Chyloptysis after Ligation of the Thoracic Duct

Shinpei Kato, Hiroki Umezawa, Toshiaki Yano, Takashi Ogasawara, Norio Kasamatsu and Ikko Hashizume

Abstract

Chyloptysis is a very rare clinical finding. We describe a 44-year-old man who presented with cough and milky-white sputum. Fiberoptic bronchoscopy revealed white sputum, which originated from the right B6 bronchus. The finding of elevated triglyceride levels in his sputum led to the diagnosis of chyloptysis. He had a surgical history of ligation of the thoracic duct for idiopathic chylopericarditis 7 years-previous. He also suffered from postoperative bilateral empyema. Since then, his pleural cavity has been adhered bilaterally. It is thought that his abnormal postoperative lymphatic flow caused the chyloptysis.

Key words: chyle, chyloptysis, chylothorax, chylopericarditis

However, he complained of dyspnea 1 month later and visited our hospital again. We detected hypoxia (SpO₂ 88% in room air) and worsened appearance of the chest radiograph. It was thought that leakage of chyle into the bronchus had worsened and thus he was readmitted. Conservative treatment with fasting and total parenteral nutrition (TPN) improved both dyspnea and chyloptysis within a few days. Fasting was discontinued 10 days after his admission. At first, we served a fat-free diet and gradually increased the fat content. The ground-glass opacity on chest radiograph gradually disappeared and he was discharged after receiving nutritional instructions. He is now eating a low-fat diet (up to 30 g fat per day) and is doing well at 9 months after discharge (Fig. 6).

Discussion

Chyloptysis is a very rare clinical finding and only a few cases have been reported in the literature (1-9) (Table). The patient complained of expectoration of milky-white and foul-tasting sputum. To confirm chyle, it is necessary to demonstrate triglyceride levels of more than 110 mg/dL and predominance of lymphocytes in the cytological analysis. Some patients expectorating bronchial casts have been reported (2, 3, 6). Causes include acquired or congenital lymphangiectasia, lymphangiomatosis, yellow nail syndrome, and Behçet’s disease (Table). Two mechanisms have been postulated for the pathogenesis of chyloptysis: 1) congenital or acquired incompetence of the lymphatic valves which causes retrograde flow from the thoracic duct into the bronchomediastinal trunks and peribronchial lymphatic plexus and 2) occurrence of bronchopleural fistula in the context of chylous effusion (7). The cause in the present case would be the former.

In general, lymphangiography is usually performed to prove an abnormal flow in lymphatic vessels. However, pneumonia caused by contrast medium has been reported in a previously published case of chyloptysis (7). In 1999, Hayashi and Miyazaki reported the method by which to delineate the thoracic duct by non-contrast MRI (10). Using MRI, we could noninvasively demonstrate a backflow of chyle into the right peribronchial lymphatics. In addition, we found an extension of lymphatic vessels by TBLB. These findings suggested that pulmonary lymphangiectasia caused chyloptysis.
Since the patient’s thoracic duct had been ligated 7 years previously, his lymphatic flow was drained through a collateral route, the capacity of which may be limited compared to healthy individuals. Due to his eating habit (high fat content) the lymphatic flow had increased. However, the abnormal flow of chyle could not leak into the pleural cavity because it had been adhered by postoperative bilateral pleural empyema. The high pressure of lymphatic flow caused a dysfunction of the lymphatic valves and resulted in a retrograde flow of chyle into the peribronchial pulmonary lymphatics. Eventually, the overloaded lymphatics ruptured into the bronchial lumen and the patient developed chyloptysis.

Patients who have been diagnosed as having chyloptysis can recover with a conservative very low-fat diet (1, 6) or medium-chain triglyceride diet (3, 5, 6). A low-fat intake is thought to reduce the flow of lymph and the size of the lymphatic channels. Medium-chain triglyceride is digested
References

1. Sanders JS, Rosenow EC 3rd, Piehler JM, Gloviczki P, Brown

The authors state that they have no Conflict of Interest (COI).

Table. Cases of Chyloptysis Previously Reported in the Literature

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>History</th>
<th>Diagnosis</th>
<th>Treatment</th>
<th>Prognosis</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>73</td>
<td>F</td>
<td>Unremarkable</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>Low fat diet, Ligation of the thoracic duct</td>
<td>Asymptomatic for 3 years</td>
<td>(1)</td>
</tr>
<tr>
<td>2</td>
<td>54</td>
<td>M</td>
<td>Unremarkable</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>Observation</td>
<td>Spontaneous improvement</td>
<td>(2)</td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>F</td>
<td>Complex cardiac anomaly</td>
<td>Lymphangiectasis due to increased venous pressure after Fontan operation</td>
<td>Heart transplantation, MCT</td>
<td>Asymptomatic for 10 months</td>
<td>(3)</td>
</tr>
<tr>
<td>4</td>
<td>40</td>
<td>M</td>
<td>Behçet's disease</td>
<td>Pulmonary involvement in Behçet's disease</td>
<td>Intensive care</td>
<td>Died of cardiac tamponade caused by chylopericarditis</td>
<td>(4)</td>
</tr>
<tr>
<td>5</td>
<td>28</td>
<td>M</td>
<td>Unremarkable</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>MCT</td>
<td>Asymptomatic for 3 years</td>
<td>(5)</td>
</tr>
<tr>
<td>6</td>
<td>70</td>
<td>M</td>
<td>Unremarkable</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>Low fat diet, MCT</td>
<td>Resolved over a 4-month period</td>
<td>(6)</td>
</tr>
<tr>
<td>7</td>
<td>39</td>
<td>M</td>
<td>OSA, Barrett's esophagus</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>Ligation of the thoracic duct</td>
<td>Resolved over a 6-month period</td>
<td>(7)</td>
</tr>
<tr>
<td>8</td>
<td>82</td>
<td>M</td>
<td>COPD, DM, HT</td>
<td>Lymphatic obstruction in yellow nail syndrome</td>
<td>Died before treatment</td>
<td>Suddenly died of AMI</td>
<td>(7)</td>
</tr>
<tr>
<td>9</td>
<td>73</td>
<td>F</td>
<td>Unremarkable</td>
<td>Lymphangiectasis due to incompetence of the lymphatic valves</td>
<td>Ligation of the thoracic duct</td>
<td>Asymptomatic for 3 years</td>
<td>(7)</td>
</tr>
<tr>
<td>10</td>
<td>20</td>
<td>F</td>
<td>Unremarkable</td>
<td>Right middle lobe syndrome due to aberrant lymphatic drainage</td>
<td>Right middle lobe resection, Octreotide, Ligation of the thoracic duct</td>
<td>Asymptomatic for 2 years</td>
<td>(8)</td>
</tr>
<tr>
<td>11</td>
<td>59</td>
<td>M</td>
<td>Idiopathic pericardial effusion</td>
<td>Anterior mediastinal mass (lymphangioma) caused by cervical mediastinoscopy</td>
<td>Resection of the anterior mediastinal mass</td>
<td>Asymptomatic for 7 months</td>
<td>(9)</td>
</tr>
</tbody>
</table>


and absorbed directly to the portal vein, which does not affect the lymphatic system. In cases of failed diet therapy, surgical ligation of the thoracic duct can also be considered. It has been reported that the patients who have undergone the medical treatment by ligation of the thoracic duct followed good progress (1, 7, 8).

In summary, we present a case of chyloptysis 7 years after ligation of the thoracic duct. The patient’s chyloptysis and dyspnea were satisfactorily eliminated with conservative low fat diet therapy. Our observations strongly suggest that the overload of lymph flow due to a habitual high fat diet may result in chyloptysis followed by the rupture of peribronchial lymphatics. Although patients with chyloptysis are very rare, physicians should be cognizant of the pathology of chyloptysis in patients suffering from milky-white sputum.

The authors state that they have no Conflict of Interest (COI).

References

1. Sanders JS, Rosenow EC 3rd, Piehler JM, Gloviczki P, Brown