

Case report

Leiomyosarcomas of the mesentery¹

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SUMMARY

We report on three cases of leiomyosarcoma of the mesentery, which were treated in our department during the period 1990-1999. They were two men and one woman aged from 50-82 years. All three patients presented with gradual weight loss and palpable abdominal mass. In addition, one patient developed a subileus symptomatology. In two cases the tumor was situated at the mesentery of the terminal ileum and in one case at the mesentery of the jejunum. The diameter of the tumors ranged between 5-12cm. No intraperitoneal spread of the tumor was found. All patients underwent curative resection of the tumor and the adjacent part of the small intestine (16-40 cm). All patients remain free of disease, 3months – 10 years after the operation.

Key words: leiomyosarcomas – GIST - mesentery

INTRODUCTION

The most common malignant tumors involving the mesentery are either metastatic tumors from other intraabdominal organs, or primary lymphomas arising from the mesenteric lymph nodes. Primary leiomyosarcomas of the mesentery are extremely uncommon. These tumors are today referred to as gastrointestinal stromal tumors (GIST). This term includes all the mesenchymal tumors of the GI tract, which tend to share common immunohistochemical characteristics. GISTs include most tumors previously designated as leiomyoma, cellular leiomyoma, leiomyoblastoma, and leiomyosarcoma. The mobility of the mesentery permits these tumors to grow

to a very large size before they present symptoms. The size of the tumor and the number of cell mitoses are the most significant prognostic factors for the long term survival of these patients.

CASE REPORTS

Case 1

A 50-year-old man presented a subileus symptomatic of three weeks duration. A hard, irregular and slightly tender mass was palpable in the lower right abdomen. The CT scan of the abdomen showed the presence of a mass of mixed density, which was adherent to jejunum loops and the cecum. The exploratory laparotomy revealed a 12x12cm mass in the mesentery of the ileum about 40cm from the ileocecal valve, compromising the lumen of the ileum. An extirpation of the tumor together with 16cm of the adjacent ileum was performed. The pathological examination showed that the tumor consisted mainly of spindle cells with high cellularity and hyperchromatic nuclei (Figure 1) and was positive for actin and desmin ; 3 cell mitoses per 50 optic fields were observed. The tumor infiltrated the intestinal wall, however the resection margins were free. The patient remains without recurrence of the disease 3 years after the operation.

Case 2

An 82-year-old man complained about repeated episodes of abdominal cramps, vomiting and weight loss. The ultrasonography of the abdomen showed a heterogeneous, hypoechoic mass anterior of the right kidney. The CT scan revealed a soft tissue mass located in the mesentery between the duodenum and the right kidney.

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During laparotomy a 5cm tumor was found in the mesentery of the jejunum about 30cm from the ileocecal valve. It was resected together with 40cm of the adjacent ileum. (Figure 2) The histopathologic evaluation of the specimen showed the presence of a leiomyosarcoma consisting mainly of spindle and round cells, as well as regions of necrosis and cystic degeneration; 5 mitoses per 50 optic fields were noted. No lymph node metastases were observed. The resection margins were free.

The patient remains without any signs of recurrence 9 years after the operation.

Case 3

A 63-year-old woman presented with a 6-month history of weight loss and recurrent episodes of meteorism and nausea. In the clinical examination a palpable, in-

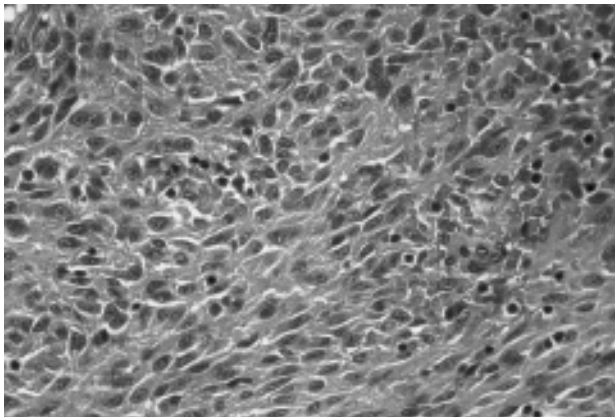


Figure 1. Microscopic view of case 1. The tumour consists of spindle cells with high cellularity and hyperchromatic nuclei

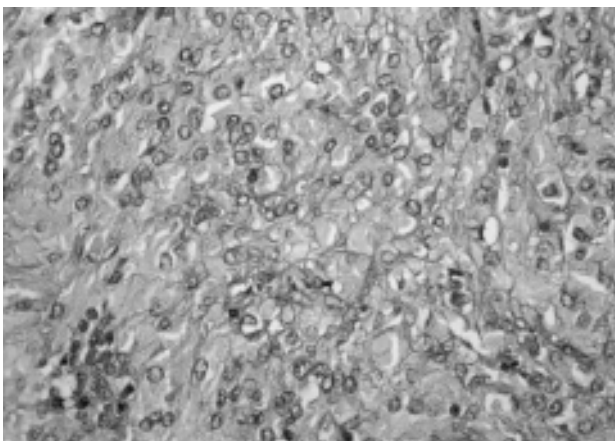


Figure 2. Microscopic view of case 2. Spindle and round cells, as well as regions of necrosis and cystic degeneration.

dent mass in the left abdomen was found. The ultrasonography showed a solid, heterogeneous mass, with no infiltration of adjacent organs. The exploratory laparotomy revealed a 10cm tumor of the mesentery of the jejunum, about 30cm from the Treitz ligament. The tumor lay in the periphery of the mesentery, and was adjacent to the jejunum. A sphenoid resection of the mesentery along with a 28cm resection of the jejunum was performed. (Figure 3) The histopathologic examination of the specimen showed a malignant stromal tumor of the mesentery with diffuse necrotic and hemorrhagic changes, and 6 mitoses/50 optic fields. The resection margins were free. The tumor was positive for actin (leiomyosarcoma).

The postoperative course was uneventful, and the patient remains asymptomatic, and without any evidence of recurrence one year after the operation.

DISCUSSION

The mesentery presents a common site of metastases of gastrointestinal malignancies. However, primary tumors originating between the leaves of the mesentery are quite uncommon, with the lymphoma being the most common. Other primary mesenteric malignancies include leiomyosarcomas, liposarcomas, fibrosarcomas, malignant teratomas or hemangiopericytomas.^{1,2}

Leiomyosarcomas present the most common mesenchymatous malignant tumor of the mesentery. These tumors are today referred to as gastrointestinal stromal tumors, and share common immunohistologic patterns including c-kit (CD117), CD34 or actin. The first three cases of leiomyosarcomas of the mesentery were described by



Figure 3. Intraoperative view of case 3.

Yannopoulos et al in 1962.³ Since then almost 30 cases have been reported in the literature.⁴⁻⁹ Since the mesentery contains no muscle fibers, the tumor is considered to arise from mesodermal elements of the mesenteric blood vessels, fibrous tissue or nerves. Approximately two thirds of these tumors are located in the mesentery of the small intestine, especially that of the ileum, but they can also arise from the transverse and sigmoid mesocolon or the gastrohepatic ligament.^{10,11} In two of our cases the tumor was located at the jejunal mesentery and in the third case in the mesentery of the ileum. The tumor is usually located peripherally in the mesentery, where it can often be adherent to the intestine.¹²

The clinical symptomatology of these tumors includes a palpable mass, abdominal distention and pain. Rarely, the tumor can present as an abscess, acute intraperitoneal hemorrhage or obstructive ileus.¹³ However, most of these tumors tend to be asymptomatic and can have grow to a large size by the time of diagnosis, since the mobility and elasticity of the mesentery allows the tumor to occupy a large intraperitoneal space without causing any obstructive symptoms. Harada reported the resection of a mesenteric leiomyosarcoma of 23cm in diameter and 2330g in weight.¹⁴ The diameter of the tumor in our cases varied from 5-12cm. A clinical characteristic of the tumor is that the mass can be easily moved from side to side, but not in the cephalocaudal direction.

The ultrasonography or CT of the abdomen can help the diagnosis. The presence of cystic or necrotic malformation within the tumor, however, can mislead the diagnosis to a mesenteric cyst, pancreatic pseudocyst or an uterine tumor.¹⁵⁻¹⁷ A barium enema or intravenous pyelography may detect a possible infiltration of the colon or the urinary tract by the tumor.

Since these malignancies are usually not responsive to either radiotherapy or chemotherapy, complete surgical resection remains the only hope for cure. When the tumor can be safely separated from the intestine, a local excision is possible. However, the resection of the adjacent intestine is usually necessary. In all our cases, we had to perform a resection of the adjacent part of the intestine, which varied in length from 16cm – 40 cm.

Ranchod showed that the mitotic activity of GISTs is the most useful indicator of malignant potential.¹⁸ The presence of more than 10 mitoses leads to a 10-year survival rate of only 14%.¹⁹ However, Hashimoto reported that ten of the 15 patients in whom the mitotic counts were less than five per 10 high power fields, developed

metastases and expired from the tumor.²⁰ It seemed, that in these cases the size of the tumor was the dominant prognostic factor.

The tumor tends to cause local recurrences or diffuse peritoneal or hepatic metastases.²¹ A careful post-operative follow-up for early detection of recurrence or metastases is essential. In our cases no recurrence or metastase occurred. This was probably due to the relatively small size of the tumors, the radicality of the resection and the low mitotic activity. The follow-up period of the third case is relatively short (12 months).

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