Extralobar Sequestration Presenting as Massive Hemothorax*

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Pulmonary extralobar sequestration is a rare anomaly, usually diagnosed during the first months of life. A case of extralobar pulmonary sequestration in an adult, manifesting itself as massive hemothorax, is presented. (CHEST 1996; 109:843-45)

Key words: cystic adenomatoid malformation; extralobar sequestration; hemothorax

Bronchial sequestration is a relatively rare anomaly, diagnosed usually at infancy. Of the two forms of bronchopulmonary sequestration, intralobar and extralobar, the latter is less frequently encountered. We describe an adult with extrapulmonary sequestration in whom massive spontaneous hemothorax was the presenting symptom.

Case Report

A 50-year-old healthy man was referred to the hospital because of a complaint of pain in the lower area of the left side of the chest and the left area of the hypochondrium. 2 h following ergometry performed as part of a periodic checkup. Physical examination revealed only a slightly distended abdomen. The patient was admitted for observation. At the time of admission, the ECG was normal and remained so. A chest x-ray film demonstrated a moderate pleural effusion on the left side. Twenty-four hours later, dyspnea and elevation of body temperature to 38°C developed, and a dull percussion sound with no air entry over the lower part of the left lung was noted. Another chest x-ray film indicated a significant increase in the amount of the pleural effusion.

Closed-tube thoracostomy yielded non-clotting blood. Since the patient was experiencing chest pain and hemothorax on the left side, the possibility of dissection of the thoracic aorta was considered. Transesophageal echocardiography did not indicate any abnormal cardiac finding or evidence of aortic dissection. However, a fairly large amount of pleural effusion was present on the left side, with a large partially moving mass within the fluid. A CT scan of the chest indicated the presence of a large left-sided pleural effusion surrounding a mass-like lesion, containing fluid with irregular, high-density foci in its periphery (Fig 1). Angiography of the thoracic aorta, celiac trunk, and left lower intercostal arteries showed no evidence of dissecting aneurysm, bleeding, or abnormal vessels supplying the lung tissue. The chest tube drained 2,800 mL bloody effusion within 48 h. Significant clinical improvement followed. However, following another episode of chest pain, a subsequent CT scan of the chest showed a marked increase in the size of the mass, with a minimal amount of fluid surrounding it.

Subsequently, the patient underwent an explorative thoracotomy. A cystic mass was found at the lower aspect of the left lung.

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References

Discussion

Balloon tamponade is a useful technique for treating hemothysis. But inserting a balloon catheter through the working channel of a flexible bronchoscope, as previously reported,1,2 has some demerits. First, insertion of a balloon catheter into the working channel reduces the suction capacity and thus makes it difficult to attain a good endoscopic view during the procedure, limiting the use of the technique to patients with slow bleeding. Second, a special balloon catheter with reattachable valve3 or complicated maneuver1 is necessary to remove the bronchoscope and then inflate the balloon. The modified insertion technique provides a better view during the procedure because only a guide wire needs to be inserted into the working channel, rendering minimal influence to suction capacity; this means that the technique could be applied to more massive bleeding cases. It requires no special catheter or complicated maneuver and is technically easy. We believe modified balloon tamponade is the preferred procedure in hemothysis patients whose bleeding is located at the lobar level or beyond.

References

Figure 2. Intercostal arteriography performed after the bleeding had been controlled. Tumor vessels draining into pulmonary vein are present.
adherent to the diaphragm. It contained brown odorless fluid. Vascular connection to the thoracic aorta was found and divided. The lesion was resected. The pathologic finding consisted of a mass 10 cm in diameter containing small cyst-like structures. The histologic features established the diagnosis of extralobar lung sequestration exhibiting cystic adenomatoid malformation (CCAM) (Fig 2). Postoperative recovery was uneventful.

**DISCUSSION**

Extralobar sequestration, an entity in which an abnormal segment of the lung enclosed within its own pleural membrane is completely separated from the tracheobronchial tree, accounts for 25% of all pulmonary sequestrations, occurring predominantly in men with a ratio of 3 to 4:1.\(^1\)\(^-\)\(^4\) Its location, usually related to the left hemidiaphragm, may vary between the lower lobe and the diaphragm, in the mediastinum, within the lung, in the pleural or pericardial spaces, or in the retroperitoneum.\(^1\)\(^-\)\(^5\)

Arterial blood supply to the sequestered lung may originate from the thoracic or abdominal aorta (>80%); from smaller arteries, such as splenic, gastric, intercostal and subclavian (15%); or from branches of the pulmonary artery and systemic vessels (5%). The venous drainage usually is connected to the azygos system; however, occasionally it can be connected to the intercostal, portal, esophageal, or adrenal veins. Pulmonary venous drainage also is possible.\(^4\)

Intriguing enough, the feeding vessel was not identified on the angiogram, nor was it detected by an independent retrospective review of the films. One could only speculate whether this was the result of vasospasm, a sharp angle of exit from the aorta, or due to the fact that no technique is infallible.

Extralobar sequestration, a developmental lesion, usually presents during the first 6 months of life (61%), often with dyspnea, cyanosis, and feeding difficulties.\(^2\) Recurrent pulmonary infections (as in intralobar sequestration) or gastrointestinal symptoms in the presence of a communication with the gastrointestinal tract (bronchopulmonary foregut malformation) are uncommon.

Very few cases of CCAM—a developmental anomaly characterized by multiple cysts of varying sizes as well as a network of interconnecting spaces—were reported in the adult.\(^6\) Extralobar pulmonary sequestration that histologically displayed the features of CCAM type 2 was reported in 14 cases, 9 of which were thoracic and 5 of which were abdominal.\(^7\) Clinical features of extralobar sequestration alone or containing CCAM type 2 do not differ significantly.\(^7\)

The occurrence of hemothorax unrelated to acute trauma is quite rare. The major diagnostic categories to be considered include defects of coagulation, malignant tumors, and primary vascular events—such as aortic dissection and pulmonary vascular bleeding. Rarely, infections, endometriosis, and costal exostoses can cause hemothorax.\(^8\)

In this patient, massive hemothorax was the presenting sign of extralobar pulmonary sequestration.

The diagnosis of extralobar sequestration in adults is rather difficult, perhaps because it is rarely thought of.\(^9\) It is intriguing that of 66 patients with extralobar sequestration who underwent surgery in the series of Savic et al.\(^2\) a correct preoperative diagnosis was made only in 6. Spontaneous hemothorax from bronchopulmonary sequestration is exceedingly rare and only has been reported in five cases.\(^10\)\(^-\)\(^12\)

To our best knowledge, all previously reported cases with spontaneous hemothorax occurred with intralobar sequestration.

In summary, a case of massive hemothorax resulting from extralobar sequestration diagnosed at thoracotomy was pre-
Use of Somatostatin Analog for Localization and Treatment of ACTH Secreting Bronchial Carcinoid Tumor*

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A 29-year-old woman presenting with an ectopic adrenocorticotropin hormone syndrome and a nodule of the upper lobe of the left lung was explored by intraabdominal somatostatin receptor scintigraphy. This showed a pathologic uptake by the nodule. Treatment with octreotide resulted in the rapid control of hypercortisolism prior to surgery. (CHEST 1996; 109:845-46)

Key words: bronchial carcinoid; Cushing’s syndrome: ectopic ACTH secretion; somatostatin receptor scintigraphy

Carcinoid tumors are responsible for 40% of cases of adrenocorticotropin hormone (ACTH) dependent Cushing’s syndrome. Bronchial carcinoid tumors usually are small (0.3 to 1.3 cm) and are difficult to localize. In addition, hypercortisolism must be controlled prior to surgery. In this report, we describe, in a patient with ectopic ACTH syndrome, the rapid normalization of hypercortisolism by octreotide-only treatment, following the localization of a bronchial carcinoid tumor by octreotide scintigraphy. Octreotide is a somatostatin analog. This case report suggests that when a paraneoplastic ACTH-dependent hypercorticism is suspected, labeled pentetreotide scintigraphy may allow the exact localization of the tumor. Furthermore, the uptake of radioactive somatostatin (octreotide [Sandostatin; Sandoz; Bäle, Switzerland]) suggests the possibility of treating patients with this drug.

Table 1—Biological Evaluation Prior to Treatment, on Day 24 of SMS Treatment, and 1 Week and 2 Months Postsurgery*

<table>
<thead>
<tr>
<th>Values</th>
<th>Initial Value</th>
<th>Treatment Day 24</th>
<th>1 Week Postsurgery</th>
<th>2 Months Postsurgery</th>
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<tbody>
<tr>
<td>UFC, nmol/24 h</td>
<td>806 (260)</td>
<td>13</td>
<td>24</td>
<td>33</td>
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<tr>
<td>Plasma cortisol, nmol/L</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>8 h</td>
<td>1,493 (220-610)</td>
<td>116</td>
<td>123</td>
<td>151</td>
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<tr>
<td>16 h</td>
<td>1,103 (100-290)</td>
<td>211</td>
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<td>Dexamethasone test1</td>
<td>1,166</td>
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<tr>
<td>Plasma ACTH, pg/mL, basal</td>
<td>214 (9-52)</td>
<td>101</td>
<td>18</td>
<td>32</td>
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<tr>
<td>CRF test2</td>
<td>246</td>
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<tr>
<td>Plasma LPH, pg/mL</td>
<td>1,674 (46-124)</td>
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<td>66</td>
<td></td>
</tr>
</tbody>
</table>

* Surgery took place on treatment day 60. Normal value ranges are shown in parentheses. UFC-urinary free cortisol; LPH-lipotropin hormone; pg-pigogram.

1 2-day high-dose dexamethasone suppression test (8 mg dexamethasone/d).

2 Corticotropin-releasing factor, 100 μg administered intravenously.