

## Case Report: Leiomyosarcoma of the First Portion of the Duodenum

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*A 51-year old male patient with a duodenal leiomyosarcoma located in the first segment and liver metastases is presented. Both were managed by radical resection and chemotherapy without pancreaticoduodenectomy. The patient required a second operation for recurrent liver metastases and survival for 9 months.*

**Key words:** Leiomyosarcoma, duodenum

### Vaka takdimi: duodenum birinci kısım leiomyosarkomu

*Duodenum birinci kısımda lokalize leiomyosarkom ve karaciğer metastazı olan 51 yaşındaki erkek hasta sunuldu. Bu iki patoloji pankreatikoduonektomi yapılmaksızın radikal rezeksiyon ve kemoterapi ile tedavi edildi. Hastanın tekrarlayan karaciğer metastazı nedeni ile ikinci bir ameliyata ihtiyacı oldu ve 9 ay yaşadı.*

**Anahtar kelimeler:** Leyomyosarkom, duodenum

Primary duodenal carcinoma is rare and is generally considered to have a low resectability rate as well as a dismal prognosis and fewer than 800 cases have been reported (1). Duodenal leiomyosarcomas are also rare tumors and account for about 10 % of duodenal malignancies (2). Approximately 100 tumors have been reported in the literature. Most duodenal leiomyosarcomas are located in the second portion. Leiomyosarcomas of the first portion are very rare. Liver metastasis and peritoneal spread are frequently seen.

We present a patient with a leiomyosarcoma measuring 10x12 cm, located in the first portion

of the duodenum and a 3x5 cm liver metastasis by radical resection.

### CASE REPORT

A 51-year old male patient complaining of epigastric pain and distention was admitted to the gastroenterology department. He had a history of peptic ulcer perforation which was treated by primary closure 4 years ago.

The physical examination revealed a large palpable mass in the epigastrium. The stomach was normal with gastroduodenoscopy while there

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was a diverticular formation in the bulbus completely lined with necrotic material. Biopsies taken from this structure showed richly cellular, disorganised smooth muscle fibres. No mitosis were detected and the diagnosis was smooth muscle tumor of undetermined malignant potential

Computerised tomography revealed a 30 mm isohypodense mass in the left medial segment of the liver. There was a 81x86 mm mass medial but separate from the liver which opacified and showed a thick wall with oral contrast.

The patient underwent laparotomy through a right paramedian incision. A nodular vascular mass measuring 10x12 cm originating from the



**Figure 1.** Low power view showing submucosal Brunner's glands and the pushing border of the tumor (x 40, H.E.).



**Figure 2.** High power view of the tumor showing fascicles of oval cells with mitotic figures and slight atypia (x 400, H.E.).

first portion of the duodenum and growing subserosally was seen. There was a metastatic mass on the anterior surface of the liver medial to the fundus of the gallbladder. The liver metastasis was resected along with margin of healthy liver tissue. The duodenal mass was dissected free of the gallbladder, transverse colon and pancreas. The distal margin of the duodenal resection was made 3 cm below the tumor, preserving the second portion of the duodenum. The tumor was resected as a 50 % distal gastrectomy along with the first portion of the duodenum. The passage was reconstructed as a Billroth II.

Pathologic examination revealed a solid-white tumor involving the duodenum with serosal invasion 2,5 cm from the distal surgical margin. Microscopic examination showed muscle bundles with prominent mitotic activity with five or more mitoses per ten high-power fields (Figure 1,2). There was no tumor invasion in the stomach. Eleven regional lymph nodes were evaluated as reactive. The tumor excised from the liver measured 3x5 cm and showed a tumor of similar morphology demonstrating expansive development.

The postoperative period was complicated by intraabdominal fluid collection treated with percutaneous drainage. Discharge of bilious, purulent material decreased progressively with continuing drainage and antibiotic therapy. Doxorubicin chemotherapy was initiated after oncology consultations.

Our patient required a second operation for removal of another large liver metastasis at the second postoperative month. He died 9 months postoperatively from massive gastrointestinal bleeding and wide spread liver metastasis.

## DISCUSSION

Duodenal leiomyosarcomas are mostly seen between ages 40-60 and are two times more frequent in males (2,3). Pain, palpable mass and recurrent melena are frequent complaints. Weight loss, nausea, dyspepsia and intestinal obstruction can also be frequently seen. Twenty percent of small intestinal leiomyosarcomas are

found in the duodenum (5), where as only 8 % of duodenal leiomyosarcomas are localised in the first portion (6).

The selected therapy in leiomyosarcomas is surgical exicion. Most of the leiomyosarcomas in the duodenum (50 %) are located in the second portion and require a Whipple's operation. Tumours located in the first, third and fourth portions are treated with radical resection (3,6,7,9). Liver metastasis and peritoneal spread are commonly seen. Spread to other organs, a tumor larger than 5 cm and high mitotic activity are associated with a poor prognosis (5). The 5 year survival in leiomyosarcomas is below 50 % (4,5,7,8). The average survival is reported between 10-38 months (3). Our case survived for 9 months.

In our case, the tumor was located in the first portion of the duodenum with extraluminal growth and a 3x5 cm liver metastasis. The tumor was resected with clean surgical margins without pancreaticoduodenectomy and the reconstruction was performed as a Billroth II type procedure. The liver metastasis was also resected along with margin of healthy liver.

We have not been able to find a leiomyosarcoma of this size located in the first portion of the duodenum with a liver metastasis in the literature. A segmental radical resection is advocated for leiomyosarcomas located in the first portion. Despite its large size, the tumor in our case was resected along with the distal part of stomach preserving the second portion of the duodenum and avoiding a Whipple procedure. While liver metastasis frequent, a single large 3x5 cm metastasis resected simultaneously

during the same operatin is also a significant finding unreported in the literature.

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