Retroperitoneal Hematoma Induced by Von Recklinghausen Disease
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Abstract: A 51-year-old male experienced left lower abdominal pain while running. He was transferred to a local acute critical care center. On arrival, he was in a state of shock. He had also multiple café au lait macules and subcutaneous nodules, suggesting neurofibromatosis-1 (NF1). His shock state was improved by the infusion of lactate ringer solution. Truncal body enhanced computed tomography (CT) revealed retroperitoneal hematoma with a sacral body tumor and enriched, enlarged tumor-nourishing blood vessels without extravasation of contrast medium. As his vital signs were stable and there was no sign of active bleeding, conservative therapy was selected. After 10 days, his condition remained unchanged without transfusion, and the volume of the retroperitoneal hematoma had decreased, so he was temporarily discharged and referred to a special tumor orthopedist to evaluate the sacral tumor. As patients with NF1 are occasionally complicated with lethal hemorrhaging in the head, neck, chest and abdomen, including the retroperitoneal space, patients with unstable circulation should be evaluated by whole-body CT to detect the bleeding source in addition to the distribution of asymptomatic tumors.

Keywords: Retroperitoneal hematoma; von Recklinghausen disease; observation.

INTRODUCTION
Von Recklinghausen's disease or neurofibromatosis-1 (NF1) is an autosomal dominant inheritable neurocutaneous disorder manifested by developmental changes in the nervous system, bones, and skin.

The gene for NF-1 is located at chromosome 17q11.2, encoding the gene product neurofibromin[1]. The diagnosis of NF1 is mainly clinical and is based on the established clinical criteria, which require two or more of the following conditions to be met: ≥6 café au lait macules, ≥2 cutaneous/subcutaneous neurofibromas or 1 plexiform neurofibroma, axillary or groin freckling, optic pathway glioma, ≥2 Lisch nodules, bony dysplasia and a first-degree relative with NF1 [2]. The course of NF1 varies considerably among patients, but the majority has a benign disease course without developing major complications [3]. However, some patients with NF1 suddenly collapse due to a number of causes, with the most frequent reasons being intracranial tumors, vasculopathy affecting the coronary arteries and spontaneous hemotherorax, the latter being likely associated with the sudden rupture of dysplastic arteries that have been described in NF1.[4]

We herein report a rare case of retroperitoneal hematoma abruptly induced by NF1.

CASE PRESENTATION
A 51-year-old male felt left lower abdominal pain while running. As he could not stand, his colleagues called an ambulance. When emergency medical technicians checked him, he was in a state of shock (blood pressure 88/66 mmHg) with paleness and a cold sweat. He was transferred to a local acute critical care center. On arrival, he had a blood pressure of 86/66 mm Hg, a heart rate of 81 beats per minute (BPM), an SpO2 of 97% under oxygen mask with 3 L per minute and a body temperature of 35.1 °C. He had mild tenderness at the left lower abdomen without peritoneal stimulation signs. He also had multiple café au lait macules and subcutaneous nodules, suggesting NF1. One of his three sons had died due to malignant intracranial schwannoma, and the remaining two also had multiple café au lait macules; they were being followed under suspicion of NF1 by a pediatric physician. He had been diagnosed with a sacral tumor in the past, but he had ignored it until he was asked about it by emergency physicians. He had undergone head magnetic resonance imaging in the past year, the findings of which were negative. As he met the clinical diagnostic criteria, he received a diagnosis of NF1.

His shock state was improved by the infusion of lactate ringer solution. Truncal body enhanced computed tomography (CT) revealed retroperitoneal
hematoma with sacral body tumor and enriched, enlarged tumor-nourishing blood vessels without extravasation of contrast medium (Figure 1). Pelvic magnetic resonance imaging revealed the tumor and hematoma more clearly than CT (Figure 2). A biochemical analysis of the blood showed no specific findings. As this hospital was not caring for the patient, he was transferred to our hospital the same day. His vital signs were stable, and there were no signs of active bleeding, so conservative therapy was selected. After 10 days, his condition remained unchanged without transfusion, and the volume of retroperitoneal hematoma had decreased, so he was temporarily discharged and referred to a special tumor orthopedist to evaluate the sacral tumor.

Fig-1: Truncal body enhanced computed tomography (CT) on arrival at an acute critical care center. CT reveals retroperitoneal hematoma (triangle) with a sacral body tumor (star) and enriched, enlarged tumor-nourishing blood vessels (arrow) without extravasation of the contrast medium

Fig-2: Pelvic enhanced magnetic resonance imaging (MRI) on arrival at an acute critical care center. MRI reveals retroperitoneal hematoma (triangle) with sacral body tumor (star) ; B- bladder
Table 1: A summary of retroperitoneal hematoma by von Recklinghausen disease

<table>
<thead>
<tr>
<th>No.</th>
<th>Year</th>
<th>Reporter</th>
<th>Sex</th>
<th>Age</th>
<th>Diagnosis</th>
<th>Source</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1975</td>
<td>Fye</td>
<td>M</td>
<td>38</td>
<td>Operation</td>
<td>Renal subcapsular a.</td>
<td>Observation</td>
<td>Survival</td>
</tr>
<tr>
<td>2</td>
<td>1978</td>
<td>Stevenson</td>
<td>M</td>
<td>39</td>
<td>Operation</td>
<td>Root of Mesocolon</td>
<td>Ligation</td>
<td>Survival</td>
</tr>
<tr>
<td>3</td>
<td>1982</td>
<td>Keenan</td>
<td>F</td>
<td>26</td>
<td>Operation</td>
<td>Inferior venal cava</td>
<td>Operation</td>
<td>Death</td>
</tr>
<tr>
<td>4</td>
<td>1983</td>
<td>Shelton</td>
<td>M</td>
<td>59</td>
<td>Operation</td>
<td>Tumor feeding a.</td>
<td>Operation</td>
<td>Death</td>
</tr>
<tr>
<td>5</td>
<td>1988</td>
<td>Wan</td>
<td>M</td>
<td>45</td>
<td>Operation</td>
<td>Erosion of the vessel</td>
<td>Operation</td>
<td>Death</td>
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<tr>
<td>6</td>
<td>1990</td>
<td>Poston</td>
<td>F</td>
<td>48</td>
<td>Operation</td>
<td>Gastroduodenal a.</td>
<td>Ligation</td>
<td>Survival</td>
</tr>
<tr>
<td>7</td>
<td>1997</td>
<td>Akuzawa</td>
<td>M</td>
<td>29</td>
<td>Operation</td>
<td>Adrenal gland</td>
<td>Observation</td>
<td>Survival</td>
</tr>
<tr>
<td>8</td>
<td>1998</td>
<td>Shimizu</td>
<td>M</td>
<td>51</td>
<td>CT</td>
<td>Lumbar artery</td>
<td>Observation</td>
<td>Death</td>
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<tr>
<td>9</td>
<td>2013</td>
<td>Niwa</td>
<td>F</td>
<td>41</td>
<td>CT</td>
<td>Renal artery</td>
<td>Embolization</td>
<td>Survival</td>
</tr>
<tr>
<td>10</td>
<td></td>
<td>Present</td>
<td>M</td>
<td>51</td>
<td>CT</td>
<td>Tumor feeding a.</td>
<td>Observation</td>
<td>Survival</td>
</tr>
</tbody>
</table>

F: Female, M: Male, a.: artery, CT: computed tomography

DISCUSSION
This is an extremely rare case of retroperitoneal hematoma induced by NF1. A PubMed and an Ichushi (Japana Centra Revuo Medicine), which collects summaries of Japanese medical articles, search was performed to identify articles using the key words "von Recklinghausen disease", "hemorrhage" and "retroperitoneum" on December 7, 2017. We found 9 articles which described individuals with retroperitoneal hemorrhaging induced by NF1. We summarized these cases, including the present case, in Table 1 [5-13]. The age ranged from 29 to 59 years, with an average age of 42.7. Male gender was predominant (7 of 10 cases). Seven cases encountered before the year 2000 were recognized during surgery. Before the era of CT, all cases were initially misdiagnosed as retroperitoneal hematoma. However, CT revealed the retroperitoneal hematoma easily. Two patients, who received observed therapy after intra-operative confirmation of retroperitoneal hematoma without signs of active bleeding, obtained a survival outcome. Four cases died, giving a mortality rate of 40%. Three of these four cases underwent surgical operation. As detecting the retroperitoneal bleeding source is difficult by operation, interventional radiological treatments might be useful.

As patients with NF1 are occasionally complicated with lethal hemorrhaging in the head, neck, chest and abdomen, including the retroperitoneal space, patients with unstable circulation should be evaluated by whole-body CT to detect the bleeding source and determine the distribution of asymptomatic tumors [4].

Conflict of interest
The authors declare no conflicts of interest in association with this study.

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