

Gastric Leiomyosarcoma: First Case Report from the Kingdom of Bahrain and Review of Literature

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Gastric leiomyosarcoma are very rare tumors. We are reporting the first case of gastric leiomyosarcoma diagnosed in Bahrain and documented in the cancer registry. Unfortunately, the patient died due to rapidly progressing nature of the disease despite the multimodality treatment.

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Gastric sarcoma is a very rare tumor accounting for 1-3% of all gastric malignancy worldwide¹⁻⁴. Few case reports have been published in the literature. Major institutes report less than 3 cases per year³. The management of this tumor is a multidisciplinary approach consisting of surgery, chemo and radiation therapy. Information regarding the treatment of metastatic tumor is limited⁵. The prognosis depends upon the extent of disease at the time of presentation.

Search of cancer registry in Bahrain revealed that this is the first reported case of gastric leiomyosarcoma in the Bahrain.

The aim of this report is to highlight the first case of leiomyosarcoma in Bahrain and to alert the physicians of its unique presentation, management and aggressive nature.

THE CASE

A thirty-nine years old Bahraini male with no significant medical history. The patient attended to a private hospital with history of chronic back pain of 3 to 4 years duration. Recently he developed upper abdominal pain associated with loss of appetite and significant weight loss of about 10 Kg over the past five months. The patient had no history of fever, night sweats, cough and change of bowel habits or pruritus.

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Clinical examination revealed a firm abdominal mass in the left upper quadrant. MRI of dorsal spine showed diffuse disc bulge (Dorsal spine 8) encroaching on the neural foramina and a large intra abdominal mass. CT Scan of the abdomen and pelvis revealed a well defined cystic mass in the left side of upper quadrant measuring 13 x 16 x 22 cm; it was closely related to anterior aspect of the pancreas and almost inseparable from it. The mass was extending to anterior aspect of the spleen and causing compression of stomach. The mass extended from the level of left diaphragm to the level of renal vessels.

Exploratory laparotomy and total removal of the mass was performed on first July 2008. Operative findings showed huge solid mass around 20 x 20 cm occupying the left upper quadrant and attached to the spleen, stomach, left lobe of liver and diaphragm. The resection included the greater curvature of stomach, left lateral lobe of liver, spleen and a portion of diaphragm. One cm nodule on the distal jejunum was also excised.

Histopathology revealed a high grade spindle cell sarcoma arising from the gastric wall with invasion of the spleen, liver and diaphragm. The tumor cells around the blood vessels showed epithelioid appearance with many osteoclast like tumor giant cells. All surgical margins were free of tumor. Excised jejunal nodule showed features of benign gastrointestinal stromal tumor.

Postoperative follow up on 3rd August 2008 did not reveal any evidence of the disease. He was admitted on 23rd August 2008 to the Salmaniya Medical Complex with history of abdominal pain and vomiting. Examination showed upper abdominal mass. No signs of acute abdomen were seen.

Repeat postoperative CT scan on 28th August 2008 revealed at least 4 lesions within the remnant of the stomach and pancreas, the largest lesion measured 10.4 cm x 7.2 cm, see Figure 1. Upper mesentery showed fat stranding with few upper mesenteric sub-centimeter lymph nodes. The size and number of the lesion had increased compared to the CT scan done on 6th August 2008. Bone scan on 5th August 2008 was negative for bone metastasis.

Histopathological slide review at Salmaniya medical complex showed tumor arising from the wall of the stomach with infiltration into the spleen, see Figure 2. Histopathology displayed all the features of a high grade sarcoma possibly leiomyosarcoma, see Figure 3, 4. The histopathological diagnosis was supported by the immunoreactivity for Vimentin and smooth muscle actin and non immunoreactive for CD34, CD117, Desmin, S100, Melan A and cytokeratin (MNF116).

The tumor in the small intestine was confirmed as gastrointestinal stromal tumor as shown by the immunoreactivity for Vimentin, CD34, CD117 and focally for Desmin and S100. Diagnostic ascitic tapping was done and it was negative for malignant cells, see Figure 5. The patient traveled to Singapore on 28th September 2008 for further management in a sarcoma center because this facility is currently not available in Bahrain. His diagnosis was reconfirmed as high grade gastric leiomyosarcoma.

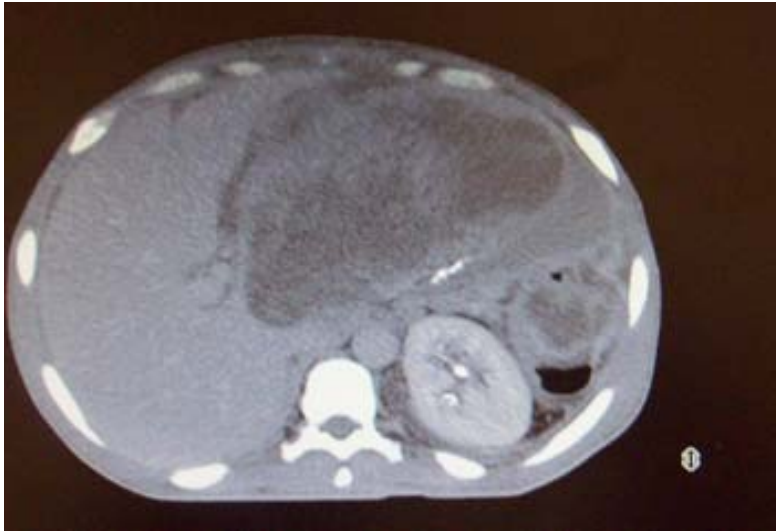


Figure 1: CT Scan Showing Massive Abdominal Mass

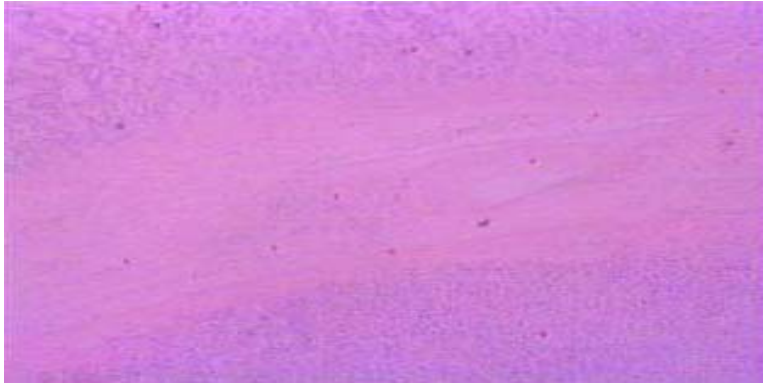


Figure 2: Photomicrograph Showing the Tumor Arising from the Muscularis Propria of the Stomach Wall. 40x Magnification H & E Stain- scanner View

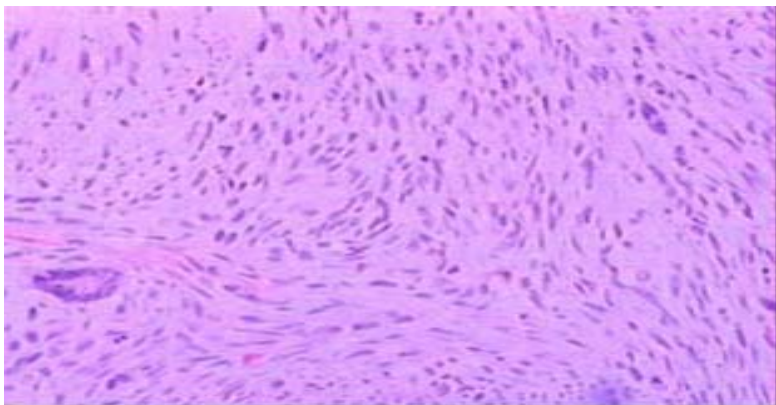


Figure 3: Photomicrograph Showing the Spindle Cells Arranged in Interlacing Fascicles with Multinucleated Giant Cell. 100x Magnification H & E Stain

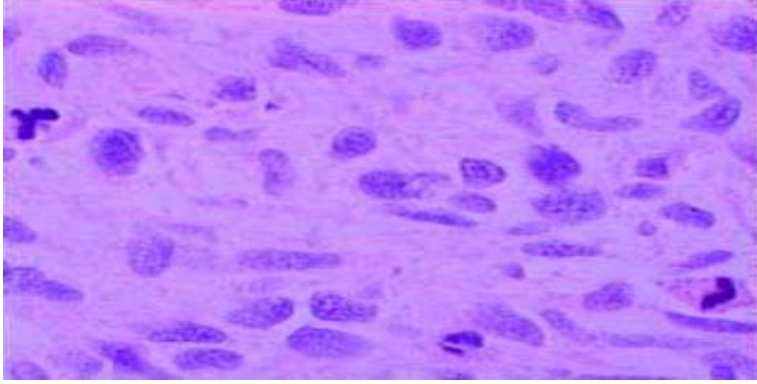


Figure 4: Photomicrograph Showing the Spindle Cells of the Leiomyosarcoma with Multiple Abnormal Mitosis. 400x Magnification H & E Stain



Figure 5: Clinical Photograph Showing Massive Abdominal Distension

PET/CT scan done on 7th October 2008 showed multiple huge peritoneal masses and hyper metabolic lesion in the dome of the liver consistent with hepatic metastasis. There was no metastasis noted in the lungs or the bones. It was decided to downsize the tumor by administering combination chemotherapy consisting of Gemcitabine and Docetaxel followed by surgery. The ascitic fluid was drained by an indwelling pigtail catheter. The pain was managed with transdermal Fentanyl and morphine. Two cycles of chemotherapy were given; his condition deteriorated quickly and had progressive difficulty in breathing. The patient died in Singapore on 16th October 2008, most likely due to pulmonary embolism. His disease had progressed in spite of the chemotherapy.

DISCUSSION

Approximately 1-2% of solid tumors are soft tissue sarcoma and leiomyosarcoma comprise 2-9% of these⁶. Twenty percent of leiomyosarcomas are found in the gastrointestinal tract, the sites are evenly divided between the stomach and the small intestine⁴. Gastric leiomyosarcomas are

rare tumor accounting for 1-3% of all gastric malignancy¹⁻⁴. Leiomyosarcomas arise from the smooth muscle of the gastric wall, mostly located in the submucosa³. They involve the pyloric end but unlike carcinoma do not usually cause a stenosis because their growth occurs extraluminally. As a rule, leiomyosarcomas do not produce the severe symptoms of carcinoma but owing to their bulk usually give a demonstrable mass earlier⁶. They often present with massive bleeding (70%)⁶. Complaints of malaise and fatigue due to anemia are often present in patients who bleed chronically⁶. Weight loss is a late feature; its incidence is 20%⁶.

Six percent of patients with leiomyosarcomas had a history of Crohn's disease⁷. The tumor size ranges from 4 cm to 20 cm^{8,9}. The tumor is generally made up of spindle shaped cells and has a high cellularity. The histological diagnosis of a leiomyosarcoma is based on immunohistochemical expression of SMA³. The definitive diagnosis of the tumor is difficult before surgery⁹.

Metastasis is primarily hematogenic. Lymph node metastasis is rare occurring in 0-15% of cases. Primarily leiomyosarcomas spread to the liver and peritoneum; the lung is less frequently affected than the liver and peritoneum⁶. This is in contrast to other soft tissue sarcomas in which the lung is the most common site of metastasis. About 20-40% of patients have metastasis at the initial laprotomy. Distant soft tissue metastases in gastric leiomyosarcoma are extremely rare¹⁰.

Complete tumor resection is the standard treatment but most surgeons remove the tumor together with a safety margin, i.e. resecting the gastric wall or partial gastrectomy^{2,10}. Crocker advises a safety margin of at least 2 cm¹¹. The surgeon must avoid rupturing the tumor to prevent peritoneal seeding. Regional lymph node dissection is not indicated because nodal metastasis is infrequent and if it occurs, it is usually accompanied by metastatic disease, which prohibits curative treatment^{3,12}.

Adjuvant combined modality treatment has been suggested for patients with local tumor invasion resected for cure⁵. Adjuvant radiotherapy has been used without effect on salvage rate^{5,13}. Treatment of advanced disease by chemotherapy is not substantiated⁵. Guidelines for adjuvant treatments had not been published⁶. Most of the available information is retrospective and it refers to small number of patients. The presence of distant metastasis and or direct invasion of adjacent structures, tumor size of more than 6 cm and high grade have been identified as unfavorable predictors of survival⁵. The mucosal ulceration indicates rapid tumor growth⁵. The three most common factors identified in disease free survival after curative resection are local extent of the tumor, tumor size and histopathological grade of the tumor^{14,15}. The median and mean survival of gastric leiomyosarcoma after curative resection ranges from 10.1-80.1 months¹⁰. Distant metastasis decreases the survival rate to 30%⁷.

CONCLUSION

In Bahrain, stomach malignancy accounts for less than one percent of all malignancy. This is the first case of gastric leiomyosarcoma in the Kingdom of Bahrain. This patient had the radical surgery in a private hospital and referred to Salmaniya medical complex for adjuvant treatment. The tumor had progressed rapidly before initiating the adjuvant treatment. This patient was sent to a specialized sarcoma center in Singapore. Unfortunately, the patient did not respond to the systemic chemotherapy and died due to the progressive and metastatic disease.

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