Situs inversus totalis (SIT) is a congenital and autosomal recessive genetic disease that is connected with X-chromosome defect, and the frequency of SIT is 1-2/10,000 newborns [1]. SIT involves a complete reversal of the normal arrangement of the thoracic and abdominal viscera [2], and it requires particular care to prevent complications during surgery. Here we describe a successful thoracoscopic lung lobectomy for a lung cancer patient with SIT.

**Case Report**

A 61-year-old woman with SIT was referred to our hospital because of an abnormal shadow on chest radiography. Chest computed tomography (CT) revealed an irregularly shaped nodule (measuring 25×12 mm in diameter) in the left upper lobe (S2) attached to the interlobar pleura (Fig. 1A) and SIT (Fig. 1B). Bronchoscopy confirmed that the left and right bronchial branches were mirror images of each other. As we were unable to obtain a definitive diagnosis by the bronchoscopic approach,
surgery was planned for diagnosis and treatment. Before the surgery, three-dimensional CT (3D-CT) images were reconstructed for a precise evaluation of the pulmonary vessels and bronchi (Fig. 2A, B). Intraoperatively, we established that the location of the superior vena cava and azygos vein mirrored the normal distribution, and that the aortic arch was absent from the left thoracic cavity. The left lung was well lobulated into three lobes and the anatomy of the left side of the pulmonary artery had features typical of the normal right side (Fig. 3A, B).

The nodule was located in the left upper lobe (S2), and we removed it by wedge resection. The frozen section revealed the proliferation of malignant cells, suggesting adenocarcinoma. Therefore, we performed left upper lobectomy with lymph node dissection (ND2a-1) using VATS (3 ports). We did not observe any malformations other than the mirror image of the organs. The operation took 3 hours and 15 minutes to complete, with minimal, intraoperative blood loss. The final pathological diagnosis was adenocarcinoma, p-T1b (12 × 12 mm) N0M0, p-
Stage IA2 (Union for International Cancer Control, 8th ed.). The patient’s postoperative course was uneventful, and she was discharged from our hospital on postoperative day 9.

Discussion

Situs inversus totalis (SIT) is an uncommon condition in which the heart and other organs of the body are transposed through the sagittal plane to lie on the opposite side from the normal position. 20–25% of this condition is associated with Kartagener’s syndrome [3]. A relationship between lung cancer and SIT has not yet been elucidated, and medical reports of lung cancer patients with SIT are extremely rare; only 16 surgical cases (13 thoracotomies and 3 VATS) have been reported so far.

SIT itself has no pathological significance, but this condition requires careful attention during surgery. In SIT, not only are the organs reversed sagittally, but there may also be some variations in the courses of the pulmonary vessels and bronchi. Therefore, it is essential for surgeons to take extreme care in identifying and handling the vascular and bronchial branches of the contralateral thoracic cavity [4]. Wójcik et al reported that “all authors confirmed the consistency of contrast-enhanced CT for detecting the anatomical conditions observed during the operation” [5]. 3D-CT is useful to understand the mirror-imaged anatomy three-dimensionally and to prevent the risk of vascular and bronchial injury.

Unlike pulmonary vessels and bronchi, neurological structures are more difficult to capture by preoperative imaging, and this difficulty is compounded by the fact that we rarely encounter patients with SIT. Because the location of the left recurrent laryngeal nerve was unclear at the beginning of the present operation, the upper mediastinal lymph node dissection had to be performed very carefully so as not to injure the recurrent laryngeal nerves. These precautions probably ensured the uneventful postoperative course.

The present patient was free of Kartagener’s syndrome, but if that condition were to exist, the case could be complicated by intrathoracic adhesions and an irregular and expanded bronchial artery due to chronic and repeated infections [6]. Surgeons should consider these possibilities and make appropriate preoperative preparations.

Conclusion

In conclusion, we present an extremely rare case of thoracoscopic lung lobectomy for a lung cancer patient with SIT. Cautious preoperative anatomical evaluation and perioperative handling are essential to prevent intraoperative injuries and complications in cases involving SIT.
Conflicts of Interest

The authors declare no conflicts of interest.

References


完全内臓逆位に合併した肺癌に対する胸腔鏡下肺葉切除術

金山 雅俊1, 井上 政昭1, 吉田 順一1, 田中 文啓2

1下関市立市民病院 呼吸器外科
2産業医科大学 医学部 第2外科学講座

要   旨: 完全内臓逆位は, 各臓器が左右逆転し正常位に対して鏡面像と位置関係にあるものをいい, 頻度は10,000人に1〜2人と稀にみられる常染色体劣性遺伝の破格である. 今回, 完全内臓逆位に合併した原発性肺癌に対して, 胸腔鏡下手術を施行した報告をする.

症例は61歳女性で, 胸部X線写真で左上肺野に異常陰影を指摘され紹介となる. 胸腹部CTにて胸腹部内臓の完全逆位に加え, 左肺上葉S2に葉間胸膜に接する25 × 12 mmの結節影を認めた. 術前検査にて左上葉肺癌疑いc-T1bN0M0 Stage IAとして手術を行う方針とした. 術前3D構築画像にて鏡面像を呈した肺動静脈および気管支の立体的構造を把握した. 手術は腫瘍を胸腔鏡下に部分切除を行い, 迅速病理検査で腺癌と診断されたため完全胸腔鏡下に左上葉切除およびリンパ節郭清を施行した. 左縦隔に下行大動脈は認めず, 左肺は上中下葉の3葉に分葉していた. 左肺鏡面像であったが, 正常解剖の際の手技. 手順で施行可能であった. 完全内臓逆位の手術においては, つねに反対側の胸腔をイメージしながらの手術となる. そのため術前に肺血管, 気管支の解剖学的評価を十分に行った上での慎重な手術操作が必要となる.

キーワード: 胸腔鏡下肺葉切除, 肺癌, 完全内臓逆位.