Ileal Shwannoma in which Blood Loss Scintigraphy was Useful for Diagnosis

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Abstract

An 85-year-old woman was hospitalized with severe melena of unknown origin. Upper gastrointestinal (GI) endoscopy and lower GI endoscopy did not detect the origin and we could not establish any diagnosis. To explore the bleeding site, 99mTc-HSA blood loss scintigraphy was performed and a tumor of the small intestine was suspected. Fluoroscopic examination of the small intestine and abdominal CT scan confirmed an ileal tumor measuring 4x3 cm. The mass was a well-demarcated tumor about 80 cm proximal to Bauhin’s valve. Partial resection of the ileum was carried out and the tumor was histologically diagnosed as schwannoma. Thereafter, there has been no recurrence of melena nor metastasis of the tumor. It is thought that blood loss scintigraphy is a useful method for unexplained exacerbation of melena. (Internal Medicine 42: 1178-1182, 2003)

Key words: ileal schwannoma, blood loss scintigraphy, immunohistochemistry, melena, S-100 protein

Introduction

Ileal schwannoma is an extremely rare disease even among small intestine tumors. In Japan, there have been no more than six cases in the literature since a report by Maejima et al in 1952 (1). Endoscopic examination is the primary choice for melena, but when endoscopy does not clearly demonstrate the cause or the procedure is difficult to perform, RI test gastrointestinal scintigraphy is useful. Here, we report a single case of ileal schwannoma in which blood loss scintigraphy was useful for diagnosing the source of hemorrhage causing severe melena, and include a short discussion of the related literature.

Case Report

The patient was an 85-year-old woman who was brought to the hospital on June 24, 1999 due to sudden severe melena. Her blood pressure was 98/52, pulse was 87/min, no abnormal heart rhythm, body temperature was 37.1°C. There was marked anemia in the bulbar conjunctiva, but no sign of jaundice. Superficial lymph nodes were not palpable. There were no abnormal cardiopulmonary findings. The abdomen was flat and there was no pain on pressure. There was no palpable liver mass. Regarding test findings upon hospitalization, RBC was 2.26 million/μl, and hemoglobin was 6.7 g/dl, indicating anemia. BUN was 38.1 mg/dl and CA19-9 was 21.7 ng/ml. Upper gastrointestinal endoscopy demonstrated only atrophic gastritis but no hemorrhaging. Endoscopy in the large intestine demonstrated large dark red blood clots throughout the large intestine and terminal ileum, but the source of the hemorrhaging was unclear. For blood loss scintigraphy using 99mTc-HSA (human serum albumin), 740 MBq of 99mTc-HSA was intravenously infused, and 5 hour later abnormal accumulation was seen in the pelvic lumen (Fig. 1). On abdominal CT, a large mass lesion measuring 3 cm in diameter showing a strong shadow effect was seen at the site identified on scintigram findings (Fig. 2). Imaging of the small intestine showed a large elevated mass 4x3 cm in diameter with a smooth surface at the same site in the ileum, suggesting a submucosal tumor (Fig. 3). Based on these findings, the patient was diagnosed with submucosal ileal tumor in the pelvic lumen, which was considered as the source of the hemorrhage. Surgery was performed on June 28.

The tumor was located 80 cm caudal to the terminal ileum, and was growing outside the tract contralateral to the mesenterium (Fig. 4A). The tumor measured 4x3 cm and had...
Heal Schwannoma

0–5 min 5 hr

Figure 1. 99mTc-HAS scintigraphy showed bleeding from a region in the pelvic lumen (arrow).

Figure 2. A; Abdominal CT demonstrated a round solid tumor (arrow). B; The tumor measuring 3 cm in diameter showed a strong shadow effect at the site identified on scintigram findings.

Figure 3. Enteroclysis showing an intraluminal filling defect in the ileum (arrow).

Discussion

Schwannomas are tumors derived from Schwann cells. These tumors are prone to occur peripherally in the head and limbs, and cause radiating pain or sensory paralysis (2). Although schwannoma rarely occur on the trunk, even a small one in the brain or spinal cord causes serious neurological symptoms. Manifestation in the gastrointestinal tract...
is rare, but when there is an occurrence, the incidence is high in the stomach; a schwannoma originating in the small intestine is rare (3). Since the report by Kondo et al in 1943 (4), there have been only 53 reported cases in Japan. However, only seven of these cases, including the present one, have been limited to the ileum, thus the incidence is indeed rare (5–9) (Table 1). It is thought that this type of tumor originates in Auerbach’s plexus or Meissner’s plexus, the site of the nerve ending in the enteric canal, and most grow outside the canal contralateral to the mesenteric attachment (10). Common symptoms/signs are abdominal pain, palpable mass, and hemorrhaging; intestinal obstruction is rare. A schwannoma is a submucosal tumor with abundant blood vessels. As the tumor grows it becomes exposed on the surface of the mucosa, and is prone to hemorrhage due to necrosis (11).

For preoperative diagnosis, CT, ultrasound, MRI, and angiography are useful for locating the tumor site. CT depicts the tumor growing outside the wall, and displays a high shadow effect (12). Imaging of the small intestine shows findings suggestive of a submucosal tumor, such as extramural pressure or tumor shadow loss (13). Angiography of the superior mesenteric artery shows hypervascular tumor vessels (14).

Blood loss scintigraphy enables the detection of hemorrhaging of at least 0.05 ml per min, and is 10 times more sensitive than angiography. It also enables the detection of intermittent hemorrhaging by obtaining scintigrams over time (15). There have also been cases reported in which blood loss scintigraphy suggested the existence of small intestinal lesions and ultimately enabled jejunal tumor
Heal Schwannoma

Figure 6. Immunohistochemical staining of the tumor cells. A; C-KIT was negative, B; CD-34 was negative, C; α-smooth muscle (SMA) was negative, D; S-100 protein was positive (x100).

Table 1. Reported Cases of Schwannoma of Ileum in Japan

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Chief complaint</th>
<th>Size (mm)</th>
<th>Operation</th>
<th>Character</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maejima (1)</td>
<td>1952</td>
<td>24</td>
<td>M</td>
<td>Abdominal pain</td>
<td>50</td>
<td>Resection</td>
<td>Benign</td>
</tr>
<tr>
<td>Nishimura (5)</td>
<td>1957</td>
<td>50</td>
<td>M</td>
<td>Abdominal pain</td>
<td>110</td>
<td>Resection</td>
<td>Unknown</td>
</tr>
<tr>
<td>Takamura (6)</td>
<td>1965</td>
<td>45</td>
<td>F</td>
<td>Melena</td>
<td>100</td>
<td>Resection</td>
<td>Malignant</td>
</tr>
<tr>
<td>Matsumoto (7)</td>
<td>1975</td>
<td>35</td>
<td>M</td>
<td>Abdominal pain</td>
<td>50</td>
<td>Resection</td>
<td>Benign</td>
</tr>
<tr>
<td>Tanabe (8)</td>
<td>1989</td>
<td>76</td>
<td>M</td>
<td>Melena</td>
<td>32x25x18</td>
<td>Resection</td>
<td>Benign</td>
</tr>
<tr>
<td>Shuto (9)</td>
<td>1996</td>
<td>66</td>
<td>F</td>
<td>Melena</td>
<td>85x76</td>
<td>Resection</td>
<td>Benign</td>
</tr>
<tr>
<td>Our case</td>
<td>1999</td>
<td>85</td>
<td>F</td>
<td>Melena</td>
<td>40x30</td>
<td>Resection</td>
<td>Benign</td>
</tr>
</tbody>
</table>

detection (16). However, it is rare to be able to pinpoint the hemorrhage site, and sometimes the hemorrhaging is stopped at the time of testing and no significant findings can be obtained. The timing of testing is therefore paramount. In the present case, upper and lower gastrointestinal endoscopy revealed no origin and we could not make any diagnosis. In order to explore the bleeding site, °Tc-HSA blood loss scintigraphy was performed and a tumor of the small intestine was detected. In cases where upper or lower gastrointestinal endoscopy cannot identify the source of the hemorrhaging, blood loss scintigraphy is a non-invasive nuclear medical test that is considered a very useful method for locating the source of hemorrhage for GI hemorrhaging of unknown cause.

In histopathological terms, the present case consisted of tumor cells with an oblong or spindle-like shape crossing...
over one another and growing in a tightly packed formation. The lack of regressive degeneration, nuclear mitosis and cellular atypia resulted in benign schwannoma. Immunohistological testing is useful for distinguishing between gastrointestinal stromal tumors (GISTs) and leiomyomas. Approximately 95% of GISTs are positive for C-KIT, while 70–80% are positive for CD34. Virtually 100% of leiomyomas are positive for α-smooth muscle actin, but are usually negative for S-100 protein. S-100 protein is found in schwannoma cell line tumors and is useful for diagnosis (17). In the present case, the tumor stained positive for S-100 protein and negative for C-KIT, CD34 and α-smooth muscle actin. Therefore, we concluded that our patient had a tumor of neural origin, schwannoma.

This disease is treated in principle by surgical resection, with chemotherapy or radiotherapy generally regarded as ineffective. In the case of malignant neurinomas, the prognosis is poor, with hematogenous metastasis to the lungs and liver prone to occur (18). Aggressive tumor resection with due consideration of malignant changes is a mode of treatment. In malignancy, surgical resection is not sufficient even for small tumors, but in the present case of a benign tumor, partial resection of the ileum was thought to have sufficed. There have not been any signs of recurrence to date, four years and seven months postoperatively.

In summary, we described a very rare case of ileal schwannoma which was diagnosed by blood loss scintigraphy. It is thought that blood loss scintigraphy is a useful method for unexplained melena.

References