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Mesenteric Desmoid Tumor of the Appendix：A Case Report and Review of the Literature in Japan

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Abstract：Mesenteric desmoid tumors, especially those originating from the mesoappendix, are very rare. We herein report the case of a perforating mesenteric desmoid tumor of the appendix in a patient with no history of either abdominal surgery or Gardner's syndrome. A 24–year–old man was admitted because of acute abdomen. At laparotomy, a perforating huge solid mass arising from the mesoappendix was found. Only 4 cases demonstrating a mesenteric desmoid tumor of the appendix have been previously reported in Japan.

Key words：**Mesenteric tumor, Desmoid tumor, Appendix, Perforating tumor**

Introduction

Desmoid tumors are rare, nonencapsulated, nonmetastasizing tumors[1)2)] which tend to arise from the musculoaponeurotic regions.[1)] The tumor is locally invasive[1)–3)] and, when uncontrolled, it can lead to severe local morbidity, even death.[3)] Due to the pattern of local infiltration, the frequency of local recurrence following an excision is high.[3)]

Mesenteric desmoid tumors are rare entity,[1)2)4)–9)] and its incidence is 0.03% in general population.[4)] Many cases of mesenteric desmoid tumors are found in the small intestinal mesentery, while some cases also occur in the colonic mesentery.[7)8)] Such tumors originating from the mesoappendix are thus considered to be very rare.[4)–9)] We herein present such a case and review 4 other similar cases[4)–9)] previously reported in Japan.

Case report

A 24–year–old man was admitted to Fukuoka University Hospital because of acute abdomen. Abdominal computed tomography (CT) and ultrasonography demonstrated a large well circumscribled, soft tissue mass measuring 20 cm in diameter in the right lower abdomen (Fig. 1). Neither a history of abdominal surgery nor Gardner's syndrome was noted. He had a fever of 39℃ and tarry stool were also present. An exploratory laparotomy revealed a perforating huge solid mesenteric tumor arising from the mesoappendix. Localized peritonitis was also noted at the area surrounding a perforating hole of the tumor. An ileocecal resection including the tumor and adjacent alimentary tracts was thus performed. The tumor measured 20×19×13 cm in size and it was poorly encapsulated. The cut surface was white in color and it had a grisly appearance. Microscopically, there was a proliferation of spindle–shaped fibroblasts within a fibrous stroma. The pathologic diagnosis was a mesenteric desmoid tumor (Fig. 2).

We considered the patient to have a high risk for recurrence. Therefore, he was followed carefully. Abdominal ultrasonography was performed every 2 or 3 months. No abnormal mass was detected by the imaging examination. Fourteen months postoperatively, an abdominal CT indicated a recurrent

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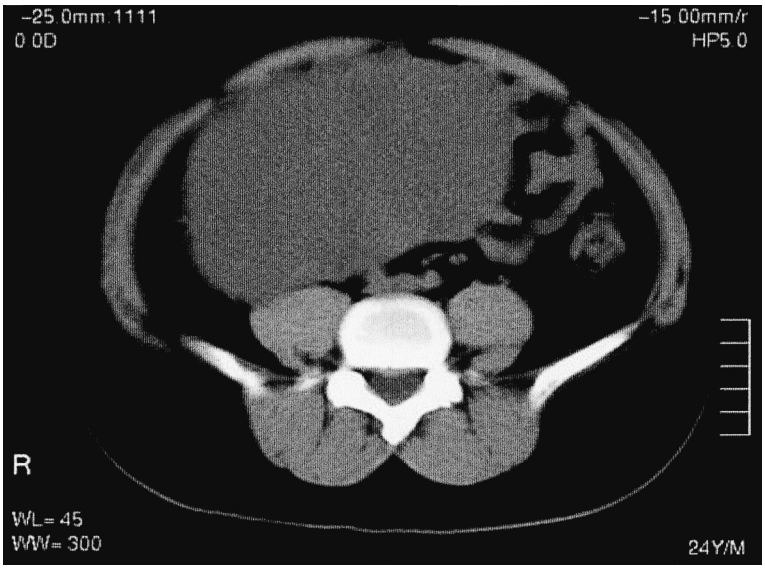

Fig. 1. An abdominal CT scan shows a huge, homogenous, round mass in the right lower abdomen.

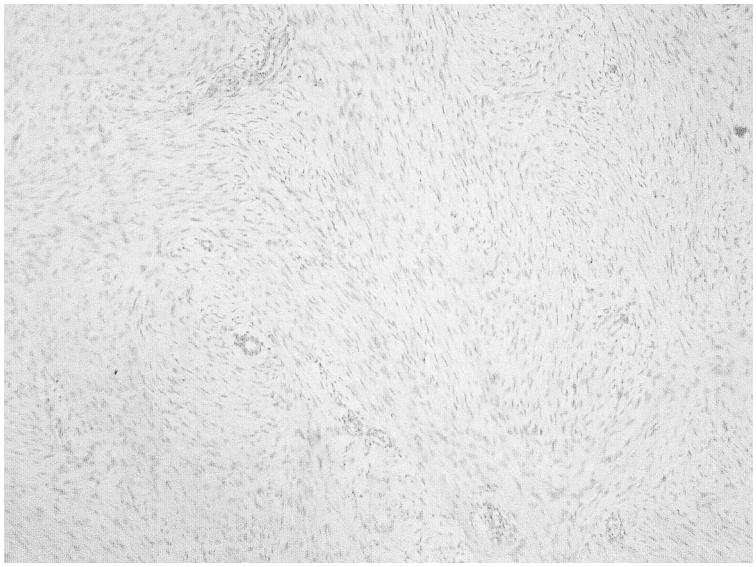

Fig. 2. The tumor was composed of interlacing bundles of proliferated spindle-shaped cells producing an abundant amount of collage.

desmoid tumor with multiple peritoneal dissemination nodules and bilateral hydronephrosis. A reoperation was thus scheduled. A main diffuse large tumor was found in the root of the mesentery. Part of the tumor had invaded the small bowel and was bundled together with the small bowel like a clover, thus resulting in multiple small bowel obstructions. Therefore, an incomplete resection of the tumor with a gross residual and ileocolostomy were performed. Although anti-estrogen (tamoxifen) therapy and radiation therapy were performed, no remission of the rapidly growing desmoid tumor was observed. Two months after the reoperation, the patient died due to massive bleeding in the gastrointestinal tract due to tumor invasion.

Discussion

The mean incidence of desmoid tumors is 2.4 new cases per 1 million population per year[2] and 0.03% to 0.13% of the total number of neoplasms.[10] Of

these desmoid tumors, 8% are mesenteric.[2] Most cases are sporadic, although there is an association with adenomatosis coli (Gardner syndrome), trauma, and estrogens.[11] Mesenteric desmoids frequently arise in young individuals.[4] Shibata et al.[8] revealed that of 75 mesenteric desmoids without a history of Gardner's syndrome in Japan only 3 cases (4.0%) originated from the mesoappendix. Therefore, a desmoid tumor originating from the mesoappendix is very rare. We found only 5 such cases, including the present case, and all 4 of these previously reported cases occurred in Japan (Table).[5-9]

The age of such cases ranged from 17–43 years old, with an average age of 30 years old. They included 3 male and 2 female patients. Three patients demonstrated an intra-abdominal mass with the other two had an intra-abdominal pain (Table). Okawa et al. reviewed 130 mesenteric desmoid tumors for the period of 1960–2004 reported in Japan.[12] The age of the 130 patients ranged from 13–79 years old, with an average age of 44 years old. They included 79 male and 51 female patients. The symptoms and chief complains of these mesenteric desmoid tumors consisted of the following : 79 had an intra-abdominal mass (60.8%), 17 had intra-abdominal pain (13.1%), 10 had abdominal fullness (7.7%), and 24 others (18.5%).

The tumors size varied from 9.5×9.0×6.0 to 20.0×19.0×13.0 cm in diameter, and the observed color ranged from grayish-white to grayish-tan. Some cases of mesenteric desmoid tumors have been described as rapidly growing and all were large masses, ranging from 2.5 to 36.0 cm with an average size of 12.5 cm at the time of surgery.[11,12] Rodriguez et al. reported the greatest diameter of the 25 lesions, which involved the gastrointestinal tract, and ranged in size from 4.6 to 27.0 cm, with an average size of 10.9 cm.[13]

The risk factors for the development of mesenteric desmoids are Gardner's syndrome, abdominal surgery, pregnancy and estrogenic hormones, trauma and an autoimmune response.[1,4-7] Three of the desmoid tumors originating from the mesoappendix, including the present case, had no etiological risk factors, while the two other cases were not well described.

Primary therapy consists of a complete local surgical extirpation of tumor. Two major problems exist with a surgical resection. First, extensive local tissue invasion may lead to technical difficulties thus limiting the complete removal of the entire tumor. Secondly, there is a high incidence of local recurrence despite apparent adequate initial resection. Local recurrence of a desmoid tumor is an indication for a re-excision when technically possible.[1] In our case, the first operation revealed a perforating tumor associated with localized peritonitis. Because we considered the patient to have a high risk of recurrence, we followed him carefully. At re-laparotomy, we tried to perform a complete resection of the residual tumor. However, only an ileocolostomy could be done because the tumor involved the superior mesenteric artery.

Table : Mesenteric Desmoid Tumors of Appendix in Japan

Patient No.	Study	Sex	Age (y)	History of surgery	Family history	Chief complaint	Size (cm)	Weight (g)	Recurrence	Prognosis
1	Otani	M	36	ND	ND	Lower abdominal pain	12	ND	ND	Alive
2	Handa	F	30	ND	ND	Abdominal mass	9.5×9.0×6.0	ND	ND	Alive
3	Ishizaki	F	17	—	—	Abdominal pain Abdominal mass	20.0×15.5×13.0	4,030	−	Alive
4	Pilichowska	M	43	—	—	Right iliac mass	ND	ND	−	Alive
5	Present case	M	24	—	—	Abdominal pain	20.0×19.0×13.0	1,700	+	Died

ND : no description

Complementary therapies have been used including radiation therapy, anti－estrogen therapy (tamoxifen) and chemotherapeutic agents.[13)14)] However, the effectiveness of these therapies on the tumor response still remains controversial.[14)] The role of radiation therapy for desmoid tumors is not clear.[1)] Some authors have judged radiation to be of limited value in the curative treatment of patients with desmoid tumors.[3)] However, radiation therapy should be considered as an adjuvant therapy to surgery when repeat surgery for a recurrent tumor is complicated by either a significant risk of morbidity.[3)15)16)] Anti-estrogen treatment is sometimes useful to reduce the patient's pain caused by rapidly growing desmoids.[17)] There is no well-established chemotherapy for desmoid tumors.[1)] The role of chemotherapy in the management of desmoid tumors is also not clear. We therefore, decided to use radiation therapy and tamoxifen. However, both therapies proved to be ineffective in the present aggressive case.

A complete local excision of the tumor remains the mainstay treatment for intra-abdominal desmoid tumors. Desmoid tumors of the mesentery and other peritoneal sites may recur and cause an intestinal obstruction and death when not resectable.[11)] The recurrence rates range from 16 to 33% and a 6% death rate has been reported for desmoid tumors in general.[11)] Mesenteric desmoid tumors should be considered as one of the causes of stenosis of the colon and small bowel, and patients should therefore receive a careful follow-up after unknown stenosis.[14)] The present patient therefore should have undergone a more careful and frequent follow-up after the first laparotomy.

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