Case Report

Case Report-A Rare Congenital Lung Defect

Christos Simoglou^{1*}; Igner Bertus²

¹Department of Thoracic Surgery, Red Cross General Hospital, Athens, Greece

²Department of Cardiothoracic Surgery Universital Hospital of Bristol, UK

*Corresponding author: Christos Simoglou

Department of Thoracic Surgery, Red Cross General Hospital, Athens, Greece.

Email: simoglouchristosdoctor@yahoo.com

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Abstract

A 72 year old male smoker presented with haemoptysis and recurrent pneumonias. Chest CT showed an emphysematous cyst and air-fluid level cavities in the left lower lobe, (Figure 1). A left lower lobectomy was performed. Intraoperative finding was an intralobar sequestration. Histopathology revealed the presence of adenocarcinoma within the sequestrated lobe.

Keywords: Complication; Pulmonary infection; Thoracoscopic procedures

Introduction

Pulmonary sequestration is a rare congenital lung defect in which nonfunctioning lung tissue is separated from the normal tracheobronchial tree and receives vascular blood supply from an aberrant systemic artery.

While it is not in itself a life-threatening condition, a pulmonary sequestration can cause health complications including cardiovascular problems, long-term infections like tuberculosis and bronchial cancer. It could be fatal if blood vessels in the begin to hemorrhage.

Case Report

We report a case of a 72 year old male patient, smoker (40 pack years), with history of haemoptysis and recurrent pneumonias. CT of the thorax showed an emphysematous cyst and multiple air-fluid level cavities in the left lower lobe, (Figure 1). A left lower lobectomy was decided due to the patient's history and extent of damage of the lobe. The intraoperative finding was an intralobar sequestration with an aberrant artery arising from the descending thoracic aorta. The histopathological examination revealed a tumor with a size of 2.7cm within the sequestrated lung tissue, which was found to be adenocarcinoma. As the tumor was not diagnosed in the preoperative CT scan, a work-up was done post-operatively. The abdominal CT showed a lymph node of 5,1cm in size around the left renal artery. The tumor was staged as stage IV (pT₁N₀M₁) and the patient received adjuvant chemotherapy. After three cycles of chemotherapy, the size of the lymph node decreased only to 3.7cm. The radiology scan also revealed infiltration of the left hemithorax and metastasis to the thoracic spine. The patient was sent for radiotherapy treatment, during which a rapid deterioration of his clinical condition occurred and finally he succumbed to his ailment 27 months after the thoracic procedure.

Discussion

The most common and recommended treatment is surgical removal of the pulmonary sequestration. Extrapulmonary growths can be removed without the loss of any normal lung tissue [1,2].

Pulmonary sequestration is a rare congenital anomaly in which nonfunctioning lung tissue is separated from normal tracheobronchial tree and is supplied by an aberrant systemic artery [3,4]. Two types of pulmonary sequestration exist: Extra Lobular Sequestration (ELS) which has its own pleural covering, and Intralobular Sequestration (ILS) which shares common pleura with the normal lung tissue [5]. Intralobular sequestration is the most common form and the sequestrated lobe usually receives blood supply from the thoracic aorta (74%).



Figure 1 Chest CT: An emphysematous cyst and air-fluid level cavities in the left lower lobe.

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Patients often present with recurrent bronchitis, pneumonia or heamoptysis. The diagnosis of pulmonary sequestration requires a high index of suspicion since in many cases the diagnosis is made intraoperatively [6].

Lung cancer associated with pulmonary sequestration is rare. Among these, The treatment of associated lung cancer with pulmonary sequestration is surgery after a thorough work up and proper staging of patient has been performed. In case of accidental finding, a post-operative work up needs to be done to stage the tumor and where necessary additional treatment (chemotherapy, radiotherapy or both), should be undergone [7].

Cases of carcinoma and lung sequestration are rare and even rarer are cases of lung cancer in pulmonary sequestration, as reported in the literature. Despite that, we suggest that frozen section biopsy should be done in every patient undergoing lung resection for suspected or diagnosed pulmonary sequestration to exclude any other alternative diagnosis [8]. The rarity of cancer and pulmonary sequestration co-existence does not justify a complete preoperative staging for all patients, but we suggest it could be optimal for surgeons managing such patients.

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