for SIT cases. We believe that this method can provide some references and ideas for thoracic surgeons when they encounter similar situations. Moreover, considering that the neurological structures may not be clearly imaged preoperatively, particular vigilance is necessary to avoid intraoperative injury to the recurrent laryngeal nerve.

The etiology of SIT has not been elucidated; however, evidence suggests that it could be associated with improper extra-embryonic fluid flow and false heart tube rotation by disturbed ciliary motility during embryogenesis [1, 5, 7]. Moreover, the correlation between SIT and neoplasms is controversial and requires further clinical and epidemiological studies, which is of great relevance for the prevention, diagnosis, and treatment of the disease.

Conclusions

Despite the anatomical variations and possible intrathoracic adhesions being extremely challenging, uniportal video-assisted thoracoscopic lobectomy can be performed safely and efficiently in SIT patients with detailed preoperative anatomic evaluation by experienced thoracic surgeons.

Abbreviations

SIT	Situs inversus totalis
3D-CTBA	Three-dimensional-computed tomography bronchography and angiography
DLT	Double-lumen endotracheal tube
PET/CT	Positron emission tomography computed tomography
UVATS	Uniportal video-assisted thoracoscopic surgery

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Author contributions

Conceptualization, XY.J., J.Z. and C.L.; methodology, XY.J., J.Z. and C.L.; software, J.Z. and C.L.; validation, XY.J., J.Z. and C.L.; formal analysis, XY.J., J.Z. and C.L.; investigation, XY.J., J.Z. and C.L.; resources, J.Z. and C.L.; data curation, XY.J., J.Z. and C.L.; writing—original draft preparation, XY.J., J.Z. and C.L.; writing—review and editing, All authors.; supervision, J.Z. and C.L.; project administration, J.Z. and C.L.; funding acquisition, J.Z. and C.L. All authors have read and agreed to the published version of the manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Informed consent

Informed consent was obtained from all subjects involved in the study.Written informed consent has been obtained from the patient(s) to publish this paper.

Competing interests

The authors declare no competing interests.

Conflict of interest

The authors declare no conflict of interest.

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