An Autopsy Case of Periaortic Malignant Fibrous Histiocytoma with Multiple Metastases to the Small Intestine

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Abstract: A 67-year-old woman was admitted to our hospital with severe abdominal pain. Emergency laparotomy was performed, and intestinal perforation was diagnosed. Wide resection of the distal ileum and cecum with ileostomy was performed. Resected specimens revealed three small perforated ulcers and two pedunculated tumors in the small intestine. The ulcers and tumors were diagnosed histopathologically and immunohistochemically as storiform-pleomorphic-type malignant fibrous histiocytoma (MFH). The patient died 1 month after surgery due to liver and renal failure with aortic occlusion. At the primary site, an encapsulated mass (4.0 × 1.0 × 2.0 cm) was situated around the wall of the aorta at autopsy. The grayish white tumor tightly encircled half of the circumference at the level of the celiac axis. It is very unusual for the metastatic foci of MFH to be found before discovery of the primary focus. In addition gastrointestinal (GI) involvement from MFH has rarely been reported. We found 11 cases of metastasis of MFH to the small intestine in the English language literature. Our case is the first reported case of MFH causing perforation of the small intestine.

Key words: malignant fibrous histiocytoma, intestinal metastasis, perforation, aorta

Introduction

Malignant fibrous histiocytoma (MFH) usually metastasizes to the lung, lymph nodes, liver, and bone; gastrointestinal (GI) involvement is rare. We found only 11 cases of metastasis of MFH to the small intestine in the English language literature. We reported a case of periaortic MFH metastases to the small intestine presenting as a perforation. We describe our case and review the previously reported cases of this disease.

Case Report

A 67-year-old woman was admitted to our hospital in July 2005 due to severe, periodic lower abdominal pain that continued for 4 days. She had begun to notice occasional abdominal pain 6 months prior to admission. Physical examination yielded a temperature of 37.4°C, blood pressure of 194/94 mmHg, and pulse rate of 98 beats/min. The patient’s abdomen was symmetrically rounded and diffusely tender with signs of peritoneal irritation.
Fig. 1. Abdominal CT image shows free air around the liver, distention of the small bowel, and a low-density mass at the anterior surface of the abdominal aorta at the level of the celiac axis.

Hematologic study revealed a hemoglobin level of 10.4 g/dl and a white blood cell count of 16,600/mm³. Results of blood chemistry tests, including liver and renal function tests, were within normal limits, but the C-reactive protein level was elevated to 25.1 mg/dl. Abdominal X-ray showed free air beneath the diaphragm, gaseous distention of the small bowel, and multiple air-fluid levels.

Computed tomography (CT) revealed free air around the liver, distention of the small bowel, and ascites. Moreover, CT showed a low-density mass at the anterior surface of the abdominal aorta, but at that time it was thought to be an intraluminal aortic thrombus (Fig. 1). Emergency laparotomy was performed, and a clinical diagnosis of intestinal perforation and peritonitis was made. During surgery, we found purulent fluid in the peritoneal cavity and four perforated ulcers in the small intestine, which were confirmed to be about 80, 160, 260, and 290 cm distal to the ligament of Treitz. Mucosal friability was noticed about 50 and 130 cm distal to the ligament of Treitz. Two pedunculated tumors were present in the ileum. There was no evidence of metastasis to the lymph nodes, liver, or peritoneal cavity. Wide resection of the distal ileum and cecum with ileostomy was performed. The small bowel looked mildly ischemic, extending about 50 cm from the proximal terminal ileum.

**Histopathologic findings**

In the resected intestine, three ulcers and two pedunculated tumors with a central crater were found (Fig. 2). The ulcerous lesions penetrated the entire wall with tumor infiltration (Fig. 3a). Tumor nodules were located in the ileum 5 cm and 34 cm proximal to the ileocecal valve, and measured 30 x 4 mm and 8 x 9 mm. The cut surfaces showed white, firm, solid tumor tissue located mainly in the submucosa, elevating the overlying mucosa and invading the muscularis propria. In the center of the nodules, small ulcers were formed by direct invasion of tumor cells (Fig. 3b). Microscopically, both the ulcerous and polyoid lesions were cellular masses composed of pleomorphic spindle cells forming bundles and
fascicles. Giant cells with bizarre nuclei were predominant, and mitotic activity was high (Fig. 3c).

Specimens were immunohistochemically positive for vimentin and CD68 and negative for cytokeratin, c-kit, CD-34, smooth muscle actin, S-100 protein, and desmin. On the basis of these findings, the tumors were identified as malignant fibrous histiocytoma (MFH) showing a pleomorphic-storiform pattern. It was impossible to distinguish between the primary site and the metastases; and there was no evidence that the primary site was in the extremities or peritoneal cavity.

Post-surgical CT revealed that the size of the aortic mass had not changed. CT angiography of the aorta confirmed the obstruction by revealing a filling defect in the entry of the celiac artery; the superior mesenteric artery and left renal artery were occluded. The superior mesenteric artery was enhanced from 3 cm distal to its origin, and the collateral veins were not enhanced. The liver and left kidney were poorly enhanced due to arterial obstruction. The left kidney was atrophic and much smaller than the right kidney. The patient developed an anastomotic leak and perforation due to small bowel infarction. Her condition deteriorated rapidly, liver and kidney failure ensued, and she died a month after surgery.

Autopsy revealed dark discoloration of the entire ileum and distal jejunum with two perforated lesions of the proximal anastomosis. The remaining portion of the small bowel was necrotic. There was a firm, longitudinally placed, grayish white tumor, 4.0 × 1.0 × 2.0 cm, which appeared to involve the adventitia of the abdominal aorta. The tumor tightly encircled half of the circumference at the level of the celiac axis thickening the aortic wall to 1.0 cm, but the overlying intima seemed largely intact. There was no direct spread to adjacent structures. The root of the celiac artery was occluded by a thrombus, and the superior mesenteric and left renal arteries were also occluded (Fig. 4). Microscopically, the histologic appearance of the periaortic tumor was similar to that seen in the surgical material. The tumor extended mainly from the adventitia through the partly destroyed media to the intima. Elastic van-Gieson staining revealed that the tumor had infiltrated the intraluminal surface of the abdominal aorta, the intima of the celiac artery, the adventitia of the superior mesenteric artery, and the media of the left renal artery. Degenerative necrosis due to thrombosis was seen in the liver and spleen and was considered the cause of death. No tumor embolus or metastasis was seen in any other organ. We realized this to be a case of periaortic MFH with multiple metastases to the small intestine.

Discussion

MFH is a soft tissue sarcoma that occurs mainly in middle-aged individuals. According to an analysis of 200 cases of MFH, the most frequent site is the extremities (lower extremities, 49%; upper extremities, 19%); the second most prevalent site is the abdominal cavity or retroperitoneum (16%) Nicholson. MFH is a high-grade malignancy with an approximately 50% chance of metastasis existing at the time of diagnosis. The most common sites of metastasis are the lungs (25%) and lymph nodes (10%). Tumor metastasis to the gastrointestinal tract is extremely rare.

Only 11 cases of metastasis of MFH to the small intestine have been reported in the English-language literature. These cases, including our case were reviewed and the findings were summarized (Table 1). Seven of the patients were male and five were female. Mean age was 53.3 years (range, 25-75 years). The common primary sites were
Table 1. Reported cases of malignant fibrous histiocytoma metastatic to the small intestine

<table>
<thead>
<tr>
<th>First author</th>
<th>Year</th>
<th>Age / Sex</th>
<th>Primary site</th>
<th>Histological subtype</th>
<th>Symptom</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Terashima*</td>
<td>1983</td>
<td>27 / F</td>
<td>Heart</td>
<td>SP</td>
<td>Anemia</td>
<td>Jejunum</td>
</tr>
<tr>
<td>Misonou†</td>
<td>1990</td>
<td>25 / M</td>
<td>Mediastinum</td>
<td>I</td>
<td>GI bleeding</td>
<td>Jejunum</td>
</tr>
<tr>
<td>Gorman*</td>
<td>1993</td>
<td>47 / F</td>
<td>Thigh</td>
<td>SP</td>
<td>Intussusception</td>
<td>Jejunum</td>
</tr>
<tr>
<td>Kanoh*</td>
<td>1998</td>
<td>45 / M</td>
<td>Back</td>
<td>P</td>
<td>Intussusception</td>
<td>Ileum</td>
</tr>
<tr>
<td>Tamura†</td>
<td>2002</td>
<td>73 / M</td>
<td>Rib</td>
<td>P</td>
<td>GI bleeding</td>
<td>Jejunum</td>
</tr>
<tr>
<td>Atmatzidis†</td>
<td>2003</td>
<td>45 / F</td>
<td>ND</td>
<td>P</td>
<td>Epigastric pain</td>
<td>Small intestine</td>
</tr>
<tr>
<td>Kashima (present case)</td>
<td>2008</td>
<td>67 / F</td>
<td>Periaorta</td>
<td>SP</td>
<td>Perforation</td>
<td>Small intestine</td>
</tr>
</tbody>
</table>

Fig. 2. Macroscopic findings of the resected intestine
Three ulcers (A, B, C) and two pedunculated tumors (D, E) were found. The two polypoid polyps, 8 × 9 mm (D) and 30 × 34 mm (E) located 34 cm (D) and 5 cm (E) proximal to the ileoceleal valve were also found.

Fig. 3. Microscopic examination of the resected intestine
a: An ulcerous lesion was associated with a tumor (A) penetrating the entire wall. (hematoxylin and eosin [H & E])
b: A pedunculated tumor (C) was located mainly in the submucosa with infiltration to the mucosal surface and muscularis propria (H & E; × 400).
c: The tumor is composed of spindle cells in a storiform pattern, with nuclear atypia and mitoses (H & E; × 400).

Fig. 4. Cross-section of the abdominal aorta
Cut surface of the autopsied specimen shows a grayish white tumor and thrombus in the celiac artery, superior mesenteric artery, and left renal artery.
the extremities and the trunk (five cases). Metastatic tumors arose in the jejunum in six patients, the ileum in two patients, and the small intestine in four patients. The common clinical symptoms were abdominal pain, and two major manifestations of GI involvement, GI bleeding (five cases) and intussusception (two cases). The presence of ulceration on the surface of tumors was documented in all cases with GI bleeding. Ours is the first reported case of MFH causing perforation of the small intestine. Metastatic tumors formed as polypoid or pedunculated tumors. The tumors of the seven patients with GI bleeding or intussusception varied in size from 0.8 cm to 13 cm (average size, 6.7 cm). Three cases were of solitary MFHs, but two cases were of multiple MFHs.

With respect to other metastatic foci in the 12 cases, six cases had lung metastases and others had liver, cerebrum, or adrenal metastases. In our case, it is remarkable that intestinal metastases existed without evidence of MFH in the lungs. We propose that our patient had primary retroperitoneal MFH surrounding the aorta and that this metastasized to the small intestine via the abdominal aorta, the celiac artery, and the superior mesenteric artery with an absence of lung metastasis. Moreover, it is highly unusual that metastatic foci were found before discovery of the primary MFH focus. Of 82 patients with metastatic MFH reported by Weiss and Enzinger1, only one had metastases discovered before the primary focus. Of 167 patients with soft tissue MFH reported by Kearney et al12, 10 had pulmonary metastases at the time of diagnosis, but none had metastases predating discovery of the primary tumor12.

The overall prognosis of MFH is poor, especially in cases in which metastatic disease is present. However, Atmatzidis et al11 described a patient who was well 2 years after resection of metastatic foci without discovery of the primary tumors. In our case, periaortic MFH produced ischemic trauma to the small intestine, liver, spleen, and kidney by causing aortic thrombosis; thus, the patient died of multiple organ failure. If the primary lesion had been found earlier and treated, the patient might have survived.

In conclusion, when multiple ulcers and tumors are found in the alimentary tract, clinicians and pathologists must include MFH in the differential diagnosis. A careful search for other lesions should always be performed, especially near the celiac and superior mesenteric arteries.

References


[Received July 12, 2007: Accepted August 8, 2007]