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A Unique Case of Lung Cancer Invading Azygos Lobe and Vein

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Abstract

In human anatomy, an azygos lobe is a congenital variation of the upper lobe of the right lung. It is seen in 1% of the population. Embryologically, it arises from an anomalous lateral course of the azygos vein in a pleural septum within the apical segment of the right upper lobe. As it has no bronchi, veins and arteries of its own or corresponding alteration in the segmental architecture of the lung, so it is not a true (misnomer), or even accessory, pulmonary lobe, but rather an anatomically separated part of the upper lobe. It is usually an incidental finding on chest x-ray or computed tomography and is as such not associated with any morbidity but can cause technical problems in thoracoscopic procedures. This is a case of a 55-year-old male presented to the hospital with cough, right chest pain and clubbing. Chest X-ray revealed a mass in the upper-middle zone of the right lung, and the presence of an azygos lobe.

Lung cancer in a patient with azygos lobe invading the azygos vein has not been reported previously. Operative strategy must take in account the anatomy of the azygos vein and its collaterals to avoid intraoperative complications. Invasion of the azygos vein should be added in the T4 group in the next UICC TNM lung cancer staging system

Keywords: Azygos; Chest X-ray; Pulmonary angiography; Spinal symptomatology

Introduction

This is a case of a 55-year-old male presented to the hospital with cough, right chest pain and clubbing. Chest X-ray revealed a mass in the upper-middle zone of the right lung, and the presence of an azygos lobe. Pulmonary angiography CT-scan confirmed the presence of 9 cm mass involving the upper and azygos lobes with invasion of the azygos vein. He underwent exploratory VATS, pneumonectomy with extended lymphadenectomy and combined resection of the involved azygos vein. Post-operative stage was pT4N2 (IIIB)

adenocarcinoma of the lung. Patient is well 3 months postoperatively.

Although involvement of mediastinum by a lung cancer is well known and the most frequently involved structures are superior vena cava, carina, left atrium, aorta and esophagus, no papers report about the surgical treatment of a lung cancer of the azygos lobe invading the azygos vein. Herewith we report such a case treated with extended right pneumonectomy and combined resection of the involved azygos vein.

Case Presentation

A 55-year-old male presented to the hospital with cough, right chest pain and clubbing. Chest X-ray revealed a mass in the upper-middle zone of the right lung and the presence of an azygos lobe. Chest CT-scan confirmed the presence of the large mass, measuring 9 cm in diameter, involving the upper, azygos lobes, azygos vein and mediastinal fat. No pleural effusion was present. Pulmonary angiography CT-scan showed obstruction of the azygos arch (**Figure 1**).

Positron Emission Tomography CT-scan was positive and showed no abnormal 18-FDG up-take in the mediastinum or presence of distant metastasis. A brain Magnetic Resonance showed only the presence of a pituitary adenoma. Clinical stage was cT4N0 (stage IIIB). CT guided trans-thoracic biopsy revealed the presence of an adenocarcinoma. Although patient underwent neoadjuvant chemotherapy, the mass did not decrease in size, and repeated CT scan and PET did not show significant modifications. Preoperative respiratory functional tests showed a FEV1 of 3.34 L/sec (93% of predicted) and a FVC of 5.21 L. Echocardiography was normal. Uniportal exploratory VATS were performed and the tumor involved the azygos lobe, the upper lobe and the azygos vein. Right postero-lateral thoracotomy and extrapleural dissection were mandatory.

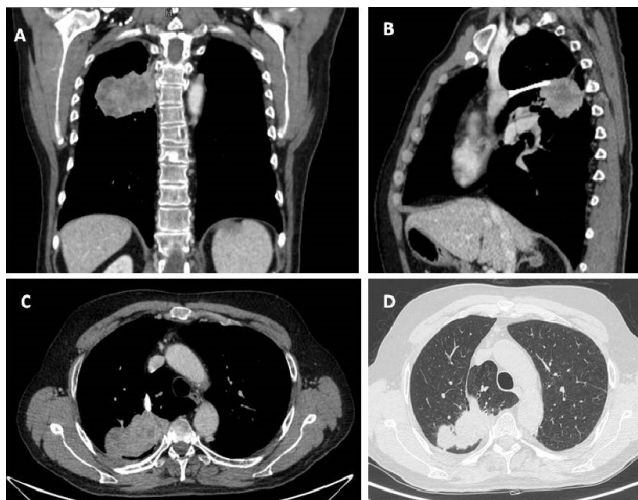


Figure 1 Chest CT-scan showing A-Coronal section showing the large tumour invading the mediastinum, B, C-Pulmonary angiography CT scan showing the tumor causing obstruction of the azygous vein, D-Axial section showing the mass and the azygos lobe.

The azygos vein was controlled by encircling the portion above and below the invasion (**Figure 2A**). An endovascular stapler was used to close and cut the vein proximal and distal to the tumour. All the intercostal veins draining to the resected portion of the azygos vein were clipped and cut (**Figure 2B**). Because an intraoperative lesion of the main pulmonary artery we have been forced to perform an en bloc right pneumonectomy with extensive lymphadenectomy (stations 4R, 7, 9). Two separated pulmonary veins for the upper and azygos lobes were present. Patient was discharged on the 7th postoperative day. Pathology showed the presence of a 10 cm adenocarcinoma. Post-operative stage was pT4N2 (IIIB). Patient died 23 months post-operatively.

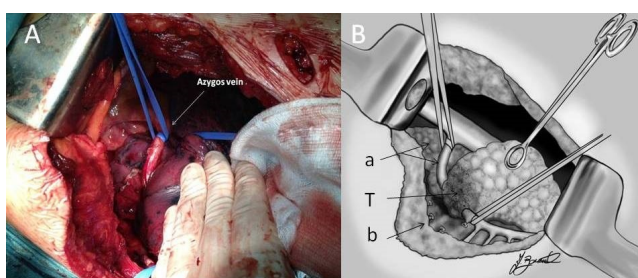


Figure 2 A-Intraoperative photo shows the azygos lobe and the azygos vein invaded by the tumour, B-Drawing shows our operative technique showing (A) azygos lobe and vein; T) tumour; (B) intercostal veins.

Discussion

The azygos lobe, described for the first time by Wrisberg in 1778 [1], is a well-known and well-described anatomical congenital malformation of the lung. Its incidence varies from

0.1% to 1.1% at autopsy, while it is discovered radiologically in 0.01% to 2.6% of the population [2]. Some authors support a vertical transmission with an autosomal dominant mode of inheritance [3]. In absence of azygos lobe, the invasion of the azygos vein showed by means of azygography, is a sign of inoperability for lung cancer because such cases are too far advanced to benefit from any form of surgery [4].

Primary lung cancer arising from an azygos lobe, however, is exceedingly rare, and only few surgical cases have been published [5-8]. None of these publications report about the invasion of the azygos vein. In our case it is impossible to determine if the tumour arised primarily from the azygos or upper lobe.

Furthermore, it is important to note that, although the neoadjuvant chemotherapy has been unsuccessfully, we were able to perform the operation because there was no progression of the disease. Nevertheless, the resection of the azygos lobe alone is not to be considered an oncologically radical resection, because the azygos lobe is not an independent lobe, but it is a partial segmentation of a normal right upper lobe by an abnormally placed azygos vein [2].

We want to emphasize that the invasion of the azygos vein has been determinant to modify our operative strategy. In fact, to remove the mass we have been forced to perform an en bloc resection of the lung with the upper portion of the longitudinal intrathoracic azygos vein and a portion of the azygos arch, as shown in the drawing (**Figure 2B**).

Conclusion

Large experiences about mediastinal T4 lung tumours report that the most frequently involved structures are superior vena cava, carina, left atrium, aorta, esophagus and vertebral bodies [9,10]. Moreover, some reports showed that there is a 20% 3-year survival in patients with involvement of superior vena cava, carina or intrapericardiac pulmonary artery, while a worse prognosis is related to the invasion of aorta, esophagus, left atrium or vertebral body. Although it has been demonstrated that there is a survival advantage after surgical extended resection in T4 NSCLC, an appropriate operative strategy has not been established [6]. The mortality rate reported in large experiences ranges from 8% at 30 days to 18% at 90 days, and morbidity rate varies from 20% to 50% [9]. On the contrary, no data on survival are available as regards T4 NSCLC invading the azygos vein.

Lung cancer in a patient with azygos lobe invading the azygos vein has not been reported previously. Operative strategy must take in account the anatomy of the azygos vein and its collaterals to avoid intraoperative complications. Invasion of the azygos vein should be added in the T4 group in the next UICC TNM lung cancer staging system.

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