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Non-small cell lung cancer in patient with visceral total inversion

Abstract

This article reports a case of non-small cell lung cancer in a 74-year-old man with visceral total inversion. The epidemiology and main anatomical differences present in this rare syndrome, as well as basic information on lung cancer, are explored. We present diagnostic procedures and their results and describe the surgical technique of lung cancer treatment performed in this rare case. The perioperative period and the histopathological findings are analysed. Finally, references to similar cases found in worldwide literature are discussed.

Key words: *situs inversus*, lung cancer, bilobectomy

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Introduction

Situs inversus is a congenital condition, with organs of the chest and abdomen being arranged in the exact mirror image reversal of the normal positioning.

Normal human development results in the asymmetrical positioning of the organs both in the chest and abdominal cavity. Typically, the heart is placed on the left (*sinistrocardia* or *levocardia*), liver on the right, left lung with two lobes, while the right lung has three. Such a positioning is known as *situs solidus*. Mirror image reversal of organ arrangement occurs in around 1 in 8500 living newborns, with a similar frequency in boys and girls [1–3]. Chest and abdomen organs are in the exact opposite position — heart in the right (*dextrocardia*) as well as two-lobe lung and spleen, while the liver and three-lobe part of lung are on the left side. Despite this condition, functional relationship is not altered and clinical problems are infrequent. Heart defects are more common in this population (3–5% in comparison to less than 1%), especially transposition of great arteries [4]

(TGA). In approximately 25% of individuals with *situs inversus*, syndrome of primary ciliary dyskinesia known also as Kartagener's Syndrome occurs [5–8].

Situs inversus is classified as the transposition of organs with dextrocardia or levocardia. This classification is based on the heart apex position. In this condition levocardia is observed infrequently (1 in 22 000) and is almost always accompanied by the heart defect. Other classifications include complete and incomplete *situs inversus* [6, 9].

Except for the complete *situs inversus*, additional types of this abnormality, known as *situs ambiguous*, have been identified. Two of them are of particular interest: asplenia syndrome (right isomerism) and polysplenia syndrome (left isomerism). In asplenia syndrome, heart atria are bilaterally right-sided and centrally positioned; symmetric liver, lack of spleen and bilaterally trilobed lungs are observed, with both descending aorta and vena cava inferior at the same side of the spine. Conversely, in polysplenia syndrome, bilaterally morphologically left heart atria, multiple spleens, both lungs bilobed and vena cava often joint with azygos and hemiazygos are present [3].

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Organ misplacement might present a diagnostic challenge with a necessity to adequately assess the position and relationship between the organs, heart atria and ventricles, aortic arch and its branches, pulmonary vessels, vena cava superior and inferior, coronary sinus, and hepatic veins. For imaging of the inappropriate organ placement, X-ray and ultrasonographic diagnostics are usually sufficient, but computed tomography is preferred in the case of full *situs inversus*, so as to allow the precise visualisation of anatomical position of the organs, heart apex and branches of large vessels.

Diagnosis of *situs inversus* is especially important before planned thoracic or abdominal surgery. Special attention should be focused on the appropriate image side labelling by radiologists, both for classic X-rays and computed tomography.

Case report

A 74-year-old patient, a long-term smoker (50 pack-years), with a previously diagnosed total *situs inversus*, generalised atherosclerosis, arterial hypertension and a history of ischemic stroke in 2006, was admitted to the Clinical Department of Thoracic Surgery of Pomeranian Medical University in Szczecin, Poland for surgical treatment of bronchogenic non-small cell left lung cancer.

Symptoms of cough, weakness and fatigue were first observed in January 2007, with no significant improvement after treatment by the general practitioner. On chest X-ray (performed in August 2007) a lesion in the lower part of the left pulmonary hilus was observed, resulting in patient referral for further diagnostics to the Department of Pulmonology in Gorzów Wielkopolski, Poland. Bronchofiberscopy revealed a tumour lesion in the bronchi of the left lower lobe, with histopathological specimens collected by the brush biopsy. No metastatic lesions were visualised in further diagnostics (abdominal and supraclavial ultrasound scans, computed tomography of the abdomen). The patient was qualified for the surgical resection.

On admission to our department, balance disturbances and vertigo were observed. Physical examination revealed emphysematous features of the chest, bilateral vesicular murmur with prolonged exhalation phase, apex beat in the right 5th intercostal space, normal heart sounds on auscultation but in the exact mirror image reversal position, and psychomotoric retardation. No neurological disturbances, after the prior stroke, were observed.

In the routine chest X-ray performed in the P-A projection, dextrocardia, right sided aortic arch, higher position of the left diaphragm dome and

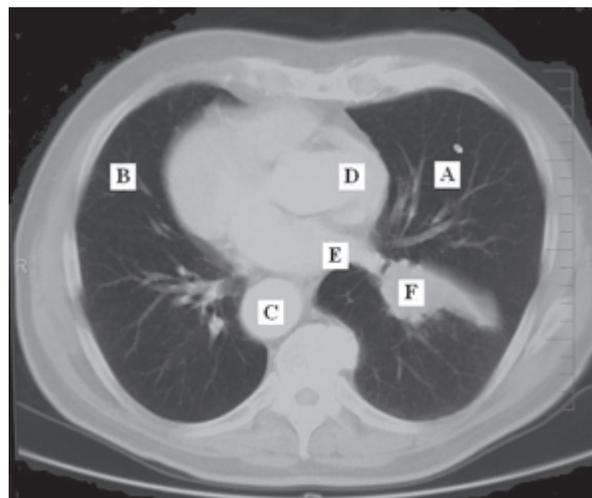


Figure 1. Tumor and its relation to surrounding structures on CT scan (A — left lung, B — right lung, C — descending aorta, D — ascending aorta, E — left pulmonary artery, F — tumor)

tumour overgrown lower part of the left lung hilum were visualised. In the lateral projection, the lesion was projecting onto the pulmonary hilum region. Bronchofiberscopy confirmed the image of bronchial tree reflecting complete *situs inversus* positioning (short mainstream bronchus on the left side, with branching left upper lobe bronchus, bronchus intermedius, and presence of middle lobe bronchus). Tumour was observed in the left lower lobe orifice. Non-small cell cancer was diagnosed histologically. *Situs inversus* in the chest was observed in CT scans as well (Fig. 1), with right-side positioning of the heart and aortic arch, left sided azygos and hemiazygos veins, three-lobed left lung and bilobed right one; additionally tumour with a diameter of 30 mm, causing partial atelectasis in the 9th segment of the left lung. Left inferior paratracheal lymph nodes of the threshold size were described.

Complete *situs inversus* was described again, with no abnormality of internal organs and retroperitoneal space detected in abdominal ultrasound scans. Routine supraclavicular ultrasound scans did not reveal any enlarged lymph nodes. Basic laboratory parameters remained within normal ranges. Because of the medical history, both internal disease and neurological consultations were requested, to exclude other contraindications for the planned surgery.

After consultations of clinical case and laboratory data analysis, the patient was finally qualified for the surgical treatment. The procedure was performed in the Department of Thoracic Surgery, Pomeranian Medical University, Szczecin, Poland. By left-sided posterolateral thoracotomy through the 5th intercostal space, the chest cavity was open.

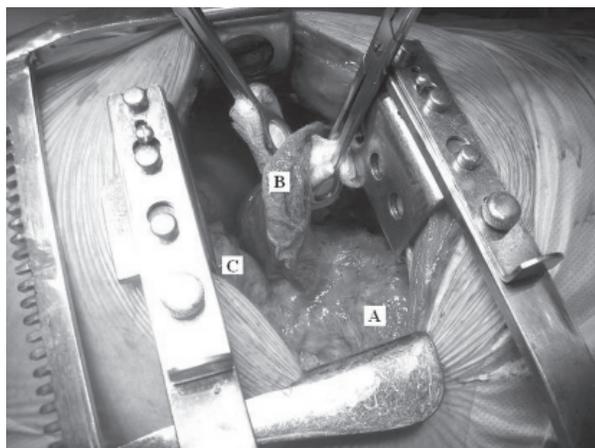


Figure 2. View of the left lung after thoracotomy (A — upper lobe, B — middle lobe, C — lower lobe)

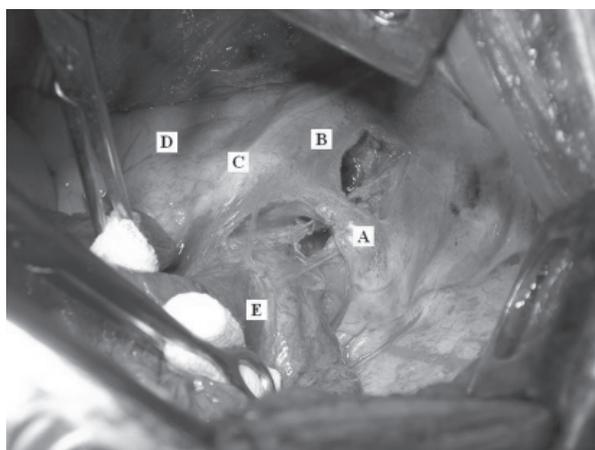


Figure 3. View after completion of the inferior bilobectomy and lymphadenectomy (A — arch of the azygos vein, B — superior vena cava, C — phrenic nerve, D — pericardial fat, E — remaining left upper lobe)

Three lobed lung was visualised in the left pleural space (Fig. 2). Complete *situs inversus* was revealed, with left lung and its bronchi, pulmonary vessels and hilum anatomy being the exact mirror image reversal of the normal right lung. The heart was placed on the right, with apex directed to the right side. Superior vena cava and azygos vein were positioned on the left, with transposition of the thoracic aorta. In the lower lobe of the left lung, in the area of hilum, the tumour, sized 3 × 5 cm, was present, infiltrating the middle lobe hilum. The operating surgeon decided to remove both middle and lower lobe of the lung, with the following groups of lymph nodes: lower paratracheal left and right, subcarinal, from the area of the left pulmonary ligament and left pulmonary hilum. Bronchial stump was closed by prolene 3-0 suture by Klingenberg, and was covered by the pedicle flap of parietal pleura (Fig. 3).

No complications were observed in the postoperative period. Reverse positioning confirming the *situs inversus* was confirmed by pathologist. A tumour sized 34 × 37 × 46 mm was situated in the peri-hilar region and surrounded by pulmonary tissue. Within the tumour both fibrosis and inflammatory infiltration were described, with necrosis of approximately 80% of the total transverse diameter of the lesion. Final histopathological diagnosis, grading and staging were: squamous cell lung cancer G1, stage pT2N0M0/IB.

The patient was discharged from the department with the planned follow-up in the Pulmonary Disease Outpatient Clinic, Szczecin, Poland and bronchofiberoptic reassessment three months after the surgery.

Discussion

Bronchogenic cancer is the most common malignant neoplasm among males with 15 248 diagnoses and 1652 deaths in Poland (according to the Polish National Cancer Registry 2005). Key factors promoting development of the cancer include carcinogenic substances present in tobacco smoke. Sadly, at the diagnosis, in 75–80% of patients contraindications to surgical treatment are present. These include advanced stage of the cancer, history of concomitant general diseases, and poor results of the functional respiratory tests. Such a clinical and pathophysiological picture is related to the long asymptomatic phase of the disease and the fact that as the most prevalent symptoms (coughing and dyspnoea) are common among tobacco smokers, these patients are not inclined to consult the physician. Routine chest X-ray and chest computed tomography are still of primary importance here, being the introduction to the more advanced diagnostics in the described case [10].

Standard surgical treatment in case of lung cancer recommends the anatomical resection of pulmonary tissue. In the presented case, bilobectomy was performed; in normal anatomical positioning such a procedure is performed on the right side [11].

In this report we reviewed the case of the operation treatment of the bronchogenic cancer in the patient with complete *situs inversus*. Because of the rarity of the syndrome and different (contralateral) manifestation of certain diseases with typical symptoms linked to the side of the body (e.g. appendicitis, cholecystitis and cholestatic diseases), the syndrome might pose a diagnostic challenge.

In the presented case, congenitally present complete *situs inversus* was diagnosed during previous hospitalisations. Diagnostic procedures per-

formed in our department aimed at confirming the complete exact mirror image reversal of the normal positioning. Additionally, it was necessary to confirm if *situs inversus* was accompanied by any other structural or functional abnormalities, especially in the anatomy of the circulatory system including pulmonary vessels and the bronchial tree, which might directly influence the surgical procedure and adaptation in the perioperative period.

Medical reports on lung cancer treatment among patients with complete *situs inversus* are uncommon, with only a few casuistic cases published so far. Thompson et al. were first to describe a case of a 58-year-old man with pre-diagnosed complete *situs inversus* and post-mortem diagnosis of right upper lobe cancer with liver and 6th rib metastases. In autopsy, *situs inversus* was found with two-lobed right lung and three-lobed left one [12].

Surgical treatment of the lung cancer in a patient with organ positioning reversal was described by Kodama, Doi and Tatsuta. The case of a 68-year-old man was presented with the initially BAL-diagnosed squamous cell cancer of the left middle lobe. The authors performed the left middle lobectomy with additional excision of S7 and S8 segments and resection of the cancer-infiltrating 6th and 7th rib. No other anatomical abnormality, except for *situs inversus*, was detected in this case either [13].

In the report by Subotich et al. from 2006, radical treatment of the adenocarcinoma of the left lung cancer was described. Left upper lobectomy was performed in this case, with anatomical situation as seen normally on the right side of the body. For more adequate pre-surgical assessment, the authors additionally performed contrast-enhanced radiological scanning of the pulmonary vessels, aorta and its branches, with no deviation from "normal" functional characteristics for *situs inversus* syndrome [14].

During review of the widely available medical reports and data, no case report on the surgical

removal of two lung lobes in a patient with *situs inversus* syndrome was found. Upper or lower bilobectomy, as mentioned above, is the procedure normally performed on the right side only. In the described case, due to the anatomical conditions, the surgery was performed on the left. Two cases described above as well as the case presented by us are the reports with the tumour excision performed on the left side. No case of right-side lung cancer related surgery was found to have been published so far.

In conclusion, we stress the importance of precise physical patient examination and image data review for diagnosis of this rare anatomical condition.

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