Spontaneous haemothorax associated with von Recklinghausen’s disease: review of occurrence in Japan

Hiroyuki Miura, Osamu Taira, Osamu Uchida, Jitsuo Usuda, Shigeru Hirai, Harubumi Kato

Abstract

The case history is presented of a 61 year old man with von Recklinghausen’s disease who developed a spontaneous haemothorax. In spite of being asymptomatic for five days after drainage, he died as a result of fatal sudden re-bleeding. The post mortem examination showed dissection and rupture of the left subclavian artery. Microscopically, disarrangement of smooth muscle and decrease of elastic fibre was observed in the ruptured artery. Haemothorax in patients with von Recklinghausen’s disease may require thoracotomy, even if the condition of the patient appears to be stable.

(Torax 1997;52:577–578)

Keywords: haemothorax, von Recklinghausen’s disease, neurofibromatosis.

Von Recklinghausen’s disease is transmitted by an autosomal dominant gene and is characterised by multiple skin tumours and abnormal cutaneous pigmentation. Benign schwannomas, osseous system involvement, malignant schwannomas, other types of cancer, and central nervous system involvement are recognised associated complications.1 Vascular lesions are rare but sometimes fatal.2 We report a patient with von Recklinghausen’s disease with haemothorax who was asymptomatic for five days after drainage but who died due to sudden re-bleeding. The cause of arterial rupture was different from those previously reported.

Case report

A 61 year old man presented at our emergency department complaining of left shoulder pain and momentary loss of consciousness. He had suffered from hypertension and atrial fibrillation for 10 years. He had suffered a transient ischaemic attack at the age of 57. Neurofibromatosis was diagnosed when he was aged 50 without known involvement other than cutaneous manifestations of the disease. There was no history of trauma. A chest radiograph showed a left pleural effusion (fig 1). A plain computed tomographic (CT) scan showed a left pleural effusion with high density areas suspicious of blood. There was no evidence of an aneurysm. Numerous subcutaneous tumours were seen on the face, trunk, and limbs. There were no abnormal neurological findings although there was tachycardia (170/min) and the blood pressure was 120/80 mm Hg. Atrial fibrillation was observed on the ECG and the haemoglobin level was 11.2 g/dl.

An intercostal drainage tube was inserted and approximately 1000 ml of blood was removed. After drainage the serous bloody discharge decreased to below 100 ml per day. Radiologically there was no evidence of an increase in the pleural effusion.

Five days after admission he suddenly collapsed and his blood pressure was unrecordable. Although cardiopulmonary resuscitation was immediately performed, he died. Blood did not flow out from the chest drainage tube. The chest radiograph taken two hours before cardiac arrest also showed no increase in the pleural effusion.

The post mortem examination revealed about 4000 ml of blood in the thoracic cavity. The tumorous lesion with haematoma, about 5 cm in diameter, in the apex was thought to have caused the dissection and rupture of the subclavian artery. Microscopically, disarrangement of smooth muscle and decrease of elastic fibres were observed in the ruptured artery. Proliferation of fibroblasts, indicating that the rupture had occurred several times, was observed at the ruptured portion. Neurofibromas were not seen at the ruptured portion.

Discussion

The incidence of vascular lesion in patients with von Recklinghausen’s disease has been reported to be only 3.6%.1 Arterial lesions are classified into five types:34 (1) the pure intimal form, (2) the intimal-aneurysmal form, (3) the periarteriolar nodular form, (4) the advanced intimal form, and (5) the epithelioid form. These lesions involved small vessels less than 1 mm in diameter. Greene et al5 investigated the arterial lesions associated with neurofibromatosis and described two categories in

Figure 1 Chest radiograph on arrival showing a left pleural effusion.
Table 1  Spontaneous haemothorax in patients with neurofibromatosis reported in Japan

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Location</th>
<th>Ruptured artery</th>
<th>Operation</th>
<th>Prognosis</th>
<th>Neurofibromatous invasion in blood vessels</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>55</td>
<td>M</td>
<td>Right Intercostal</td>
<td>Resection</td>
<td>Alive</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>43</td>
<td>F</td>
<td>Left Internal thoracic</td>
<td>Ligation</td>
<td>Dead</td>
<td>−**</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>32</td>
<td>F</td>
<td>Right Subclavian</td>
<td>Ligation</td>
<td>Dead</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>38</td>
<td>F</td>
<td>Left Subclavian</td>
<td>Ligation</td>
<td>Alive</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>37</td>
<td>F</td>
<td>Left Subclavian</td>
<td>Thoracotomy</td>
<td>Dead</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>48</td>
<td>F</td>
<td>Right Unknown</td>
<td>Resection and implantation of descending aorta</td>
<td>Alive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>31</td>
<td>F</td>
<td>Right Intercostal</td>
<td>Ligation</td>
<td>Alive</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>55</td>
<td>F</td>
<td>Left Subclavian</td>
<td>Thoracotomy</td>
<td>Dead</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>53</td>
<td>M</td>
<td>Right Innominate</td>
<td>Resection and aorta-annominate bypass</td>
<td>Dead</td>
<td>−</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>48</td>
<td>F</td>
<td>Left Internal thoracic</td>
<td>Resection</td>
<td>Alive</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>67</td>
<td>F</td>
<td>Left Intercostal</td>
<td>Haemostasis</td>
<td>Alive</td>
<td>?</td>
<td></td>
</tr>
<tr>
<td>12*</td>
<td>61</td>
<td>M</td>
<td>Left Subclavian</td>
<td>Thoracotomy</td>
<td>Dead</td>
<td>−</td>
<td></td>
</tr>
</tbody>
</table>

NP = not performed.

* Present case.
** Neurofibromatous tissue surrounded ruptured artery.

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2 Leier CV, Dewan CJ, Anastasia LF. Fatal hemorrhage as the subclavian artery were the most common complication of neurofibromatosis. Vasc Surg 1972;6:98–101.
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