

Surgery for Primary Giant Acinic Cell Carcinoma of Right Lung with Severe Mediastinal Deviation: A Case Report

Xueyu Chen

Ruijin Hospital Shanghai Jiaotong University School of Medicine <https://orcid.org/0000-0002-3940-4921>

Nengchong Zhang

Ruijin hospital Shanghai jiaotong university School of Medicine

Fangxiu Luo

Ruijin Hospital, Shanghai jiaotong University School of Medicine

Lianggang Zhu (✉ rjhnwx@163.com)

Ruijin Hospital Shanghai Jiaotong University School of Medicine

Case report

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Abstract

Background: Primary lung acinic cell carcinoma is very rare. Here we report a young female patient who suffered the largest primary lung acinic cell carcinoma and severe mediastinal deviation at the same time which has never been reported before.

Case presentation: A 27-years-old married female patient presented with recurrent coughing and hemoptysis for more than ten years came to our hospital. A chest computed tomography was performed in our hospital and showed a giant space-occupying lesion in the hilum of right lung. After a thorough and detailed preoperative examination, the patient then was performed a radical right pneumonectomy and mediastinal lymph node dissection. The specimens and pathology showed that the size of the tumor was about 8.6×4.5×4.4cm. The morphology of the tumor cell under microscope demonstrate a primary acinic cell carcinoma of right lung. The immunohistochemistry of the tumor showed AE1/AE3(+), Ki-67(2% +), CK7(+), Calponin(+/-), Vimentin(+), CK19(+), a-ACT(+), PAS(partial+). The patient was discharged two weeks after the operation. So far, the patient has been followed-up for one year, all the clinical test indexes are normal and no evidence of tumor recurrence or metastasis was observed.

Conclusions: The primary acinic cell carcinoma of lung in this case is the biggest one ever been reported. The patient also had a very rare condition of severe mediastinal deviation. After surgical treatment, the patient recovered uneventfully and had stable disease without recurrence after one year of follow-up. This case indicates that acinic cell carcinoma of lung is of low malignancy, the prognosis and therapy effect of surgical treatment are satisfied.

Background

Acinic cell carcinoma (ACC) is a very rare epithelial malignant tumor of salivary gland, accounting for 10%-17% of all malignant salivary gland tumors [1]. It was initially proposed as an independent type of salivary gland tumor by Foote and Frazell in 1953 [2]. In addition to salivary gland, acinic cell carcinoma also has been observed in lung, breast and other organs. Among them, primary lung acinic cell carcinoma is even rare [3] and was firstly reported by Fechner in 1972 [4]. So far, there were only 25 cases of primary lung acinic cell carcinoma have been reported in literature [12]. Here we report a case that the patient suffered a giant acinic cell carcinoma in upper lobe of right lung with severe mediastinal deviation and the patient was treated successfully by surgery in our thoracic surgery department.

Case Presentation

A 27-years-old married female patient presented with recurrent coughing and hemoptysis for more than ten years first came to our hospital in January 2019. According to the medical history information provided, the patient was previously diagnosed as pneumonia and treated symptomatically in local clinical institution. The symptoms of patient were once relieved after the treatment, but recurrent frequently. The patient has no family history of primary lung malignancy and genetic disease and was

155 cm in height and 42 kg in weight with a lean body shape, symmetrical thorax without deformity. The trachea of the patient deviated to the right slightly and there was no obvious rhonchi and moist rale as well as wheezing rale were heard. The heart rhythm was regular, the auscultation area of heart sound deviated to the right thorax obviously and no obvious abnormality was found in abdominal physical examination.

A chest computed tomography was performed for the patient and showed a giant abnormal space-occupying lesion in the hilum of right lung. The tumor was closely related to the right pulmonary artery and bronchus (Fig. 1). The right lung was atelectasis and some of the left lung as well as heart were obviously deviated to the right thoracic cavity. A Chest MRI showed a mass abnormal signal tumor in the region of right hilum about 10.0 × 4.5 cm in size. The T1WI was isointense, while T2WI and DWI were both hyperintense. A 3D reconstruction of the hilar structures have shown a complete anatomical disorganization of right pulmonary artery and vein (Fig. 2). A neoplasm with smooth surface was observed in the right main bronchus by bronchoscopy examination, but a biopsy was not performed. Pulmonary function showed a severe obstructive mixed ventilation dysfunction. The forced expiratory volume in 1 second (FEV1) was 1.36L, accounting for 45% of the predicted value, and maximal voluntary ventilation (MVV) was 27.63L, accounting for 43% of the predicted value. The renal function, electrolyte, coagulation function, arterial blood gas analysis were all normal. Because of the continuous hemoptysis for quite a long time, the patient's blood routine examination showed a moderate anemia and hemoglobin was 69 g/L, biochemical examination showed moderate malnutrition and prealbumin was only 85 g/L, albumin was 31 g/L. In this patient, CA125 was 434.77 u/ml, CEA was 12.43 ng/ml, CA724 was 233.3 u/ml, while CA242, CA199, AFP, SCCA, NSE were all normal. No sign of abnormality or metastasis was found in enhanced MRI of brain and bone scan. No abnormal and enlarged lymph nodes were observed in neck and supraclavicular region by ultrasound examination.

We diagnosed the patient with a huge tumor in the right lung and firstly considered a special type of malignancy. In view of the patient's personal willingness and equipment limitations, a preoperative biopsy of the tumor was not performed. After discussion with a multidisciplinary team which included thoracic surgeons, radiologists, and respiratory physicians, we decided to perform a surgical treatment for the patient with her permission.

A thoracotomy was performed under general anesthesia with left lung ventilation. The patient was in left 90° lateral position, a 20 centimeters long posterolateral incision at the 5th intercostal space was made overlying the right chest wall. Surgical exploration revealed a complete atelectasis of the right lung with obvious consolidation in lung tissue. The lingual segment and anterior segment of left upper lobe herniated into right upper thoracic cavity while the heart of patient also deviated into right lower thoracic cavity obviously. The lymph nodes of each group in mediastinum were checked in the operation and no significantly enlarged lymph nodes were observed. Because of the size and invasion of the tumor, a right pneumonectomy and mediastinal lymph nodes dissection was initially considered before surgery. The right main bronchus was significantly thicker than normal and the outer diameter of the right main bronchus was 2.4 cm. The bronchial arteries around the bronchus were extremely twisted and dilated.

The upper lobe of the right lung was completely consolidated into a mass, the middle and lower lobes of the right lung were atelectatic. Subsequently, we performed a right pneumonectomy for this patient.

We checked the surgical specimens and found that the tumor originated from the right main bronchus, about 1 cm away from the tracheal carina, grew distally along the lumen of the bronchus and completely blocked the lumen. The tumor was about 8.6 × 4.5 × 4.4 cm in size (Fig. 3) and pathological diagnosis showed a right main bronchus malignant tumor. The morphology of tumor cells under microscope demonstrated a primary acinar cell carcinoma of right lung. Three lymph nodes (LNs) in group 2 and 4, two LNs in group 7, three LNs in group 9 and one LN in group 10 were dissected and no tumor metastasis was found in the above lymph nodes. Immunohistochemistry of the tumor showed AE1/AE3(+), Ki-67(2%+), K7(+), Calponin(+/-), Vimentin(+), CK19(+), a-ACT(+), PAS(partial+). While CD56, S-100, P63, TTF-1, CDX2, CK5/6, SYN, Dog-1, SOX-10, Mammaglobin were all negative. (Fig. 4).

The patient complained of dyspnea on the second day after operation. Some significant inspiratory wheezing sounds were heard on retrosternal auscultation. The patient had no previous history of asthma and was unresponsive to treatment with bronchodilator. A chest computed tomography then was performed and we were unexpected to find that the heart and mediastinum further deviated into the right thoracic cavity after right pneumonectomy. Because of the gravity of the heart, when the patient lay flat the left main bronchus was clamped between the mediastinum and the thoracic vertebra which resulting in an obvious compression on the left main bronchus (Fig. 5). The narrowest region of airway stenosis of the left main bronchus was only 3 mm.

Then we tried to turn the patient to left semi-prone and prone position to relieve the compression of the left main bronchus, and the patient's symptoms were partially relieved after changing body position. So we believe that some methods should be taken to reposit the patient's heart and mediastinum back to the left which may help to alleviate the patient's symptoms. After 600 ml of air was injected into the right thoracic cavity of the patient through drainage then clamped drainage tube, the symptom of the patient was partially relieved which effect is similar to the changing of body position. Over the next few days, we kept the patient's drainage clamped so that exudate and gas in the right thoracic cavity could alleviate the deviation of mediastinum. After clamping the drainage several days and confirming the symptoms did not recur, we removed the drainage on the 8th day after surgery and the patient discharged on the 14th day postoperatively.

At present, the patient has been followed up for 12 months after surgery. and there has been no evidence of recurrence and metastasis observed. All the clinical indexes and physical condition of the patient are normal.

Discussion And Conclusions

Primary acinar cell carcinoma of the lung is extremely rare, and so far very few cases have been reported worldwide. The pathogenesis of primary acinar cell carcinoma still remains unclear. Florian Haller believes that the pathogenesis of the disease may be related to the upregulation of NR4A3 [5]. Primary lung acinar

cell carcinoma is a kind of low-grade malignant tumor that usually presents as an isolated tumor adjacent to or close to the bronchus with little lymph node metastasis[6–7]. Generally, the surface of tumor is covered with normal tracheal mucosa, it is difficult to get cytological diagnosis by routine bronchoscopy brush examination and biopsy[8]. A postoperative pathological examination is critically needed to confirm the diagnosis. Meanwhile, Kazuo Watanabe[9], a Japanese scholar, believes that transbronchoscopic fine-needle aspiration (FNA) may help to obtain samples of tumor tissue to make a definite pathological diagnosis of the disease preoperatively. Primary lung acinic cell carcinoma can occur from juvenile to old age and it is most common in the 30–75 years-old population (median age 49.5 years), but there are also two cases reported in young girls[10]. The largest tumor size of primary acinic cell carcinoma ever reported is 5.2 cm [11], while the size of the tumor in this case is 8.6 cm, which is larger than that. At the same time, it is also rare for the patient in this case have a herniation of compensatory left lung expansion and heart into the right thoracic cavity. The clinical symptoms and signs of this kind of patients are lack of specificity. It often manifested as cough, expectoration, hemoptysis, dyspnea, wheezing. Therefore, primary lung acinic cell carcinoma was often misdiagnosed before operation. In some cases patients have a history of inhaling foreign bodies and the incidence of primary lung acinic cell carcinoma of right lung is higher than left. Thus, some scholars speculate that the occurrence of primary lung acinic cell carcinoma may be related to long-term chronic stimulation of inhaling foreign bodies[12].

In this case, the patient has an aggravated hemoptysis, and the chest CT examination had showed a huge neoplasm in the right lung. These symptoms and examination results had constituted an adequate indication for surgery. At the same time, we fully communicated with the patient and solicited her own willingness, so a percutaneous lung biopsy was not performed before operation.

Surgical resection of the tumor is the main therapy to treat primary lung acinic cell carcinoma[13]. During the operation, the lung function should be preserved as much as possible on the premise of ensuring the complete resection of the tumor. The mediastinal and hilar lymph node metastasis rate of patients with primary lung acinic carcinoma is relatively low, so it is unnecessary for adjuvant chemotherapy after operation in N0 case. However, some studies have shown that the expression of Ki67 antigen is related to the recurrence of the disease. The tumor is easy to recur when $Ki67 > 10\%$, and mediastinal lymph nodes metastasis can also be found in patient with tumor recurrence. Therefore, we performed systematic lymph node dissection in the operation. For patients whose tumor can not be completely resected, or patients who are physically weak, have poor cardiopulmonary function and unable to tolerate operation, a radiation therapy for tumors is also considered as an alternative treatment.

Primary lung acinic cell carcinoma is generally considered as a low-grade malignant tumor, with low recurrence, metastasis and mortality after operation. At present, the patient in this case has been followed up for one year after operation, no obvious sign of tumor recurrence and metastasis has been observed. All the clinical test indexes and physical condition of the patient are normal. With the regression of local edema in left bronchus and the adaptation of the body, the patient had no complaints of discomfort and

did not suffer dyspnea and wheezing any more. This case indicates that the prognosis and therapy effect of surgical treatment for primary lung acinic cell carcinoma are satisfied.

Abbreviations

ACC: Acinic cell carcinoma

FNA: Fine-needle aspiration

LNs: Lymph nodes

FEV1: Forced expiratory volume in 1 second

MVV: Maximal voluntary ventilation

CEA: Carcinoembryonic antigen

AFP: Alpha-fetoprotein

NSE: Neuron-specific enolase

SCCA: Squamous carcinoma cells antigen

Declarations

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Authors' contributions

Each author has made an important scientific contribution to the surgical procedure and has assisted with the drafting or revising of the manuscript. The author(s) read and approved the final manuscript.

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Availability of data and materials

All data generated or analyzed during this manuscript are included in this published article.

Consent for publication

All the authors have Consented to publish this manuscript.

Ethics approval and consent to participate

Ethical approval was given by the Ruijin Hospital, Shanghai Jiaotong University School of Medicine.

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Figures

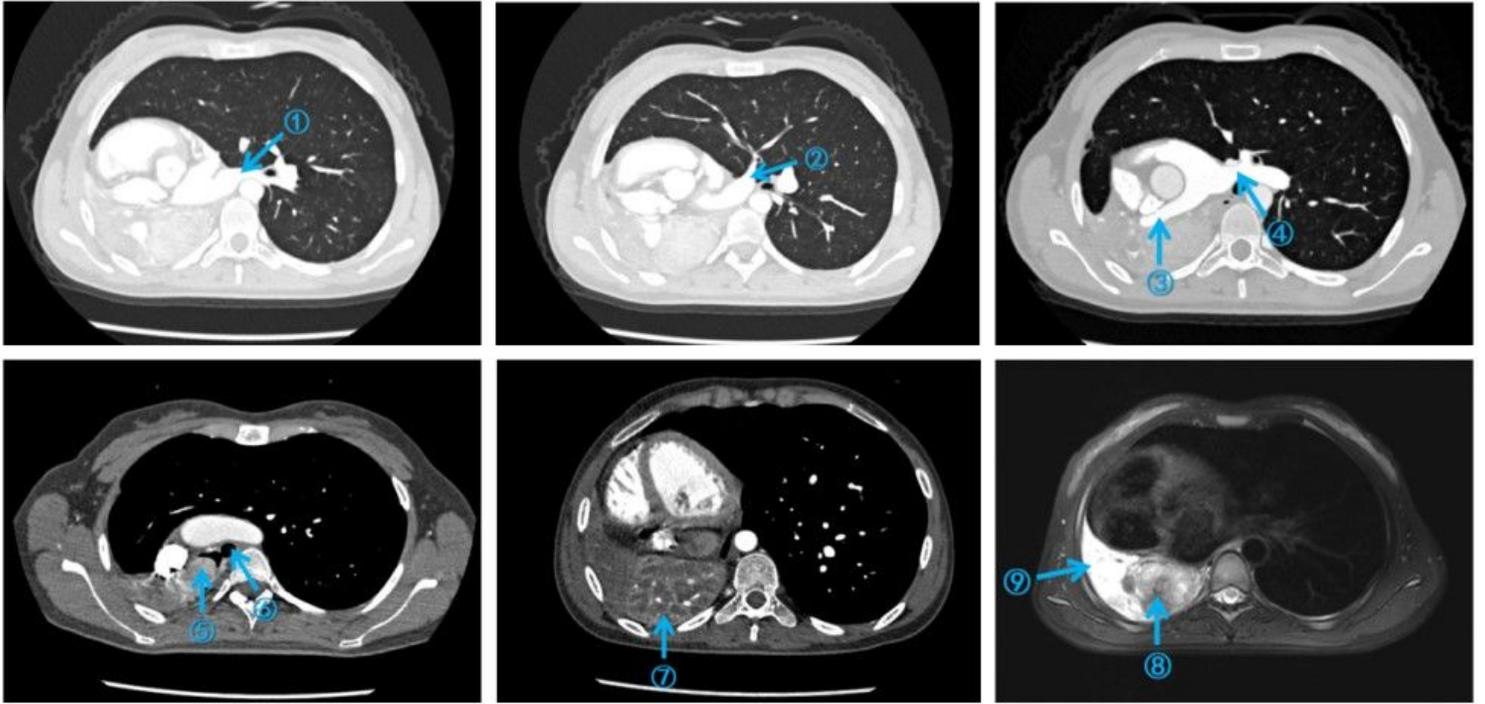


Figure 1

Enhanced CT and enhanced MRI images of the patient ☐ left inferior pulmonary vein; ☐ left superior pulmonary vein; ☐ left pulmonary artery; ☐ right pulmonary artery; ☐ tumor in right main bronchus; ☐ left main bronchus; ☐ tumor in right lung (enhanced CT); ☐ tumor in right lung(enhanced MRI); ☐ atelectasis of right lung

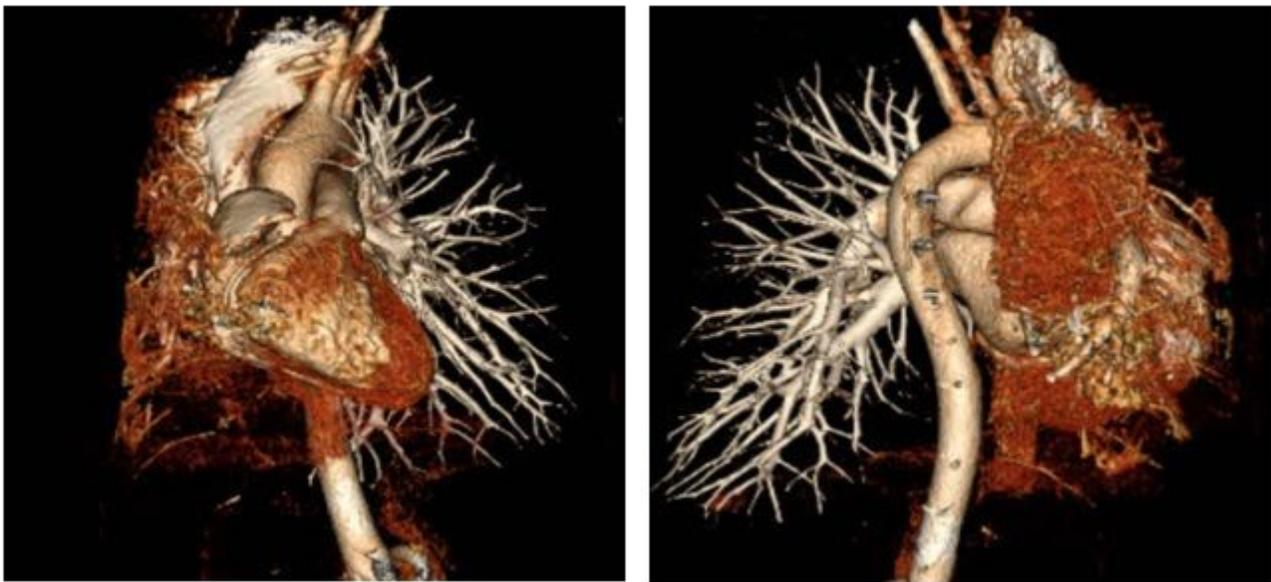


Figure 2

3D reconstruction of the patient's right hilar structures The vascular anatomical structure of the right hilum is completely disorganized and difficult to recognize

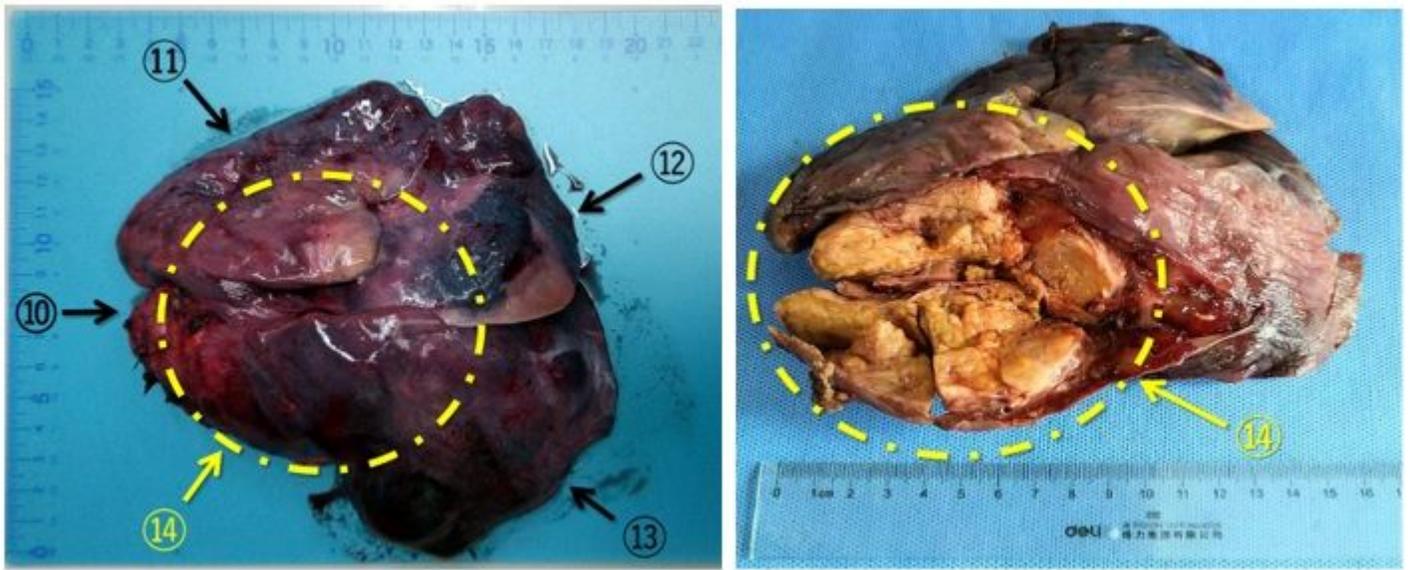


Figure 3

Surgical specimens of right lung ☒ margin of right main bronchus; ☒ right upper lobe; ☒ right middle lobe; ☒ right lower lobe; ☒ tumor tissue

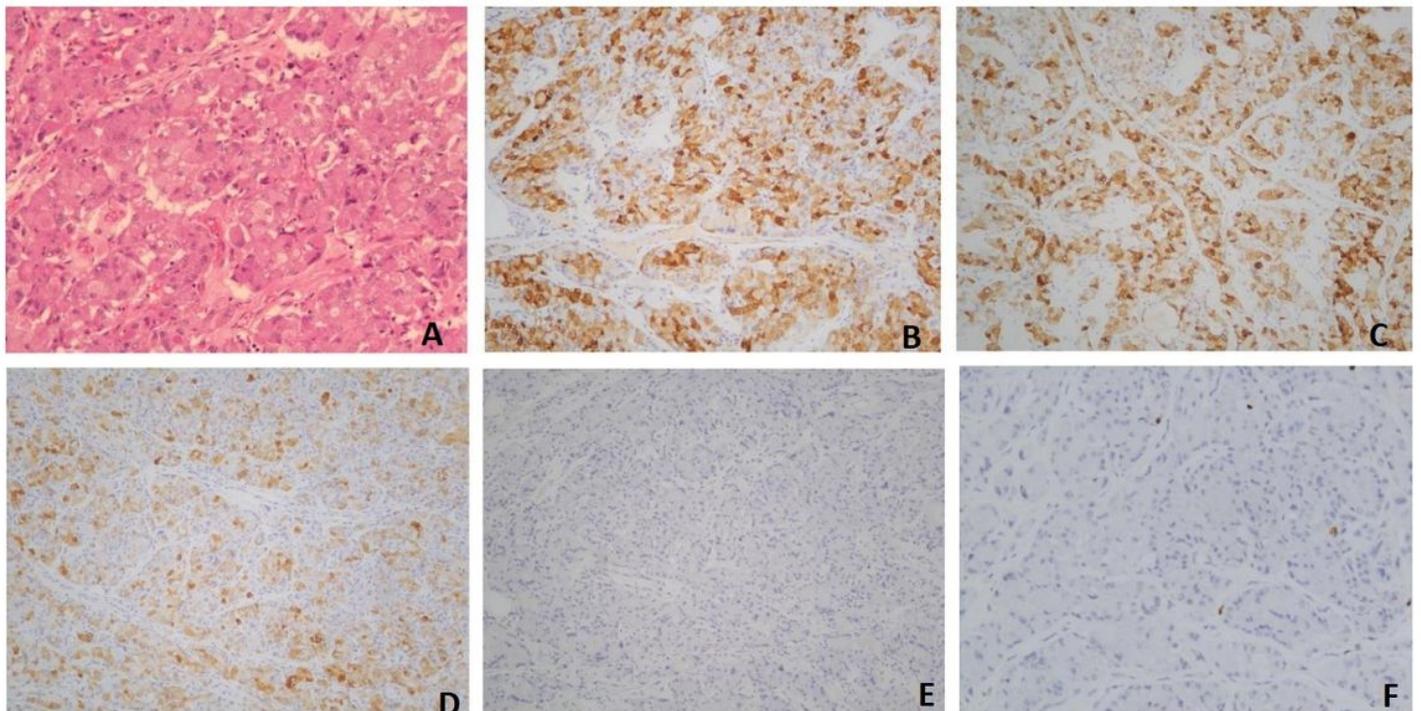


Figure 4

Immunohistochemistry of the tumour A: He staining; B: AE1 / AE3 (+); C: CK7 (+); D: CK19 (+); E: CK5 / 6 (-); F: Ki-67 2%+

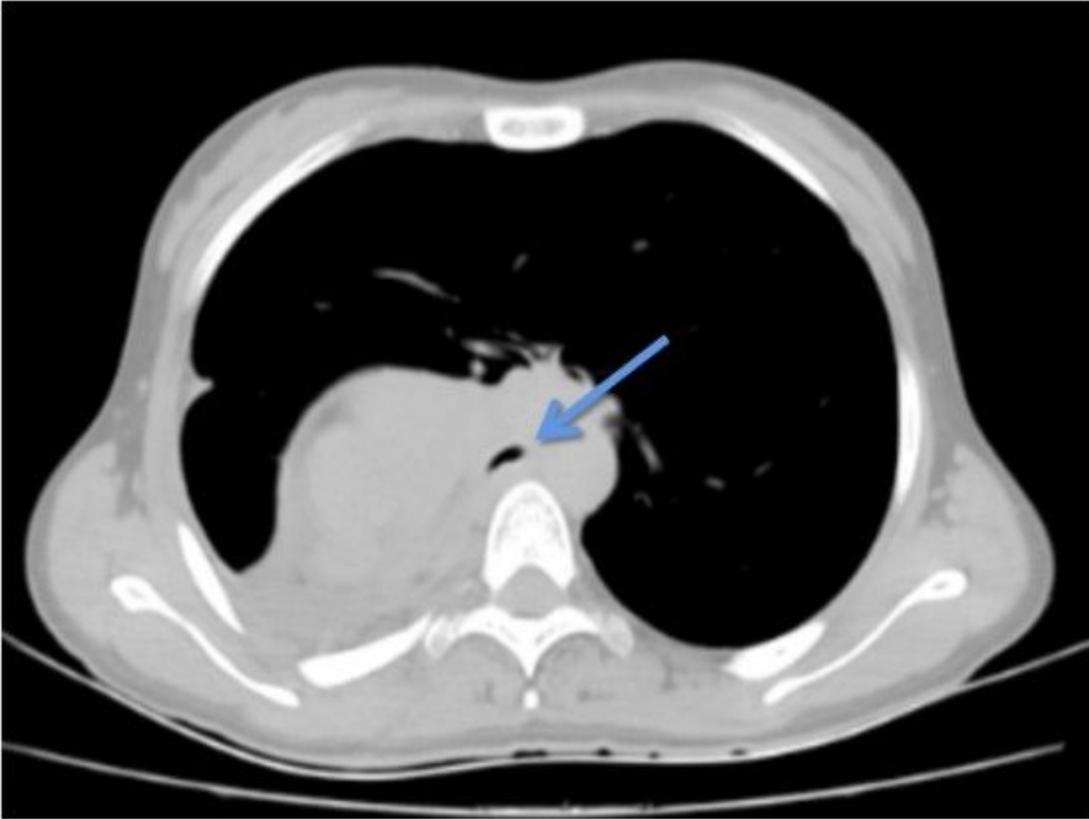


Figure 5

Stenosis of left main bronchus after operation The blue arrow points to a narrow, stenosing area of the left main bronchus

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