PULMONARY SEQUESTRATION – ANALYSIS OF DIAGNOSTIC AND THERAPEUTIC DIFFICULTIES

JOLANTA HAUER1, JACEK ALCHIMOWICZ2, MARIA HARAZDA1, JULIUSZ PANKOWSKI1,2, JAROSŁAW KUZDŻAL1, MARCIN ZIELIŃSKI1, TOMASZ GRODZKI2

Department of Thoracic Surgery, Pulmonary Hospital in Zakopane1
Ordynator: dr hab. M. Zieliński

Department of Thoracic Surgery, Regional Hospital for Lung Diseases, Szczecin-Zdunowo2
Ordynator: dr hab. med. T. Grodzki

The aim of the study was to review the clinical characteristics of operated patients with respect to pulmonary sequestration and analyze diagnostic and therapeutic problems connected with this anomaly.

Material and methods. We performed a retrospective analysis of 23 patients operated on because of pulmonary sequestration between 1963 and 2004 in the Departments of Thoracic Surgery in Zakopane (10 patients) and Szczecin-Zdunowo (13 patients).

Results. The study group comprised 8 men and 15 women and the mean age was 28 years. 15 patients (65.2%) presented with clinical symptoms such as coughing, fever and chest pain. Preoperatively, pulmonary sequestration was suspected in 5 patients. Amongst the remaining patients, the preoperative diagnosis was lung cancer (9 patients), metastasis from testicular neoplasms (n=1), emphysematous bullae (n=2), pulmonary cyst (n=4), bronchiectasis (n=1) and lung abscess (n=1). None of the patients were subjected to arteriography and 6 patients underwent contrast-enhanced computer tomography examination, although it did not lead to a proper diagnosis. We found 20 intralobar sequestrations and 3 extralobar sequestrations, which were situated above the diaphragm. There were 11 sequestrations on the right side and 12 on the left side. The operations performed included: lobectomy (14 patients), segmentectomy 1+2 (n=1), wedge resection (n=3), sequestrectomy (n=3), lower bilobectomy (n=1), and middle lobectomy (n=1). Blood supply from the thoracic aorta was found in 17 patients and the abdominal aorta in the remaining 6 patients. In 5 patients, the pulmonary sequestration was supplied by more than one artery. Complications included hemorrhage from supplying arteries in 8 patients and phrenic nerve palsy in one patient. One patient required mechanical ventilation after the operation. There was no further morbidity and mortality. Distant surgical results were good.

Conclusions. 1. Pulmonary sequestration in adults is difficult to diagnose before the operation, even with a detailed computer tomography examination. 2. Intralobar sequestration is much more common (87%) than extralobar and is often situated in the basal segments with comparable incidence on the right and left sides. 3. The greatest danger during the operation is major bleeding from the supplying artery. 4. Prognosis after the operation is favorable.

Key words: pulmonary sequestration, surgical treatment, complications

Pulmonary sequestration is a congenital anomaly of the lungs and its most characteristic feature consists of abnormal blood supply from the systemic circulation. The sequestration may be localized to intralobar and extralobar sections without a pulmonic connection. Due to the rarity of this anomaly in adults, the diagnosis of sequestration is rarely considered before the operation.

There are few publications in the literature concerning pulmonary sequestration and the majority of them are case reports. In this study, we presented our results concerning the treatment of patients with pulmonary sequ-
Pulmonary sequestration – analysis of diagnostic and therapeutic difficulties

MATERIAL AND METHODS

We retrospectively analyzed treatment results considering 23 patients operated on because of pulmonary sequestration during 1963 and 2004 in two Departments of Thoracic Surgery: Zakopane (10 patients) and Szczecin-Zduńowo (13 patients), with emphasis on the difficulties with proper preoperative diagnosis and treatment.

RESULTS

Mean patient age amounted to 28 years (ranging from 14 and 59). The study group comprised fifteen female patients (mean age of 25.6) and 8 male patients (mean age of 32.7).

The most common symptoms were as follows: recurrent infections, productive cough with seropurulent sputum, chest pain and hemoptysis. Based on diagnostic examinations, such as chest X-rays, tomograms computer tomography, bronchography, and bronchoscopy, a preoperative diagnosis of pulmonary sequestration was possible in five patients. In the remaining 18 patients, the preoperative diagnosis was as follows: lung cancer (n = 9), lung abscess (n = 1), lung cysts (n = 4), emphysematous bullae (n = 2), bronchiectasis (n = 1) and testicular cancer metastasis (n = 1). Intralobar sequestration was observed in 20 patients, while extralobar sequestration was observed in three. Among the 20 intralobar sequestrations, 17 were localized to the basal segments, two in the middle lobe and one in segments 1+2. All 3 cases of extralobar sequestration were localized within the pleural cavity above the diaphragm. There were 11 pulmonary sequestrations on the right side and 12 on the left.

Lower lobectomy was performed in 14 patients (3 – wedge resections, 3 – sequestrectomies, 1 – lower bilobectomy, 1 – middle lobectomy and in 1 – segmentectomy).

In 8 of 18 patients (44%) without preoperative suspicion of sequestration, there was major intraoperative bleeding, which required blood transfusions during the procedure or shortly afterwards. Such bleeding did not occur in any of the patients with sequestration diagnosed preoperatively.

Blood supplementation came from the thoracic aorta in 17 cases and the abdominal aorta in 6 cases. In most cases, there was one blood vessel 2–4 mm in diameter. After the operation in one patient, we noticed a phrenic nerve palsy (considering available medical records there are no data regarding whether the palsy was temporary or permanent) with atelectasis of the middle lobe. Moreover, one patient required mechanical ventilation after the surgical procedure (tab. 1).

DISCUSSION

There are two major groups of pulmonary anomalies:

– anomalies associated with developmental disturbances of the foregut, from which the bronchial buds develop,
– anomalies associated with developmental disturbances of the sixth branchial cleft, from which the pulmonary vessels are formed.

Pulmonary sequestration belongs to the first group, amongst other anomalies such as agenesis, aplasia and hypoplasia of the lungs, congenital cystic disease of the lungs, emphysema and congenital broncho-pulmonary cysts. Sequestration has no communication with the normal tracheo-bronchial tree and its blood supply comes from the systemic circulation (1, 2, 5).

Such blood supply was first described by Huber in 1777, and the term “pulmonary sequestration” and its different types was proposed by Pryce in 1946.

There are two types of pulmonary sequestration: intralobar and extralobar. There are many differences between these two types of sequestrations: localization, blood supply (arterial and venous) and associated anomalies (1, 2, 4, 5, 7). The intralobar sequestration (20 patients in our series) appears as a mass within a normal lung. The lung parenchyma is atelectatic, and cysts and bronchiectasis within the sequestration have no connection with the rest of the lung. If such a connection exists, there is risk of secondary cystic or bronchiectatic infection. This type of chronic infection may lead to bronchiectasis formation within the surrounding normal lung parenchyma. The intralobar sequestration is usually supplied by a relatively large vessel, approximately 15 mm in diameter, branching from the descending aorta, the abdominal aorta, the intercostal arteries or the abdominal aortic arch (7). In our series of 20
<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Preoperative diagnosis</th>
<th>Side</th>
<th>Type</th>
<th>Localization</th>
<th>Vascularization</th>
<th>No of vessels</th>
<th>Operation</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/63</td>
<td>44</td>
<td>M</td>
<td>bronchiectasis</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>3</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>2/82</td>
<td>18</td>
<td>F</td>
<td>abscess</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>3/88</td>
<td>27</td>
<td>M</td>
<td>emphysematous bula</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>4/90</td>
<td>24</td>
<td>M</td>
<td>cyst</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>5/03</td>
<td>52</td>
<td>M</td>
<td>tumor</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>3</td>
<td>wedge resection</td>
<td>none</td>
</tr>
<tr>
<td>6/82</td>
<td>20</td>
<td>F</td>
<td>emphysematous bula</td>
<td>R</td>
<td>SZ / EL</td>
<td>antero-inferior part of the pleural cavity</td>
<td>abdominal aorta</td>
<td>1</td>
<td>sequestrectomy</td>
<td>respiratory insufficiency</td>
</tr>
<tr>
<td>7/98</td>
<td>17</td>
<td>M</td>
<td>tumor</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>abdominal aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>8/99</td>
<td>43</td>
<td>M</td>
<td>tumor</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>wedge resection</td>
<td>none</td>
</tr>
<tr>
<td>9/72</td>
<td>22</td>
<td>F</td>
<td>tumor</td>
<td>L</td>
<td>SZ / EL</td>
<td>posterior mediastinum, supradiaphragmatic</td>
<td>thoracic aorta</td>
<td>1</td>
<td>sequestrectomy</td>
<td>none</td>
</tr>
<tr>
<td>10/72</td>
<td>14</td>
<td>F</td>
<td>cyst</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>abdominal aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding + phrenic nerve palsy</td>
</tr>
<tr>
<td>11/99</td>
<td>59</td>
<td>F</td>
<td>tumor</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>abdominal aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>12/01</td>
<td>39</td>
<td>F</td>
<td>tumor</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>abdominal aorta</td>
<td>1</td>
<td>wedge resection</td>
<td>none</td>
</tr>
<tr>
<td>13/99</td>
<td>19</td>
<td>F</td>
<td>tumor</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>2</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>14/87</td>
<td>20</td>
<td>M</td>
<td>testicular cancer meta</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>2</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>15/01</td>
<td>16</td>
<td>F</td>
<td>sequester</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>none</td>
</tr>
<tr>
<td>16/95</td>
<td>21</td>
<td>F</td>
<td>tumor</td>
<td>L</td>
<td>SW / IL</td>
<td>superior lobe</td>
<td>thoracic aorta</td>
<td>4</td>
<td>segmentectomy 1+2</td>
<td>none</td>
</tr>
<tr>
<td>17/84</td>
<td>25</td>
<td>F</td>
<td>sequester</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>2</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>18/74</td>
<td>16</td>
<td>F</td>
<td>sequester</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>19/81</td>
<td>21</td>
<td>F</td>
<td>cyst</td>
<td>R</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>abdominal aorta</td>
<td>2</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>20/71</td>
<td>42</td>
<td>F</td>
<td>tumor</td>
<td>R</td>
<td>SW / IL</td>
<td>middle lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>middle lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>21/76</td>
<td>32</td>
<td>M</td>
<td>sequester</td>
<td>L</td>
<td>SW / IL</td>
<td>lower lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower lobectomy</td>
<td>intraoperative bleeding</td>
</tr>
<tr>
<td>22/64</td>
<td>40</td>
<td>F</td>
<td>cyst</td>
<td>L</td>
<td>SZ / EL</td>
<td>supradiaphragmatic posterior mediastinum</td>
<td>thoracic aorta</td>
<td>1</td>
<td>sequestrectomy</td>
<td>none</td>
</tr>
<tr>
<td>23/78</td>
<td>13</td>
<td>F</td>
<td>sequester</td>
<td>R</td>
<td>SW / IL</td>
<td>middle lobe</td>
<td>thoracic aorta</td>
<td>1</td>
<td>lower bilobectomy</td>
<td>intraoperative bleeding</td>
</tr>
</tbody>
</table>

M - male, F - female, IL - intralobar sequestration, EL - extralobar sequestration
intralobar sequestrations, 15 presented with blood supply from the thoracic aorta and 5 from the abdominal aorta. In most cases, the vessels were nearly 3mm in diameter.

According to literature, 60% of pulmonary sequestrations are localized to the basal segments, more frequently on the left side. The venous outflow occurs by means of the pulmonary veins (2, 3, 4, 5). In our series, the lower lobe sequestration was found in 17 patients and accounted for 85% of cases of intralobar sequestration. The extralobar sequestration (3 patients in our series) is separated by its own pleura from the rest of the lung and could be localized not only within the pleural cavity, but also within the pericardium, mediastinum and even within the peritoneal cavity. If sequestration is localized within the pleural cavity, it almost always lies between the lung and diaphragm (2, 4, 5, 13). In our series, every extralobar sequestration was observed in this area. Blood supply of an extralobar sequestration comes from the abdominal aorta and venous outflow to the azygos vein on the right side and hemiazygos vein on the left side. Venous flow to the intercostal veins has also been described (3, 7).

Considering two cases of extralobar sequestration in our series, the blood supply came from the thoracic aorta, and in one case, from the abdominal aorta. The supplying arteries constitute the greatest danger during the operation, unless the sequestration is diagnosed before the operation. Since the initial step of each lung resection consists of the division of any adhesions, which are usually avascular, the surgeon may cause serious bleeding while dividing an artery supplying the sequestration that was mistakenly considered to be an adhesion. At this stage of the operation, when the lung is not fully mobilized, exposure of the bleeding vessel may be troublesome, resulting in substantial blood loss. In our opinion, the best way to avoid such complications is too establish proper preoperative diagnosis. Many patients included in this analysis were treated before 1980, when the diagnostic modalities available were much less accurate than now. Nowadays, only a small percentage of lung cancer patients are subjected to surgical intervention without tissue diagnosis of malignancy. In all such cases, the surgeon should be aware of the risk of other unsuspected pathologies including pulmonary sequestration.

Clinical symptoms of pulmonary sequestration are scant and include coughing, expectoration of purulent sputum, hemoptysis (if there is a connection between the sequestration and bronchial tree), as well as chest pain (4, 5, 8, 11). The most common symptoms in our patients were recurrent infections and chest pain, which was consistent with literature. The chest X-ray and CT findings may be misleading. Atelectatic lung parenchyma, usually containing pseudocysts filled with fluid or air may suggest infection, bronchiectasis, abscesses, tumors or a diaphragmatic hernia (3). In our series, the most common preoperative diagnosis was lung cancer. Aortography or selective arteriography are very useful modalities in the diagnostic work-up of pulmonary sequestration patients. Preoperative identification of abnormal vessels is very important and may help avoid massive bleeding during the operation (9). Nowadays, these vessels can be identified by using CT, MRI, and Doppler imaging methods (8).

Extralobar sequestration can coexist with congenital anomalies of the heart, esophagus or diaphragmatic hernias (2, 3, 4, 8, 11).

According to literature, the most common operation is sequestrectomy, which was performed in only three of our patients. This discrepancy may be explained by the fact that most relevant papers published to date concern the pediatric population, while our series included only patients older than 14 years. Considering adults, sequestrectomy is easy to perform in case of extralobal sequestration because the sequestration is separated from the normal lung by its own pleura. In case of intralobar sequestration, post-inflammatory fibrosis may obtuse the intersegmentary planes making sequestrectomy difficult. Often, a lobectomy is performed (in our study among 20 patients with intralobar sequester in 14 cases lobectomy was performed). Another cause of the substantial percentage of lobectomies in our series was the preoperative diagnosis of lung cancer, usually considered in adult patients with radiological abnormalities. Since lung cancer is extremely rare in children, such radiographic findings suggest developmental anomalies, rendering correct diagnosis easier.

Over time, refinement of the videothoracoscopic technique has led to a larger number of minimally invasive lung resections being per-
formed. The VATS sequestrectomy was described by several authors (6, 14, 15).

The basic difficulty in the treatment of pulmonary sequestration consists of proper preoperative diagnosis, which is often missed in adults because the surgeon focuses on the diagnosis of lung cancer and other common diseases, while such rare diseases like pulmonary sequestration are rarely considered.

This study was based on the retrospective analysis of patients treated over a period of 41 years. Some of the medical records were lost and we were unable to find the pertinent information. We are aware that this is a serious drawback of the publication but such situations are inevitable when one attempts to perform a collective analysis of many cases of an uncommon disease.

**CONCLUSIONS**

1. Pulmonary sequestration in adults is difficult to diagnose before the operation, even when computer tomography is performed.
2. Intralobar sequestration is much more common (87%) than extralobar sequestration, and is most often localized to the basal segments, equally often on the right and left sides.
3. The greatest danger during the operation is major bleeding from the supplying artery.
4. Prognosis after the operation is favorable.

**REFERENCES**