

# Primary Large Exophytic leiomyosarcoma of the stomach: A case report with review of literature

## Abstract

Gastric leiomyosarcoma accounts for 1% of all gastric tumors. This type of tumor typically arises from the muscularis propria and is most often found in the body, fundus, cardia, and pyloric antrum of the stomach. Gastric leiomyosarcoma commonly presents as a polypoidal mass, exophytic mass, or ulcerative mass. The primary symptoms include bleeding leading to hematemesis, anaemia, abdominal pain, weight loss, and the presence of an abdominal mass.

Effective diagnostic modalities include endoscopic ultrasonography, contrast-enhanced CT scan, and MRI. Immunohistochemically, the tumoral cells in gastric leiomyosarcoma show positive immunoreactivity to smooth muscle actin and desmin while testing negative for CD117 (C-kit) and CD34. The most effective treatment is complete surgical resection with negative margins, typically achieved through either open or laparoscopic partial gastrectomy.

We report the case of a 57-year-old male who presented with a large mass in the epigastric and left hypochondriac region. Diagnosis was confirmed via CT scan of the abdomen, and histopathological examination verified a high-grade gastric leiomyosarcoma.

## Key words

Leiomyosarcoma, stomach tumor, gastric tumors, GISTs.

## Introduction

Gastric leiomyosarcoma was first described by Margagni in 1762. Leiomyosarcoma of the stomach arises from the smooth muscle cells of the muscularis propria. These tumors are most frequently seen in adults with median age of 50 to 70 and with more men than women being affected 2:1 ratio. [1,2,3]

The etiology of gastric leiomyosarcoma has been associated with high risk factors such as ionising radiation, Epstein Barr virus, various chemical exposures, trauma to the affected region and immunocompromised patients. There is also a potential role of helicobacter pylori in its development. [2,38]

### **Leiomyosarcoma of stomach may present the following signs and symptoms.**

- Small size tumors may be asymptomatic
- Most tumors masses appear as polypoidal mass intraluminal or extraluminal on the stomach wall.
- Commonly observed symptoms are abdominal pain, discomfort, weight loss and fatigue.
- Large tumors may develop palpable abdominal lumps.
- Large tumors may ulcerative and bleeds intraluminally causing hematemesis, melena and anaemia. [1,2,3]

### **Diagnosis of leiomyosarcoma of stomach.**

- Complete physical examination with evaluation of medical history
  - Ultrasonography to detect the stomach mass.
  - Abdominal CT scan –provides more details of soft tissues, blood vessels and internal organs.
- Typically, large mass is usually exophytic, endogastric or dumbbell shaped masses can be detected.

The most helpful CT feature in gastric leiomyosarcoma is the presence of a large, heterogenous mass with areas of low attenuation, representing necrosis. Calcification is rare in these tumors.

- Upper GI Endoscopy -detect the intraluminal polypoidal mass in the stomach.
- Endoscopic ultrasonography – during this procedure, fine needle aspiration biopsy. This a good technique for tumor detection including tumor invasion parameters and whether nearby lymph nodes are affected. [1,2,5]
- Whole body PET scan- detect cancer spread to other organ systems.
- Tissue biopsy, such as fine needle aspiration(FNAC), core biopsy, open biopsy to assess laboratory or pathological examination.[2,6,8]

### Case Report

A 57-year-old male was admitted to our center on January 15, 2020, presenting with complaints of an abdominal lump and pain in abdomen for the past six months. Physical examination revealed normal vital signs. There was no history of hematemesis, jaundice, or vomiting. Abdominal examination identified a large palpable mass in the epigastrium and left hypochondrium, with mild tenderness. Blood tests, including liver and renal function tests, were within normal limits.

An abdominal CT scan revealed a solid, exophytic, nodular mass measuring 25 x 15 x 10 cm. The mass was ill-defined, heterogeneous, and lobulated, with its main bulk in the left hypochondrium, originating from the gastric body. It exerted pressure on the stomach, spleen, and pancreas and diagnosis was gastric mass.

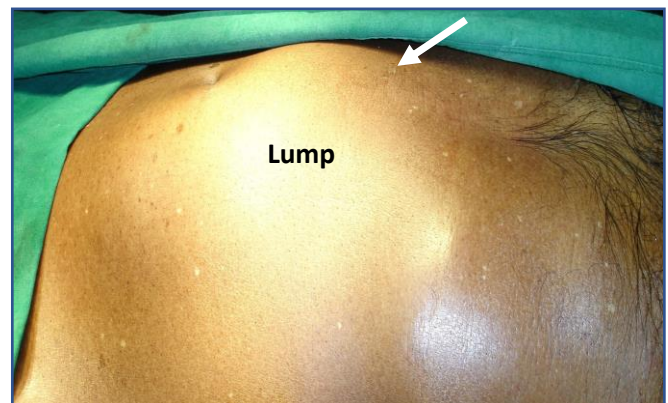
We performed an exploratory laparotomy via a midline abdominal incision with the aim of complete tumor removal, which is the standard treatment for gastric leiomyosarcoma. The patient underwent a partial gastrectomy, removing the large 25 x 15 x 10 cm multinodular mass with a 5 cm margin of normal gastric tissue along with the greater omentum.

Gross examination revealed a multinodular mass weighing 3.5 kg. Microscopic examination showed spindle-shaped tumor cells with hyperchromatic pleomorphic nuclei and a high mitotic index. Immunohistochemistry was positive for smooth muscle actin and desmin, but negative for CD117.

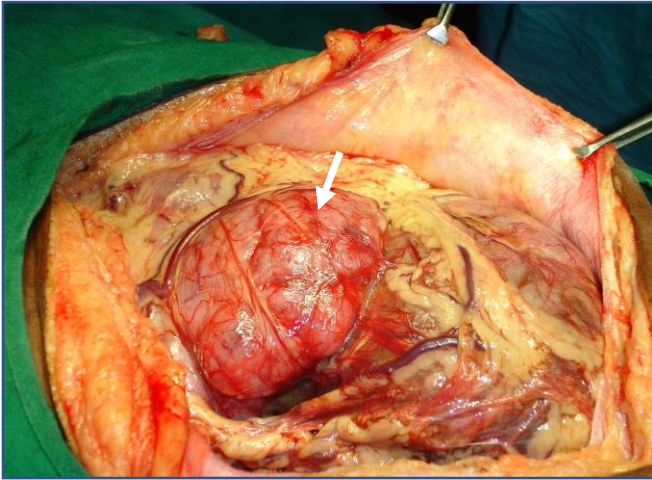
The patient was discharged on the tenth post-operative day and was advised to pursue chemotherapy at a cancer hospital due to the high-grade nature of the gastric leiomyosarcoma. Unfortunately, the patient passed away one year after surgery at home. (Fig 1-10)



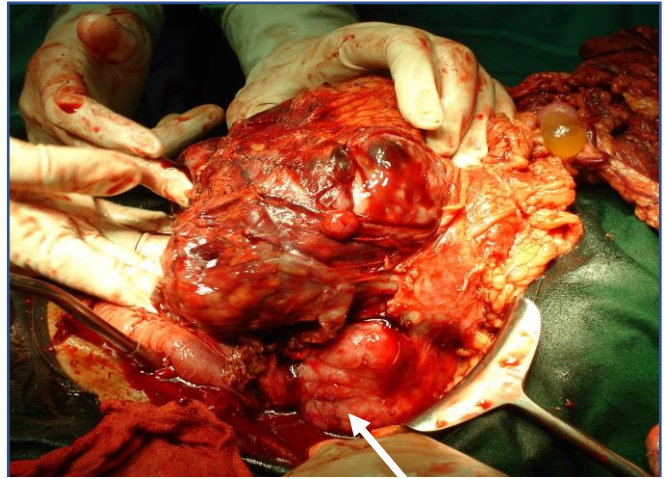
**Fig-1** CT abdomen showing a large heterogeneously enhancing exophytic mass arising from the body of the stomach



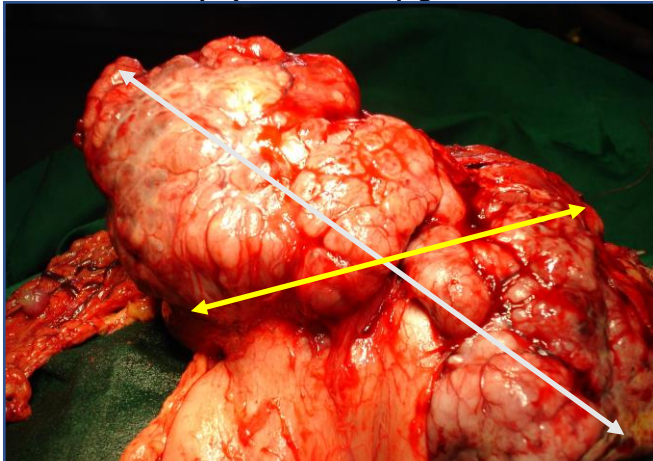
**Fig-2** Photograph showing a huge lump at epigastrium and Left hypochondrium



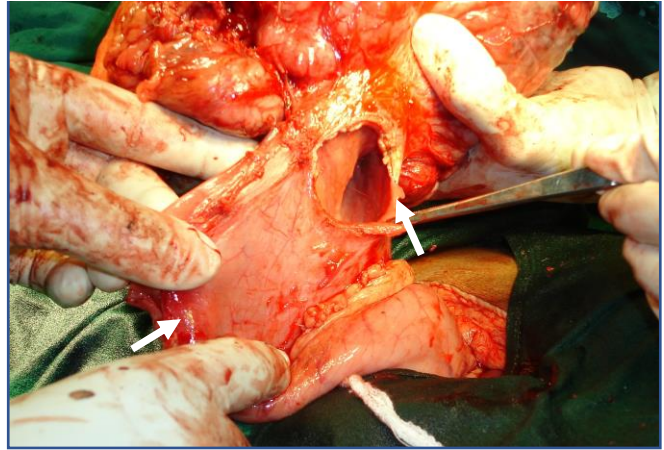
**Fig-3 Intraoperative photograph showing an exophytic mass at epigastrium**



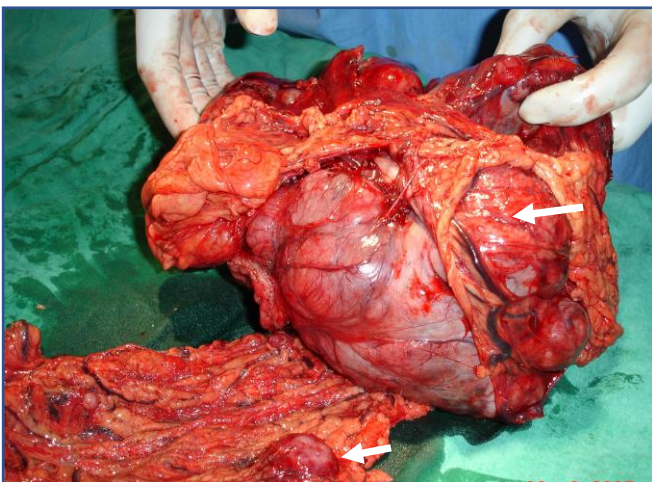
**Fig-4 Intraoperative photograph showing a huge, multinodular mass**



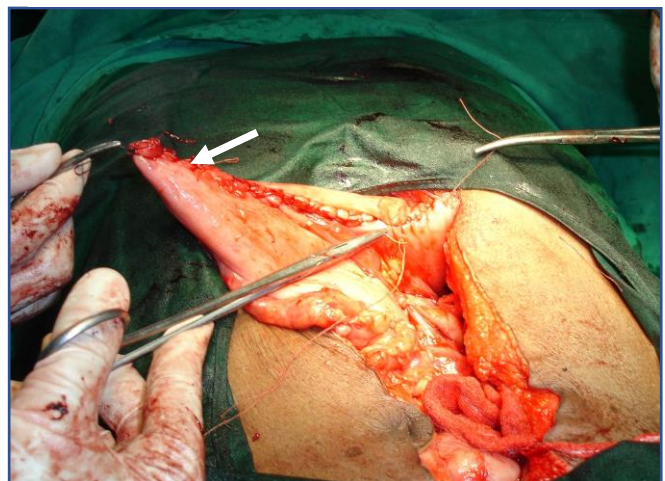
**Fig-5 Intraoperative photograph showing a huge multinodular mass arising from body of the stomach**



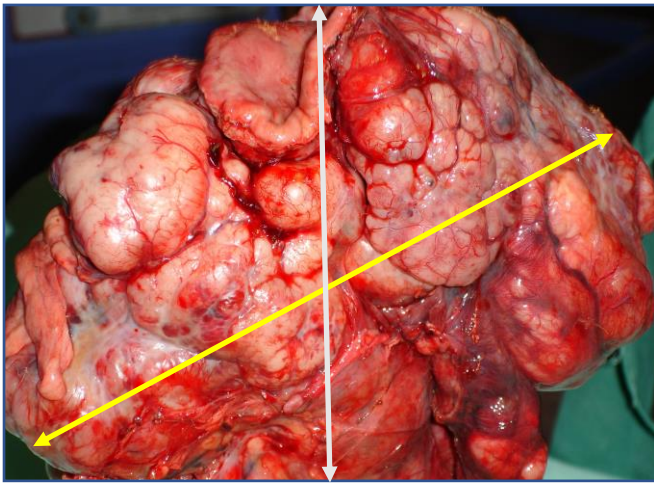
**Fig-6 Intraoperative photograph showing a wide excision of gastric mass**



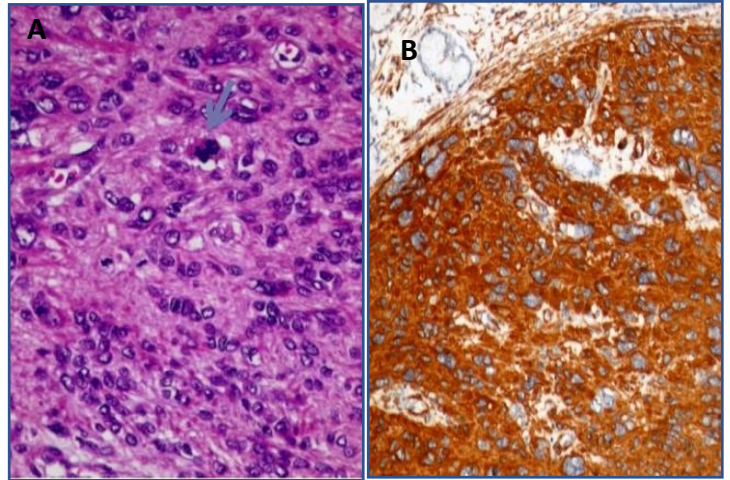
**Fig-7 Intraoperative photograph showing a large, polypoidal mass with omental excision**



**Fig-8 Intraoperative photograph showing a partial gastrectomy done**



**Fig-9 Photograph showing a large multinodular mass measuring 25x15x10 cm weighing 3.5 kg**



**Fig-10 A. Photograph showing smooth muscle cells with nuclear hyperchromasia and numerous mitoses. B. Well differentiated leiomyosarcoma, neoplastic cells positive (brown) for desmin**

### **Discussion and review of literature**

Gastric leiomyosarcoma typically originates from the muscularis propria or muscularis mucosa layer in various parts of the stomach, including the body, fundus, antrum, cardia, and pylorus. Common clinical signs and symptoms include abdominal pain, melena, weight loss, and the presence of an abdominal mass. Tumor sizes can range from 1 to 18 cm. [1,2]

Effective diagnostic tools include endoscopic ultrasonography, contrast-enhanced CT scans, and MRI. These imaging modalities help in detecting and assessing gastric leiomyosarcomas. For definitive diagnosis, histopathological examination is essential. Simple endoscopic biopsies might be inadequate; therefore, endoscopic ultrasound-guided fine needle aspiration cytology is recommended for accurate diagnosis, with a success rate of up to 97%. [3,4,7]

Since the early 2000s, only 19 cases of gastric leiomyosarcoma have been reported, with our case being the 20th. Approximately 90% of these tumors arise from the body or fundus of the stomach, with 10% originating from the gastric antrum. [2,7,10]. Immunotherapy with agents such as nivolumab and pembrolizumab is also being explored. Ongoing phase I and II trials are investigating new drug combinations with radiation and chemotherapy. [2,6]

Surveillance imaging with CT or MRI of the chest, abdomen, and pelvis is recommended every six months for the first two years and then annually thereafter. The main prognostic factors include histopathological grading, tumor size (>5-7 cm), evidence of synchronous metastasis, and gastric wall infiltration. The 5-year survival rate for patients with gastric leiomyosarcoma is approximately 22%. [1,2,4].

Leiomyosarcoma of the stomach is extremely rare and accounts for 1% of all gastric tumors. Most of the cases reported in the “Pre-kit era” as leiomyosarcomas of the stomach were actually gastrointestinal stromal tumors (GISTs’) of the stomach. Since the established of kit

immunohistochemistry in the Late 1990s, we have realized that primary gastric leiomyosarcoma is extremely rare. [1,2,4]

The diagnosis of gastric leiomyosarcomas relies as pathological examination, in general SMA, desmin and h-caldesmin are positive in the majority (70%) of leiomyosarcoma cases and CD117. CD 34 and DOG1 are negative. DOG1 is the best marker for GIST tumors. CD 117 and DOG1 are most sensitive and specific marker that can cover 99 % of GISTs. CD34 and 5-100 also positive in GISTs. GISTs originate from the intestinal cell of Cajal and leiomyosarcoma originates from smooth muscles spindle cells of stomach wall. [2,3,5]

Leiomyosarcoma treatment depends on the location and size of the tumor.

1. Surgery – it is the best option for leiomyosarcoma. The goal is to remove the entire tumor, so that the cancer does not come back.
2. Chemotherapy – This treatment is often recommended when the tumor is large or when cancer spread to other parts of the body.
3. Radiation therapy – This treatment may be used before surgery to shrink the tumor or after surgery to kill any remain cancer cells. [1,2,6]

Currently, the preferred treatment method in surgical is wedge resection with negative margin of 2.5 to 5 cm, or partial or total gastrectomy. Pauser et al, also reported submucosal endoscopic resection of leiomyosarcoma of the stomach. Laparoscopic gastrectomy and endoscopic submucosal dissection have been used for the removal of gastric leiomyosarcoma, called Laparoscopic and endoscopic co-operative surgery (LECS). The margin of the tumor was marked endoscopically and laparoscopic partial gastrectomy done. [3,6,8]

Gastric leiomyosarcoma is a cruel disease that does not respond to chemotherapy or radiation multimodality therapy involving radiotherapy and chemotherapy is benign inverted and currently recommended for cases with high local recurrence, high grade and metastasis disease. Doxorubicin and docetaxel chemotherapy drugs are used for the first line treatment. Surveillance imaging involving CT or MRI of the chest, abdomen and pelvis is recommended every 3 to 6 months for 2 to 3 years. Every 6 months for the next 2 years and then annually. The 5-year cancer survival rate is 22 to 60%. [2,3]

The prognosis depends upon stage of tumor, health of individual and old age, individual with bulky disease may have poorer prognosis. The 5 years survival rate for patient with leiomyosarcoma is 22% and more careful clinical follow up is advised. [1,2]

## **Conclusion**

Gastric leiomyosarcoma are extremely rare and diagnosis is based on histological examination with immunohistochemistry for marking and accurate diagnosis. The standard treatment for gastric leiomyosarcoma is complete resection of the tumor, a wedge resection, partial or total gastrectomy with En-bloc resection if adjacent organs are involved.

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