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Mini Review

Azygos Lobe: Clinical and Surgical Importance

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Received: September 14, 2018; **Published:** October 17, 2018

Abstract

Azygos lobe (AL) is a rare normal anatomic variant of the lung with a frequent location at the right upper lobe. It may be encountered during thoracic surgery particularly with thoracic sympathectomy for hyperhidrosis, and it may be associated with lung tumors, spontaneous pneumothorax or pulmonary sequestration. This review of literature highlights the issues of embryogenesis, imaging features, clinical importance and surgical implications of AL. This anomaly implies a specific clinical importance for respiratory care physicians regarding therapeutic drainage of secretions from AL. Also, awareness of the anatomic basis of AL may affect the surgical plan and decision during thoracic surgery.

Keywords: *Anatomic Variation; Lung; Azygos Lobe*

Introduction

The lung is a vital organ of respiration which is divided on the right side into upper, middle and lower lobes separated by oblique and horizontal fissures, while the left lung is divided into upper and lower lobes separated by an oblique fissure [1]. Azygos lobe (AL) is a rare but normal anatomic entity of the right upper lobe, which had its first autopsy description by Heinrich Wrisberg in 1877. The incidence of AL ranges from 0.2% to 1.2%, which approximates 0.4% on radiography and 1% on cadaver anatomical dissection [2,3].

The specific importance of AL is related to its association with a difficulty during thoracoscopic sympathectomy, in addition to its coexistence with other conditions including lung tumors, pneumothorax and pulmonary sequestration [3-6]. It is important to radiologists and thoracic surgeons to be aware of the anatomic variations of pulmonary lobes including AL, as it may affect the surgical plan and decision [7].

Embryogenesis

The embryological development of the lung buds from the ventral wall of the foregut begins at the 4th week of gestation, with formation of the bronchopulmonary segments (10 on the right and 8 on the left side) and hence formation of lung lobes by the 6th week [8].

Embryologically, the azygos venous components originate from the last portion of the posterior cardinal veins. The right posterior cardinal vein, a precursor of the azygos vein, lies laterally to the apex of the right lung bud and then it migrates medially over the apex of the lung. Failure of the medial migration of the right posterior cardinal vein gives a rise to AL. The anomalous lobe is separated from the remaining part of the right upper lobe by azygos vein lying in azygos fissure and enclosed in a mesentery derived from the mediastinal pleura called meso-azygos (Figure 1) [2,9,10]. The meso-azygos results from the indentation of the lung and its overlying pleural layers by azygos vein, with a subsequent formation of a new structure (mesentery) constitutes of two folds of parietal and two folds of visceral pleura [11].

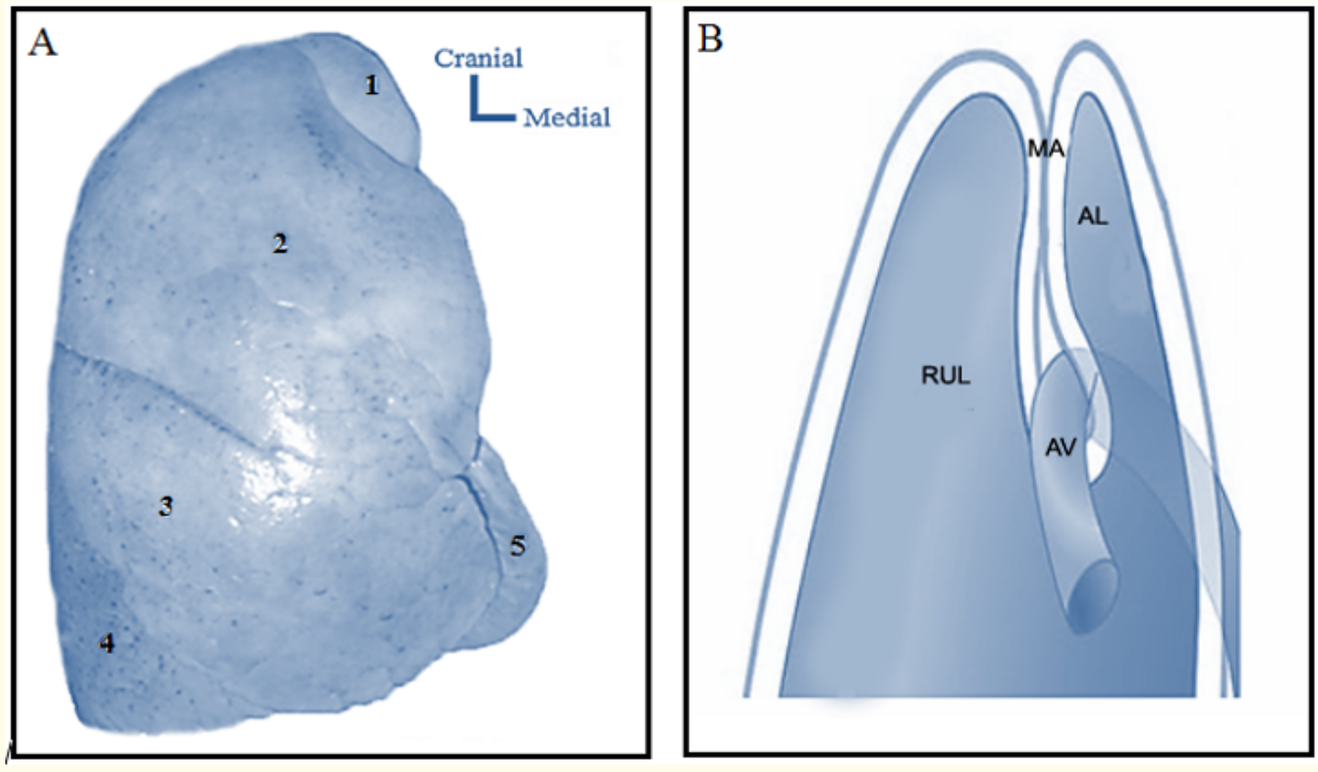

Figure 1: *(A): An autopsy photograph of azygos lobe located cranially in the right upper lung lobe: 1 Azygos lobe, 2 Upper lobe, 3 Middle lobe, 4 Lower lobe, 5 Hilum. (B): An illustration showing azygos vein (AV) enclosed in meso-azygos (MA) between the azygos lobe (AL) and the right upper lobe (RUL).*

In very rare instances, AL may locate on the left side probably as a result of embryonic cleft in the apical segment which encounters an azygos vein during its upward growth [12,13]. Azygos lobe on the right or left side was referred to as an accessory lung lobe, but at any side AL is not a true anatomic lobe because it receives its bronchial and arterial supplies from the apical or posterior segments of the upper lobe of the lung [11].

Imaging features

On chest radiographs, AL appears as a parenchymal part separated from the upper lobe by a fine line (azygos fissure) convex to the mediastinum and crosses the apex of the right lung which is triangular in shape at its upper portion (trigonum parietale). The azygos arch is normally located at the junction between the trachea and the main bronchus; however, in cases with AL the vein is located in the lowermost part of the azygos fissure as a tear-like shadow.

Radiologically, AL can be classified into three types according to its shape (Figure 2): type A where the trigonum parietale is located in the lateral aspect of the pulmonary apex with the azygos fissure as a fine vertical line in front of the posterior wall of the SVC, type B where the mesoazygos has a vertical path simulating a lung mass, and type C where the trigonum is located medially simulating a mediastinal mass [11,14].

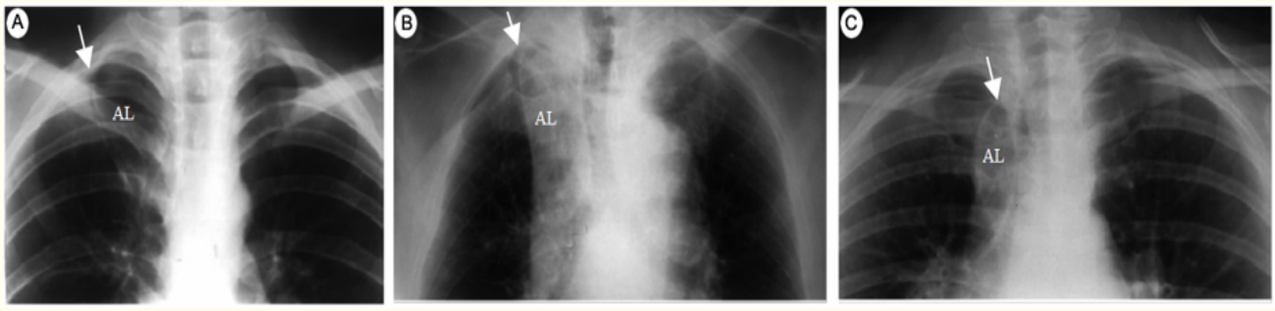

Figure 2: *Chest radiographs showing azygos lobe (AL) and its different types: (A) type A with trigonum parietale (arrow) located in the lateral aspect of the apex of the right lung lobe, (B) type B with a vertical path of meso-azygos (arrow) simulating lung mass, and (C) type C with medially located trigonum (arrow) simulating a mediastinal mass. AL, azygos lobe.*

In some cases, AL may appear on computed tomography (CT) but not visible on upright chest radiographs, probably due to lenticular shape of the great right mediastinal veins which results in difficult visualization of the azygos vein and AL [15]. Also, the presence of an azygos lobe can be missed on chest X-ray when a pneumothorax occurs [5].

Another challenge in the diagnosis of AL on chest radiographs is an increase in the opacity in AL which may be interpreted as a pathologic process, but opaque AL may be seen without any explanation of the possible cause of opacity [15,16]. Moreover, the azygos fissure may be simulated by lines resulted from other conditions such as scars, walls of bullae, displaced fissures, supernumerary fissures or the wall of a dilated esophagus, but the normal location of azygos vein excludes the presence of AL [15].

The imaging study by chest CT is helpful to confirm the diagnosis of AL (Figure 3), where the azygos fissure may be visible as C-shaped in large AL or slightly undulated in small AL. The fissure extends from the lateral aspect of the vertebral body posteriorly to the right brachiocephalic vein (RBV) and SVC anteriorly. The azygos vein appears thicker following the course of the fissure, and ends in the SVC or RBV. The azygos arch has higher position than its usual intramediastinal position and may simulate a pulmonary nodule if it has undulated appearance on its posterior portion [15,17].

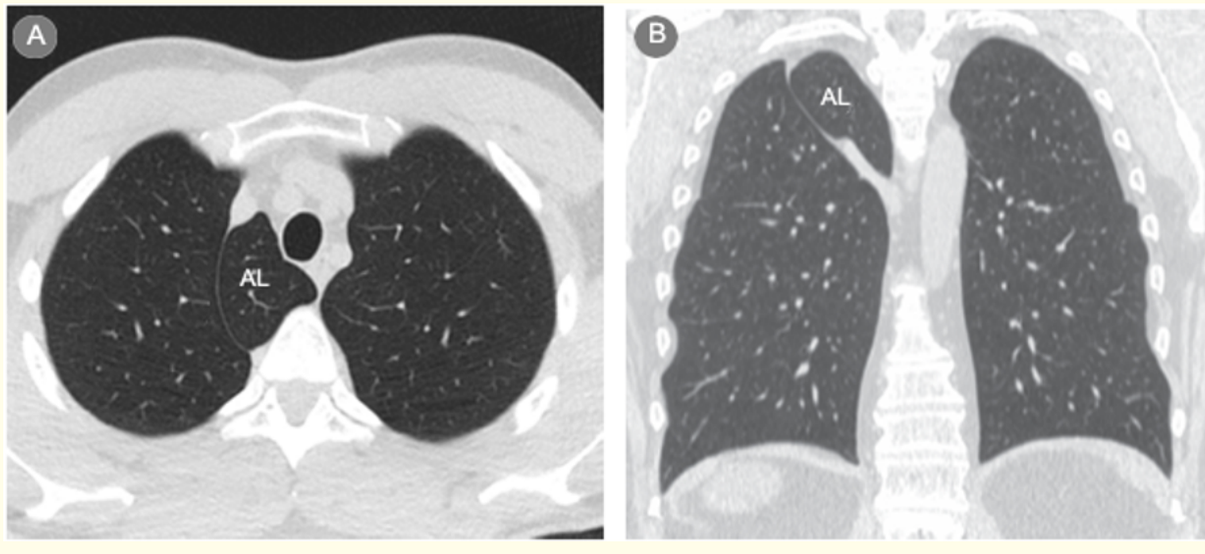

Figure 3: *Computed tomography images showing azygos lobe: (A) axial view and (B) coronal view. AL, azygos lobe.*

The azygos fissure may be empty on CT study due to an increase of the intrathoracic pressure by cough or vomiting, iatrogenic pneumothorax, or short meso-azygos resulting in displacement of the azygos vein to the mediastinum [16,18].

Clinical importance

Azygos lobe may be clinically confused with other conditions such as pulmonary bullous disease, pulmonary nodule, lung abscess or lung mass [2]. Moreover, AL may coexist with spontaneous pneumothorax [5,19], extralobar pulmonary sequestration [6], non-small cell lung cancer (NSCLC) [20-23], superior sulcus tumor [4], pulmonary artery sling [24] and tracheoesophageal fistula [25].

A rare sporadic association of AL with spontaneous pneumothorax has been reported, since AL can reduce the mechanical stress to the apex of the lung. Therefore, the presence of pneumothorax with AL may be attributed to an iatrogenic cause in association with insertion of central venous lines and surgical interventions [26].

Azygos lobe may associate with atelectasis and bronchiectasis particularly when there is a deep azygos fissure which may alter the bronchial wall and the quality of bronchial drainage in AL [3]. Notwithstanding, AL has no separated bronchial supply, thus bronchial deformation in the apical segment of the right upper lobe not that in the AL predisposes to atelectasis and bronchiectasis [27].

The study by Arakawa., *et al.* [3] proposed a suggestion of a respiratory care for patients with AL based on the detailed anatomical understanding of the bronchial branches in AL. The study revealed that the azygos lobe received bronchial branches from the medial components of the apical and anterior branches of the apical segmental bronchus (B1a and B1b) which bent excessively in a medial direction from the parent bronchial trunk. For therapeutic drainage of secretions from a right AL, a 45 degrees upright sitting posture with the neck flexed to the right side of at least 30 degrees is recommended, because in this posture the bronchial branches to the AL assume a more vertical orientation to facilitate greater dependent drainage of this lobe [3]. However, a controversy exists regarding the proper bronchial drainage of AL as it is difficult to suppose from a cadaveric study if a mere 30 degrees flexion could prove beneficial [28].

Surgical implications

The presence of AL may increase the risk of neurovascular injuries with excessive bleeding or phrenic nerve injury during thoracoscopic procedures for sympathectomy or pneumothorax cure [4,10,27-29]. Great care should be taken in cases with AL undergoing these procedures to avoid injury to the azygos vein since the subsequent fatal bleeding may indicate a rapid thoracotomy to achieve haemostasis [10]. Moreover, AL may prevent a clear view of the sympathetic chain during thoracoscopic sympathectomy [10,30].

Therefore, it is important to provide a good viewing in such cases (Figure 4). Valuable precautions have been recommended by Kauffman., *et al.* [30] which can be summarized as: (1) endotracheal intubation using a double-lumen probe to reduce the lung volume; (2) opening or creating window in the pleural curtain to expose the sympathetic chain better; (3) ligation of intercostal descending veins inside the curtain, if exist, before exposure of the sympathetic chain; (4) pushing the azygos vein aside or its ligation with extreme caution as it is a very fragile structure; and (5) assuring the restoration of AL back to its original location at the end of VATS to avoid the possibility of atelectasis and the subsequent postoperative pneumonia if it expands outside of its original location. Another possible approach, to gain better exposure of the sympathetic chain, was suggested by Reisfeld [31] to divide the azygos vein using multiple clips or intra- or extracorporeal sutures; however, the effect of azygos vein ligation is not known.

The surgical importance of AL extends beyond the associated difficulty during thoracoscopic sympathectomy to its implication on the surgical performance during other thoracic surgeries. Asai., *et al.* [5] performed thoracoscopic bullectomies uneventfully for a right primary spontaneous pneumothorax with a previously identified AL. The azygos arch was found dangling in the free inferior border of the meso-azygos which had a potential surgical hazard.

Azoury and Sayad [19] reported a case of recurrent right-sided spontaneous pneumothorax caused by rupture of AL bullae. Surgical management was successfully completed using VATS, through resection of the bullae harboring AL and subsequent pleurodesis, sparing the meso-azygos and azygos vein. The successful exposure of AL was attributed to successful deflation of the right lung and easy gentle pulling of AL inferiorly under the azygos vein in the absence of adhesions.

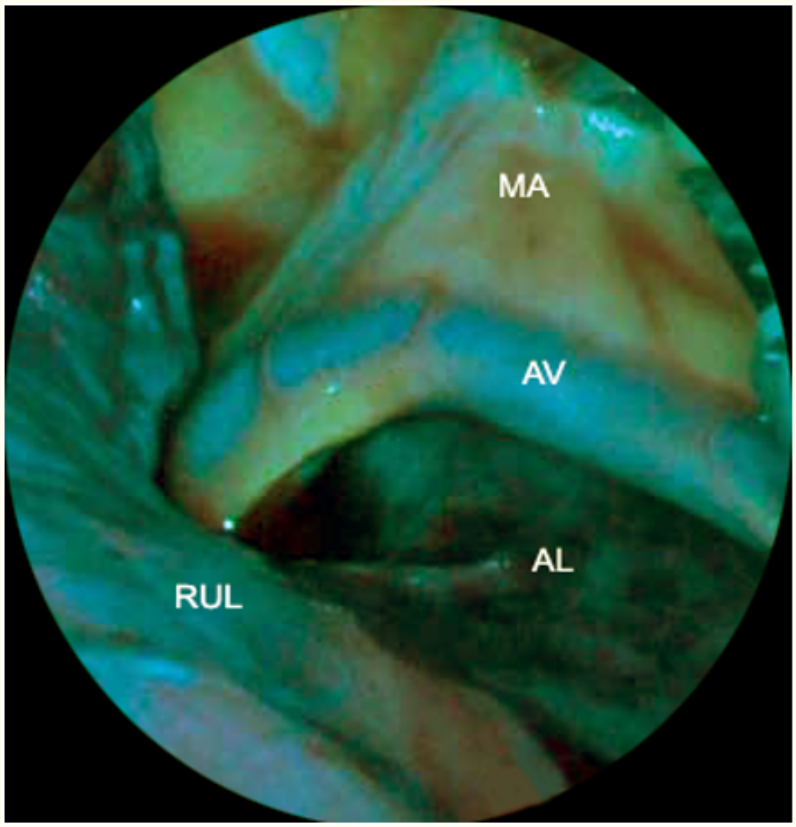

Figure 4: *Video image during VATS showing azygos lobe (AL), azygos vein (AV), meso-azygos (MA) and right upper lobe (RUL).*

Primary lung cancer associated with AL is extremely rare. Arai., *et al.* [20] performed right upper lobectomy and lymph node dissection with VATS for a case of lung adenocarcinoma without transecting the vein which was well mobilized by a cotton finger. In general, thoracotomy is preferable over VATS in such cases to avoid the risk of bleeding, if there are dense pleural adhesions around the azygos fissure. Also, Samancilar., *et al.* [21] described a case of a patient with AL and NSCLC in the upper lobe of the right lung underwent successful right upper lobectomy with mediastinal lymph node dissection using VATS.

In addition to upper lobe tumors developing next to AL, surgery may be indicated for tumors originate directly from AL. Delalieux., *et al*. [4] reported NSCLC presenting as a superior sulcus tumor in AL, for which complete resection after induction chemoradiotherapy was feasible by a transcervical-transmanubrial approach. The AL was resected after ligation of its specific branches from the pulmonary artery and superior pulmonary vein.

In some instances, better surgical results may be achieved by combination of thoracoscopic and open approaches, as reported by Papiashvili., *et al.* [32] who performed both approaches for a case of synchronous primary adenocarcinoma of AL and primary squamous cell carcinoma of the left upper lobe.

The extension of removal of lung cancer in AL may add more surgical challenge for such cases. Shakir., *et al.* [22] performed VATS for a case of presumptive lung cancer of AL. Those authors advocated that removal of AL alone may not be the best therapeutic option given the risk of locally recurrent disease, but in a select group of patients such as those with impaired lung function it may be the best available option in order to preserve postoperative pulmonary function. In addition to careful dissection, Shakir., *et al.* [22] used gentle caudal traction on the azygos lobe without significant mobilization of the azygos vein, to allow circumferential exposure to the lobe and identification of the bronchovascular pedicle. The AL was removed after division of its bronchovascular pedicle using thick tissue stapler.

Moreover, Fukuhara., *et al.* [23] described robot-assisted azygos lobectomy for adenocarcinoma arising in an azygos lobe. Those authors considered lobectomy of AL without concurrent resection of the right upper lobe is to be a limited resection and may not be an adequate oncological resection; however, an azygos lobectomy with mediastinal lymph node dissection may be an acceptable therapeutic alternative for elderly individuals with significant medical comorbidities or poor pulmonary function, to preserve postoperative pulmonary function and reducing the morbidity and mortality.

Summary and Conclusion

Azygos lobe is not a true anatomic lung lobe that may develop abnormally in embryo after failure of the medial migration of the right posterior cardinal vein. The anomaly may appear on chest radiographs separated by a fissure from the upper lobe, while chest CT is helpful to confirm its diagnosis with more details about the location of azygos vein, azygos arch and azygos fissure.

Clinically, AL should be differentiated from other pulmonary diseases especially bullous disease, tumor and abscess. If spontaneous pneumothorax coexists with AL, an iatrogenic cause should be suggested. The presence of atelectasis or bronchiectasis with AL adds a challenge in therapeutic drainage of secretions from the right AL which is recommended to be done during upright sitting posture at 45 degrees with neck flexion to the right side of at least 30 degrees.

The visual and traumatic challenges with AL during thoracoscopic sympathectomy or other thoracic surgeries on upper or azygos lobe can be reduced by careful dissection, double lumen intubation, successful deflation of the right lung, and gentle caudal traction of the AL. There are scant surgical case reports in literature describing the successful resection of the bullae harboring AL, lung adenocarcinoma in the right upper lobe, and removal of superior sulcus tumor or adenocarcinoma of AL. In a particular group of patients with presumptive lung cancer of AL and impaired lung function removal of AL alone may be the best available option to preserve the pulmonary function.

The radiologists, thoracic surgeons, and respiratory care physicians should be aware of the anatomical basis and the clinical implications of AL to insure proposing of the best surgical plan and decision in such normal challenging variant.

Conflict of Interest

None.

Funding

None.

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Volume 1 Issue 3 November 2018