An Autopsy Case of Azygos Lobe and the Extra-pulmonary Course of the Bronchial Veins in Man

By

Shoji CHIBA, Takao SUZUKI, Daisehachiro TAKAHASHI and Tatsuo KASAI

Second Department of Anatomy, School of Medicine, Hirosaki University
Hirosaki 036, Japan

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Summary: The present case represents the second report of an aberrant azygos lobe, following Adachi’s (1940) first case.

The authors describe the characteristic features of an azygos lobe observed in the right lung of a 65-year-old man as well as the extra-pulmonary course of the bronchial arteries and veins.

The literature on the azygos lobe reported in Japan and the extremely rare cases in which an azygos lobe has been found on the left side, are summarized.

Upon dissecting the pulmonary parenchyma, it became clear that the azygos lobe was supplied by definitive segmental branches (B1a and B2a, A1a and A2a, and V1a and V2a) of the tracheo-bronchial tree and pulmonary vessels. The authors review and summarize the patterns of the segmental bronchi and vessels in the azygos lobe and also discuss the development of this anomaly.

The azygos lobe is a comparatively rare anomaly of the lung caused by an aberrant course of the azygos vein. In this anomaly, the azygos vein is always enclosed within the right upper lobe except in a few cases where it has been observed on the left. Its frequency varies from 0.01% to 2.6%, according to estimates of previous authors. This anomaly was first recorded by Wrisberg in 1777 on both sides of a 3-year-old boy (Stibbe, 1919; Boyden, 1952).

Two or three characteristic features are observed in roentgenograms of the thorax, i.e., 1 a “tear-drop” or “comma” shaped linear shadow, corresponding to an aberrant fissure between the lobes, 2 a “triangle” shaped parietal shadow, indicating the beginning of the above-mentioned fissure, and 3 a denser “ovoid” shadow, indicating the confluence of the azygos vein with the superior vena cava (Bendick and Wessler, 1928; Hidaka and Isome, 1978). More than 1,936 cases of this anomaly have been reported (Anson and McVay, 1984), and 90 cases have been found in Japan. Bluntschli (1905), Stibbe (1919), Cairney (1923) and others have reviewed cases of this anomaly.

In the present paper, the authors describe the appearance of an azygos lobe in the right lung, an aberrant vein opening into the left atrium, and the extra-pulmonary course of the bronchial vessels, and review the previous cases reported in Japan and also the reports of left-sided azygos lobe. Moreover, the authors examine the bronchial and vascular supplies to the azygos lobe and review the
hypotheses for the origin of the azygos lobe published hitherto.

Materials and Methods

An azygos lobe was found in the right lung of a 65-year-old man during a practice dissection at Hirosaki University in 1985. The lung had no connections with the parietal membrane of the pleural cavity. No horizontal fissure between the upper and middle lobes was observed, except for a slight groove on the outer surface of the lung. No other congenital abnormalities were noted, but a small bronchial vein entered directly into the left atrium of the heart.

The segmental distribution of the bronchi and pulmonary vessels in the abnormal lobe was examined using a stereoscopic microscope.

Findings

1. Azygos lobe, meso-azygos and azygos vein

The azygos lobe was one of the largest ever found. It was separated from the remainder of the upper lobe of the lung by a 5-cm deep fissure on the outer surface. This fissure took a semi-circular course with a convexity laterally, and passed horizontally and medially to the pulmonary hilum (Figs. 1-3 in Plate I). The course of the fissure coincided with Type "a" of Stibbe's classification (1919).

Within this fissure, a fold of the parietal pleura was suspended like mesentery from the inner postero-lateral surface of the thorax and divided the pleural cavity into large medial and small lateral portions, enclosing the azygos lobe in the medial space. This fold, which has been referred to as the "meso-azygos" (Bluntschli, 1905; Hjelm and Hultén, 1928), consisted of two layers of parietal pleura. Four layers of visceral and parietal pleurae were inserted within the fissure. Attachment of the meso-azygos to the thorax commenced at the right side of the 5th thoracic vertebra, and then passed obliquely upwards across the posterior parts of the upper intercostal spaces and ribs to reach the first rib anteriorly (Fig. 4 in Plate I, and Fig. 5). The root of the meso-azygos extended along the inner surface of the first costal-cartilage medially and merged into the mediastinal pleura of the right lung.

The azygos vein, instead of arching over the right pulmonary hilum, took a longer course away from the 5th thoracic vertebra where the 3rd and 4th intercostal veins emptied into it. The azygos vein, about 1 cm in diameter, was contained within the free margin of the meso-azygos, and passed forwards and medially to enter the lateral part of the superior vena cava beneath the junction of the two brachiocephalic veins, at the level of the first intercostal space (Figs. 4-6).

2. Broncho-pulmonary segments and pulmonary vessels

The azygos lobe was supplied by two apical
Fig. 6. Anterior view of the pericardial cavity, showing the abnormal course of the azygos vein, with the mediastinal pleura reflected laterally.

The aberrant bronchial vein (indicated by the asterisk), which was observed at the superior part of the oblique sinus, passed through the venous mesocardium to enter the left atrium between the openings of the right and left pulmonary veins.
rami of the apical (B¹) and posterior (B²) segmental bronchi, accompanied by the pulmonary artery of the same segment. The two branches of the pair consisted of larger anterior and smaller posterior members. Each of these was distributed to the azygos lobe and to its surroundings. The branches showed a slight concavity at the base of the fissure (Figs. 3 and 7).

The arterial branches (A¹a and A²a) supplied the azygos lobe as well as the surrounding tissues. The vein (V¹a), after collecting blood from the entire azygos lobe except the small portion described below, entered the superior pulmonary vein, arching over the main stem bronchus and pulmonary artery. Blood from the posterior part of the azygos lobe drained into the apical ramus (V²a) of the posterior segmental vein. The venous branches, with the exception of the one on the medial surface, also collected blood from the surrounding tissues. The branches of the pulmonary vein mainly passed between the broncho-pulmonary and arterial segments, but the intra-pulmonary bronchial veins (Yamasita, 1954) supplied the walls, including those of the pulmonary veins (Figs. 7 and 8).

The right main stem bronchus, after a short course, divided into three: the apical, posterior and anterior segmental bronchi (Fig. 9). Both the apical (B¹) and posterior segmental bronchi (B²) further divided in two to form vertical and lateral branches. The azygos lobe was supplied by the ascending halves of the vertical branches of the apical and posterior segmental bronchi.

3. Aberrant bronchial vein draining into the left atrium

A small vein, 3 mm in diameter, at the superior part of the oblique pericardial sinus, passed through the venous mesocardium to enter the left atrium near the opening of the right superior pulmonary vein (Fig. 6 and Fig. 10 in Plate II). The aberrant vein was considered to be a bronchial vein, not a pulmonary vein, since it collected blood from the lymph nodes, trachea, bronchi, and mediastinum (Fig. 11).

According to Zuckerkandl (1881), the abnormal course of the bronchial vein which was observed in the present case is not rare and the venous mesocardium often offers a route for the bronchial veins. Horiguchi et al. (1988) stated that a small bronchial vein occasionally drained into the left atrium.

Abbreviations for Figures 1 ~ 6.

az: azygos vein
BD: right main stem bronchus
CCS: left common carotid artery
C², C⁴ and C⁸: 3rd, 4th and 8th cervical nerves
jd: right internal jugular vein
L: broncho-pulmonary lymph nodes
Ph: right phrenic nerve
pds: right superior pulmonary vein
pss: left superior pulmonary vein
T: trachea
TP: pulmonary trunk
Ts: sympathetic trunk
vcs: superior vena cava
Vd: right vagus nerve
4 ~ 8: 4th to 8th intercostal arteries and veins
1 ~ V: 1st to 5th ribs

ARC: arcus aortae
BC: brachiocephalic trunk
CCD: right common carotid artery
PD: right pulmonary artery
pdi: right inferior pulmonary vein
psi: left inferior pulmonary vein
sd: right subclavian vein
Ti: right internal thoracic artery
vci: inferior vena cava
4. Extra-pulmonary course of the bronchial vessels

The extra-pulmonary course of the bronchial arteries in the human body was investigated by Cauldwell et al. (1948), Kasai and Chiba (1979) and others, but only a few studies on the bronchial veins have been reported, i.e., by Zuckerkandl (1881) and Yamasita (1954). In the present case, the origin and course of the bronchial vessels were dissected in detail (Fig. 11). There were three bronchial arteries on each side. On the right, the first artery arose from the right subclavian artery and passed downwards along the antero-lateral surface of the trachea, the second artery arose from the left inferior thyroid artery and passed along the left recurrent laryngeal nerve forming a common trunk with the left bronchial artery, and the third artery arose from the right first aortic intercostal artery which was destined to become the 3rd and 4th intercostal arteries. The first bronchial artery entered the pulmonary hilum along the superior margin of the right main stem bronchus, the second artery passed along the inferior margin after crossing the tracheal bifurcation anteriorly, and the third artery arose from the right first aortic intercostal artery which was destined to become the 3rd and 4th intercostal arteries. The first bronchial artery entered the pulmonary hilum along the superior margin of the right main stem bronchus, the second artery passed along the inferior margin after crossing the tracheal bifurcation anteriorly, and the third artery passed transversely between the esophagus and the vertebral column ventral to the thoracic duct and entered the hilum along the postero-inferior surface of the bronchus.

On the left, two bronchial arteries arose from the thoracic aorta beneath the obliterated ductus arteriosus: one of them entered the pulmonary hilum along the superior margin of the left bronchus, and the other entered the hilum along its inferior margin, passing through the left vagus nerve plexus. The former connected with a branch of the left inferior thyroid artery near its origin. On the anterior surface of each main stem bronchus, a small communicating branch was found connecting the above-mentioned arteries.

The bronchial veins consisted of one or two small vessels which took almost the same course as the bronchial arteries but diverged from them near their junctions with the major veins and around the thoracic aorta. The bronchial veins on both sides communicated with each other to form a complicated venous plexus surrounding the bifurcation of the trachea, the bronchi, the esophagus, and the thoracic aorta. Because of these connections, venous blood from both pulmonary hila was easily conveyed to the other side. There were about 9 drainages of the bronchial veins on both sides, judging from their course and openings. The usual openings of the bronchial veins into the azygos vein were not confirmed.

On the right side, the first bronchial vein drained into the confluence of the two brachiocephalic veins, the second one into the left atrium, and the third into the inferior thyroid vein accompanied by the corresponding artery partially. The fourth vein, which had no accompanying arteries, drained into the left pericardiacophrenic or brachiocephalic vein, the fifth into the highest right intercostal vein, and the sixth into the right 5th intercostal vein, being accompanied by the right first aortic intercostal and the 5th intercostal arteries. The first 4 veins coursed...
along the anterior surface of the trachea and bronchi, while the last 2 coursed along the posterior surface of the right bronchus.

On the left side, the first and the second bronchial veins drained into the left 3rd to 6th intercostal and accessory hemiazygos veins after passing through the venous plexus surrounding the thoracic aorta. The third vein emptied into the anomalous vein which opened into the left atrium, the fourth and the fifth veins into the left inferior thyroid and brachiocephalic veins, and the sixth vein into the left highest intercostal vein.

Discussion

1. Reports on and frequencies of the azygos lobe

Anatomical findings concerning the azygos lobe have been summarized by Bluntschli (1905), Stibbe (1919), Cairney (1923), Velde (1930), Anson and Smith (1936), Adachi (1940), Foster-Carter (1946) and Boyden (1952). So far, about 120 autopsy cases of this anomaly have been reported. Boström (in a private letter to Mäusert, 1899) found 17 cases in a total of 1,600 human dissections (frequency, 1.06%), and Boyden (1952) found one in 500 cadavers (0.207%). The incidence varied from 0.1% to 1.1%.

The present example, which was found in one of 424 cadavers dissected (0.23%; Table 1), represents the second autopsy case reported in Japan, the first having been reported by Adachi (1940).

Radiologically, Bendick and Wessler (1928) first confirmed that an abnormal shadow observed in the thorax was due to an azygos lobe of the right lung. The radiographic incidence varied from 0.01% to 2.6%, as follows: 2 out of 23,334 cases examined (0.01%; Tachibana, 1955), 130 out of 50,000 (0.26%; Etter, 1947), 10 out of 3,660 (0.27%; Velde, 1930), 20 out of 5,652 (0.35%; Becanu, 1938), 70 out of 10,000 (0.7%; Günther und Müller, 1980), and 15 out of 580 (2.6%; Bourdelles et Jalet, 1931). Koshimura (1941) summarized 664 reported cases of the azygos lobe including his own cases, and Anson and McVay (1984) claimed that the anomaly amounted to 1,936 cases in 323,614 persons examined (0.59%).

Underwood and Tattersall (1933) stated that all the published descriptions of the azygos lobe up until 1923 were based on post-mortem findings, whereas the anomaly was subsequently reported more frequently radiologically, and that cases were found in about 25% of fetuses in a reference to Débré and Mignon (1931). Cascelli (1931) believed that the anomaly occurred more frequently in human embryos and fetuses than in adults. The frequency of appearance of the azygos lobe in men is 3 times higher than that in women (Velde, 1930). The azygos lobe is found normally in the porpoise (Cairney, 1923; Adachi, 1940).

Most azygos lobes were observed in the right lung, but in a few cases they were found either on the left or on both sides, i.e., 11 cases on the left and 5 cases on both sides (Table 2). Schmitz-Cliever (1950) reported the existence of the anomaly on the left by

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Fig. 8. Medial view, showing the pulmonary hilum and the branching of the bronchi and pulmonary vessels of the right lung.

The azygos lobe was supplied by the apical rami (B¹a and B²a) of the apical and posterior segmental bronchi and by the accompanying arterial branches. The veins from the azygos lobe drained into the superior pulmonary vein, passing either on the medial surface (a large part of V¹a) or on the lateral (V¹a and V²a).

In this figure, the intra-pulmonary bronchial veins (Yamasita, 1954) can be seen in the lower lobe (indicated by the asterisk).
laminography, and Koshimura (1941) and Mex (1954) summarized such cases.

The course of the azygos vein and attachment of the meso-azygos were almost the same in all the cases described in the literature, except in those reported by Gruber (1880) and Müller (1928), in which the azygos lobe was enveloped with pleura which had no connection with the pleural cavity and was supplied by an independent bronchus from the right upper lobe.

According to Stibbe (1919), Nelson and Simon (1931), the entrance of the azygos vein into the superior vena cava is slightly below the bifurcation of the two brachiocephalic veins but is shifted upwards in most cases of the azygos lobe when compared with normal subjects. The above findings were also confirmed in computed tomography (CT) scans by Speckman et al. (1981), and the presence of the azygos lobe was found to alter the contour of the right mediastinum.

Most cases of this anomaly are accompanied by no other congenital defects. In the cases described by Fischer (1899), Müller (1928) and Hasimoto et al. (1962), the middle lobe of the right lung was unusually small or less sharply demarcated from the upper lobe. In the case described by Fukabori (1932) in a 14-year-old boy, congenital absence of the pectoralis major muscle and syndactyly, or fusion of the fingers in the hand, were also found. Recently, Pomeranz and Proto (1986) reported a case in which the inferior vena cava drained into the azygos vein.

The position of the azygos fissure was classified into three types of Stibbe (1919). These are: Type “a”, in which the fissure passes into the lateral surface of the lung below the apex; Type “b”, in which the vertical fissure passes into the apex; and Type “c”, in which it passes into the medial surface below the apex. The size of the azygos lobe generally depends on the site of the fissure, and it is larger in Type “a” than in the other two types. The incidence of each type varies according to whether the specimens were examined roentgenologically or at autopsy (Table 3).

2. Bronchial supply of the azygos lobe

The bronchial and vascular supplies to the azygos lobe have not been investigated in detail. Cairney (1923) prepared a model of the branching of the eparterial bronchus and found that the apical bronchus (B\(^1\)) had three divisions: a medial division, which supplied the abnormal lobe; and a posterior and a lateral division, which supplied the remainder of the apical segment of the right upper lobe. Bendick and Wessler (1928) examined the branching pattern of the bronchus in the

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Fig. 9. Dorsal view of the right tracheo-bronchial tree.

The apical rami (B\(^1\)a and B\(^2\)a) of the apical and posterior segmental bronchi, which supplied the azygos lobe as well as its surroundings, were encircled by the abnormal azygos vein at the roots of the above two branches (indicated by arrows).

**Abbreviations for Figures 7 ~ 9.**

BD: right main stem bronchus  
L: broncho-pulmonary lymph nodes  
PD: right pulmonary artery  
pdi: right inferior pulmonary vein  
psd: right superior pulmonary vein  
B\(^1\)a: apical ramus of the apical bronchial segment  
B\(^1\)b: anterior ramus of the apical bronchial segment  
B\(^2\)a: apical ramus of the posterior bronchial segment  
B\(^2\)b: posterior ramus of the posterior bronchial segment  
B\(^3\) ~ B\(^{10}\): 3rd to 10th bronchial segments of the right lung  
V\(^1\)a: apical ramus of the apical segmental vein  
V\(^1\)b: anterior ramus of the apical segmental vein  
V\(^2\)a: apical ramus of the posterior segmental vein  
V\(^3\): anterior segmental vein of the right upper lobe
azygos lobe by the injection method. Foster-Carter (1946) summarized the reported cases and confirmed that the bronchus supplying the abnormal lobe was kinked downwards by the azygos vein which took an "N"-shaped course in the meso-azygos (Nelson and Simon, 1931).

Boyden (1952) collected 12 cases in which the bronchial supply was described and confirmed that the azygos lobe was aerated by the mediastinal branches of the apical segmental bronchus (B1) and occasionally by branches of the apical portion of the posterior segmental bronchus (B3). He also reported that the smaller lobes tended to be supplied by a single branch (Table 4). According to Boyden and Scannell (1948) and Wells and Boyden (1954), the apical segmental bronchus consists of apical and anterior subsegmental rami (B1a and B1b) and the posterior segmental bronchus consists of apical and posterior subsegmental rami (B3a and B3b). Since the abbreviation of the posterior segmental bronchus in Nomina Anatomica (1983) is B2, it would be necessary to convert it to the position of B2 from B3 in Boyden's (1952) figures and table. In the present case, the large azygos lobe was aerated by the halves of the two medial vertical branches of the apical (B1) and posterior (B2) segmental bronchi, employing the terminology of Nomina Anatomica (1983). The branching pattern of the broncho-pulmonary segments was almost the same as that described by Foster-Carter (1946) and by Boyden (1952).

3. Vascular supply of the azygos lobe

The vascular supply of the azygos lobe has not been described previously, except by Bluntschli (1905), Foster-Carter (1946) and Boyden (1952), as indicated in Table 4. Bluntschli (1905) stated briefly that the vascular supply of the azygos lobe coincided with its bronchial supply. In the 2 cases reported by Foster-Carter (1946), the azygos lobe was supplied by branches of the pulmonary artery accompanying the divisions of the apical bronchus (B1) and by a "recurrent" branch (Appleton, 1945) accompanying those of the posterior bronchus (B2). The azygos lobe was supplied by the arterial branches A1 and A2 in case 4 and by the branch A1 in case 5. In these cases, the veins of the azygos lobe joined with those from the remainder of the apical and posterior broncho-pulmonary segments before draining into the superior pulmonary vein.

In the case described by Boyden (1952), the azygos lobe was supplied by the branches (A1 and A2) of the pulmonary artery which accompanied the apical and posterior bronchial segments, and the veins of the azygos lobe drained into the superior pulmonary vein after...
Table 1. Summarized data on the azygos lobe reported in Japan.

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Total 90

* — Quoted by Adachi (1940).
** — Quoted by Kimura and Omori (1966).
*** — This report.
Table 2. Cases of azygos lobe observed either on both sides or on the left.

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<td>both sides</td>
<td>autopsy</td>
<td>1</td>
</tr>
<tr>
<td>Boström*</td>
<td>both sides</td>
<td>autopsy</td>
<td>1</td>
</tr>
<tr>
<td>Merkel (1899)</td>
<td>both sides</td>
<td>autopsy</td>
<td>1</td>
</tr>
<tr>
<td>Mex (1954)</td>
<td>both sides</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Bourdellès &amp; Jalet (1931)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Bourdellès et al. (1932)**</td>
<td>left</td>
<td>X-ray</td>
<td>2</td>
</tr>
<tr>
<td>Veyssi (1937)**</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Becanu (1938)</td>
<td>left</td>
<td>X-ray</td>
<td>2</td>
</tr>
<tr>
<td>Křivinka (1939)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Suchomel (1949)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Schmitz-Cliever (1950)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Weston (1954)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
<tr>
<td>Gassman (1958)</td>
<td>left</td>
<td>X-ray</td>
<td>1</td>
</tr>
</tbody>
</table>

* — Quoted by Mäusert (1899).
** — Quoted by Koshimura (1941).

Table 3. Frequency of each type of the azygos fissure according to Stibbe's classification (1919).

<table>
<thead>
<tr>
<th>Reported by</th>
<th>Number of cases examined</th>
<th>Type of azygos fissure</th>
<th>Subjects examined</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>a</td>
<td>b</td>
</tr>
<tr>
<td>Cairney (1923)</td>
<td>22</td>
<td>9</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(41%)</td>
<td>(32%)</td>
</tr>
<tr>
<td>Underwood &amp; Tattersall (1933)</td>
<td>12</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(33%)</td>
<td>(42%)</td>
</tr>
<tr>
<td>Koshimura (1941)</td>
<td>129</td>
<td>41</td>
<td>69</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(32%)</td>
<td>(53%)</td>
</tr>
<tr>
<td>Etter (1947)</td>
<td>130</td>
<td>53</td>
<td>42</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(41%)</td>
<td>(32%)</td>
</tr>
<tr>
<td>Günther &amp; Müller (1980)</td>
<td>70</td>
<td>9</td>
<td>37</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(13%)</td>
<td>(53%)</td>
</tr>
<tr>
<td>Chiba et al. (1990)</td>
<td>41</td>
<td>7</td>
<td>12</td>
</tr>
<tr>
<td>(this report)</td>
<td></td>
<td>(17%)</td>
<td>(29%)</td>
</tr>
</tbody>
</table>
Table 4. Segmental constitution of the bronchi and pulmonary vessels in the azygos lobe. The nomenclature of the segments is based on Boyden's table (1952), using the terminology of Nomina Anatomica (1983).

<table>
<thead>
<tr>
<th>Reported by</th>
<th>Size of azygos lobe</th>
<th>Bronchial supply</th>
<th>Pulmonary vessel supply</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gruber (1870)</td>
<td>smaller</td>
<td>B(^1)a two distinct branches</td>
<td>branches of A(^1) and V(^1)</td>
</tr>
<tr>
<td>Matthews (1898)</td>
<td>larger</td>
<td>B(^1)a</td>
<td>A(^1)a and V(^1)a</td>
</tr>
<tr>
<td>Dévé (1899) case 2</td>
<td>smaller</td>
<td>B(^1)a</td>
<td></td>
</tr>
<tr>
<td>Bluntschli (1905)</td>
<td>larger</td>
<td>small branch of B(^1)a</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>branch of the eparterial bronchus</td>
<td></td>
</tr>
<tr>
<td>Geddes (1910)</td>
<td>smaller</td>
<td>B(^1)a</td>
<td></td>
</tr>
<tr>
<td>Cairney (1923)</td>
<td>smaller</td>
<td>B(^1)a</td>
<td></td>
</tr>
<tr>
<td>Bendick &amp; Wessler (1928)</td>
<td>smaller</td>
<td>small branch of B(^1)a</td>
<td></td>
</tr>
<tr>
<td>Nelson &amp; Simon (1931)</td>
<td></td>
<td>branch of the eparterial bronchus</td>
<td></td>
</tr>
<tr>
<td>Brock (1944) case 1</td>
<td>larger</td>
<td>B(^1)a and B(^1)b</td>
<td>A(^1)a and A(^2)a,</td>
</tr>
<tr>
<td>case 2</td>
<td>larger</td>
<td>B(^1)b</td>
<td>superior pulmonary vein</td>
</tr>
<tr>
<td>Boyden (1952)</td>
<td></td>
<td>B(^1)a and B(^2)a</td>
<td>A(^1)a, superior pulmonary vein</td>
</tr>
<tr>
<td>Hasimoto et al. (1962)</td>
<td></td>
<td>B(^1)b</td>
<td></td>
</tr>
<tr>
<td>Chiba et al. (1990)</td>
<td></td>
<td>B(^1)a and B(^2)a</td>
<td></td>
</tr>
<tr>
<td>(this report)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

collecting blood from the remainder of the right upper lobe. The venous branches (V\(^1\) and V\(^2\)), which were located either on the medial surface of the azygos lobe or on the lateral surface adjacent to the remainder of the upper lobe, were not accompanied by the segmental bronchus or the pulmonary artery. In the present case, the azygos lobe was supplied largely by the same branches (A\(^1\) and A\(^2\), V\(^1\) and V\(^2\)) of the pulmonary vessels as described by previous authors.

4. Etiology of the azygos lobe

Hypotheses concerning the origin of the azygos lobe have been summarized by Bluntschli (1905), Stibbe (1919), Velde (1930), Koshimura (1941), Foster-Carter (1946), Lutz (1951) and Boyden (1952).

Cleland (1870), Collins (1874) and Lindsay (1910; quoted by Gillaspie et al., 1916) suggested that the azygos lobe was derived from a parietal splitting of the normal right upper lobe due to an abnormal course of the azygos vein.
Motti (1893), Sappey (quoted by Stibbe, 1919), and Nakagawa and Mori (1958) believed that the lung bud had not grown sufficiently. Geddes (1910) ascribed its origin to supernumerary branching of the right bronchus. Matthews (1898) suggested that a slightly higher position of the outgrowing lung bud would cause the upper lobe to impinge on the vein instead of clearing it. He also stated that the same condition might result from a temporal alteration in development, i.e., by lung outgrowth being slightly delayed or the descent of the heart being slightly advanced.

Dévé (1899) believed that the upper part of the posterior cardinal vein passed more laterally and emptied into the lateral part of the right ductus cuvieri rather than into the posterior as in normal cases. Orth (1887; quoted by Lutz, 1951) attributed its origin to the higher level of the junction between the azygos vein and the superior vena cava.

Wells (in a private letter to Boyden, 1952) stated that the abnormal displacement of the arch of the posterior cardinal vein was caused by delayed closure of the pleuropericardinal canal. Lutz (1951) believed that the azygos lobe was a result of an abnormally lateral deviation of part of the posterior cardinal vein which was destined to be the cranial part of the future azygos vein.

In a series of transverse sections of 10.5-mm human embryos, Bluntschli (1905) found that the cranial part of the pleural cavity was divided in two and the opening of the posterior cardinal vein into the ductus cuvieri was situated between the two parts of the cavity, not medially as indicated in previous papers. In 15-mm, 22-mm and 23-mm human embryos, however, the position of the opening was shifted medial to the pleural cavity as seen in normal cases. If the condition observed in the early stage of the embryo mentioned above continued, it would form an azygos lobe, since the pleural fold would be pulled downwards along with the descent of the thoracic viscera, dividing the upper lobe into two. The abnormal lateral position of the posterior cardinal vein was considered to be a result of delayed development of the right subclavian vein which passed beneath the brachial nerve plexus. Bluntschli also pointed out that the cranial part of the cavity on the left was found at the lower margin of the first rib, and it was about half a thoracic vertebra segment lower than on the right. Bluntschli’s study (1905) clarified the development of the azygos lobe and of the rare cases of left-sided azygos lobe.

Holtby (1915) reported that in 5-mm to 20-mm human embryos, the posterior cardinal vein passed through a well-defined pleural fold outside the upper part of the lung and that it was then possible for the apex of the lung to grow on either side of the vein. When this happened, part of the right upper lobe would be separated from the remainder by a mesentery-like fold of pleura enclosing the azygos vein.

In the normal development of the lung of a 4-mm human embryo, Boyden (1955) indicated that during the descent of the heart it is drawn forwards near the anterior part of the posterior cardinal vein. During this period, the lung bud has just commenced to bifurcate, and subsequently, in 5- to 7-mm embryos, the apical lobe of the right lung grows out from the right bronchus to take its normal position forming the future apex of the lung after passing beneath the arch of the future azygos vein.

Stibbe (1919) believed that in cases of azygos lobe, the lung bud failed to clear the venous arch mentioned above due to either (1) a primary abnormal position of the posterior cardinal and common cardinal veins, or (2) maldevelopment of the lung. He suggested that the immediate cause of this anomaly might be a positional alteration of the lung to the developing azygos vein.

At present, the theories expounded by
Bluntschli (1905), Holthy (1915) and Stibbe (1919) concerning the development of the azygos lobe are commonly accepted, and it is believed that the azygos lobe occurs fortuitously as a result of failure of the azygos vein to slide medially over the growing lung apex (Bendick and Wessler, 1928; Foster-Carter, 1946; Boyden, 1952 and 1955; Hollinshead, 1971; Gray and Skandalakis, 1972; Anson and McVay, 1984). However, Suess (1928), Underwood and Tattersall (1933) and Postmus et al. (1986) have suggested the possibility of hereditary factors being involved in the etiology of the azygos lobe.

**Acknowledgement**

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**References**

24) Fischer, E.: Seltener Verlauf der Vena azygos


Explanation of Figures

Plate I

Fig. 1. Photograph of the present case. Lateral aspect of the azygos lobe, showing the azygos fissure between the aberrant lobe and the remainder of the right upper lobe as well as the shallow groove corresponding to the horizontal fissure.

Fig. 2. Medial view, showing the right pulmonary hilum and a tunnel (indicated by the asterisk) for the azygos vein at the base of the azygos fissure. A slight notch (indicated by the arrow) in the anterior margin of the lung coincided with the horizontal fissure.

Fig. 3. Superior and postero-lateral view, showing the deep azygos fissure.

Fig. 4. Anterior view of the right pleural cavity. The azygos vein was cut near its opening into the superior vena cava and pulled laterally, being contained within the free margin of the meso-azygos. The meso-azygos consisted of two layers of parietal pleura, being suspended like mesentery from the inner surface of the first rib and cartilage to divide the cavity into medial and lateral portions.
Plate II

Fig. 10. Dorsal aspect of the heart.

The aberrant vein (indicated by the white arrow), 3 mm in diameter, drained into the left atrium near the opening of the right superior pulmonary vein. This vein collected blood from the trachea, bronchi, lymph nodes and mediastinum, as shown in Fig. 11.

Abbreviations for Figures 10 and 11.

ARC: arcus aortae  Ad: right atrium
AT: thoracic aorta  As: left auricle
az: azygos vein  bcd: right brachiocephalic vein
bcs: left brachiocephalic vein  CCS: left common carotid artery
BS: left main stem bronchus  E: esophagus
CCS: left common carotid artery  Lig: ligamentum arteriosum
E: esophagus  pds: right superior pulmonary vein
Lig: ligamentum arteriosum  pss: left superior pulmonary vein
pds: right superior pulmonary vein  Rd: right recurrent laryngeal nerve
pss: left superior pulmonary vein  sc: coronary sinus
Rd: right recurrent laryngeal nerve  sd: right subclavian vein
sc: coronary sinus  ss: left subclavian vein
SD: right subclavian artery  Ti: internal thoracic artery
SS: left subclavian artery  TP: pulmonary trunk
T: trachea  ty: inferior thyroid vein
TY: inferior thyroid artery  Vs: left vagus nerve
Vd: right vagus nerve  vsc: superior vena cava
vci: inferior vena cava
1 ~ 7: 1st to 7th intercostal arteries and veins