

# A Case of Duodenal Leiomyosarcoma

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## Abstract

Duodenal leiomyosarcoma is a rare disease with poor prognoses for patients. The disease does not have a clear set of signs and symptoms that allows easy diagnosis. Diagnosis is made on the basis of histopathological identification of a mesenchymal lesion composed of malignant tumor cells and immunohistochemistry positive for smooth muscle actin, desmin, muscle specific actin, calponin and caldesmon. We report a case of a 56 year old woman with primary duodenal leiomyosarcoma which had metastasized to the pancreas and the celiac artery.

## Keywords

Leiomyosarcoma, duodenum, small intestine, mesenchymal tumors.

## INTRODUCTION

Gastrointestinal mesenchymal tumors are a group of neoplasms which have similar histologic features but have different patterns of immunohistochemistry (1, 2). Gastrointestinal stromal tumor (GIST) tumors are the most common mesenchymal neoplasms that occur in the gastrointestinal tract. They are followed by smooth muscle and neural tumors (1).

Gastrointestinal smooth muscle tumors are rare. Arising in the muscle of the mucosa, they are most commonly located in the esophagus and then in the colon and rectum (3). These tumors are divided into leiomyomas which are benign and leiomyosarcoma tumors which are malignant tumors and rarer (3). Gastrointestinal leiomyosarcomas occur most commonly in the stomach. This type of tumor in this location accounts for 0.1% to 3% of all gastric tumors (4). The next most common location is the small intestine (5). Within the small intestine the most common locations

are the jejunum and ileum with less frequent occurrence in the duodenum (6).

We report a case of a 56 year old woman who was diagnosed with primary leiomyosarcoma of the duodenum, and include a literature review of cases reported with adequate histopathological diagnoses.

## CASE REPORT

The patient was a 56 year old woman with a history of non-insulin dependent diabetes mellitus, hypertension and hypercholesterolemia. She came to the hospital after 20 days of epigastric pain associated with watery stools, emesis, fatigue and weight loss.

An abdominal CAT scan showed concentric thickening of the wall of the third portion of the duodenum wall together with a lesion with irregular margins measuring approximately 94mm x 72mm x 48mm that extended into

the periduodenal tissue. In addition, multiple round and oval hypodense lesions were found in the topography of the celiac trunk (Figure 1). Complementary esophagogastroduodenoscopy (EGD) and endoscopic ultrasound (EUS) showed stenosis in the third portion of the duodenum.

Histopathology of an excisional biopsy of the duodenal mass revealed a malignant mesenchymal tumor composed of epithelioid and fusiform areas (Figure 2). The tumor had markedly atypical with cytopathology with a mitotic count of 27 in 10 high power fields and necrosis compromising 20% of its area. An additional pancreatic biopsy showed that it was also compromised by a tumor (Figure 3).

Immunohistochemistry of spindle cell areas was predominantly positive for smooth muscle actin, desmin, calponin and keratins. In addition, epithelial and spindle cell areas were also positive for CD 99 and CD138 (Figure 4), and positive for diffuse BCL2, with a proliferation index of 60% for Ki67. In addition, tests were negative for HMB45, S100, CD117, DOG1, CD34, EMA, INI1, and TL1.

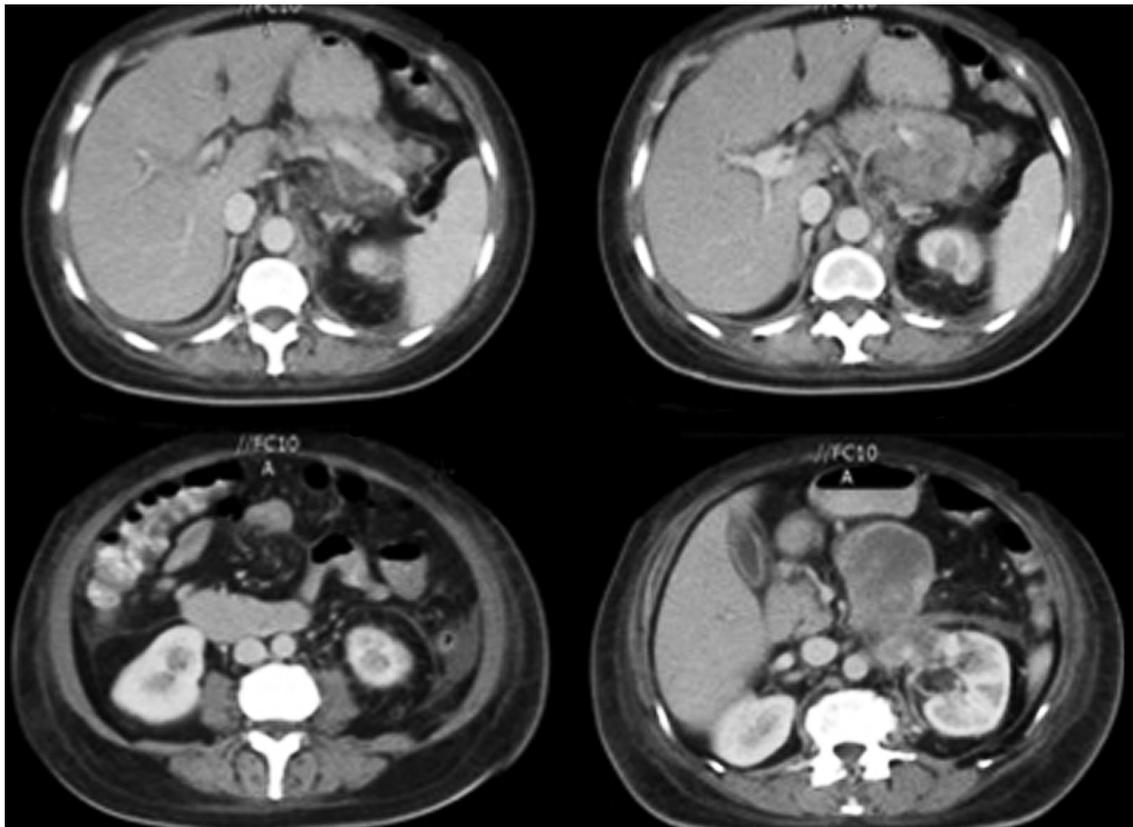
The patient was treated for her comorbidities and remained hospitalized for twenty days at which time she passed away.

## DISCUSSION

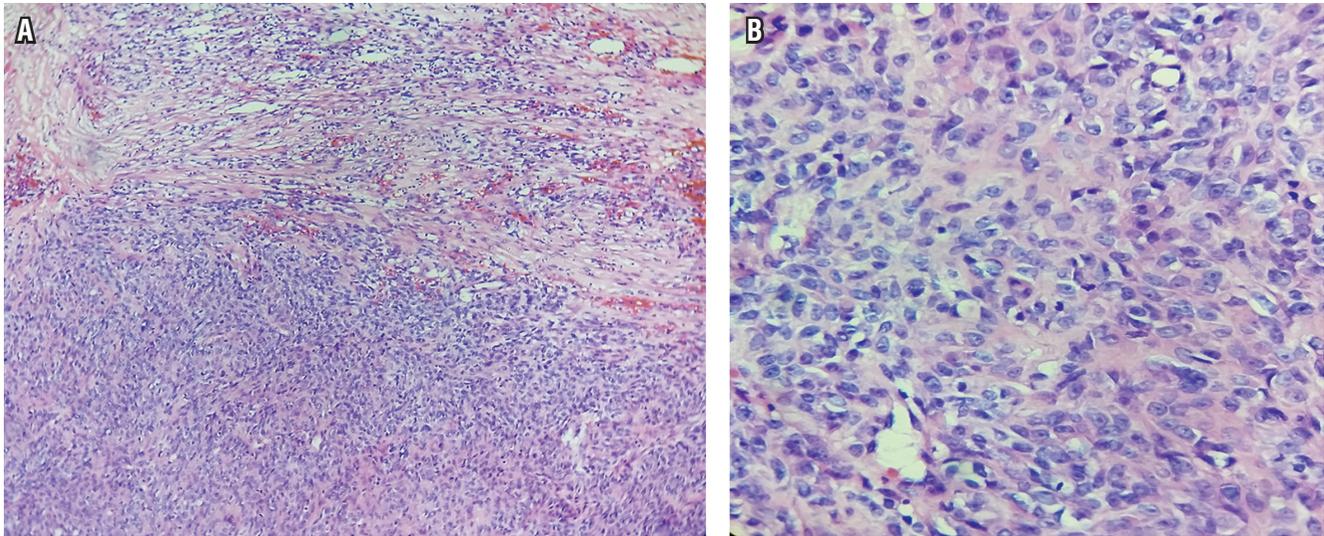
Gastrointestinal mesenchymal tumors are a group of neoplasms that arise from the stromal elements of the wall of the gastrointestinal tract. While they are histologically similar, the immunohistochemical profiles of the different types of gastrointestinal mesenchymal tumors vary (1). These tumors include GISTs, smooth muscle tumors, glomus tumors, schwannomas, inflammatory myofibroblastic tumors, plexiform fibromyxomas, synovial sarcomas, and Kaposi's sarcomas (7). These tumors are characterized by specific anatomical regions although they may not be exclusive to those regions (8).

The first reported case of primary leiomyosarcoma of the duodenum was published in 1920 by Von Salis (9). About 170 cases have been reported in the medical literature (6, 10-13). Cases of duodenal leiomyosarcomas that have metastasized to the corpus uteri have also been reported (14, 15).

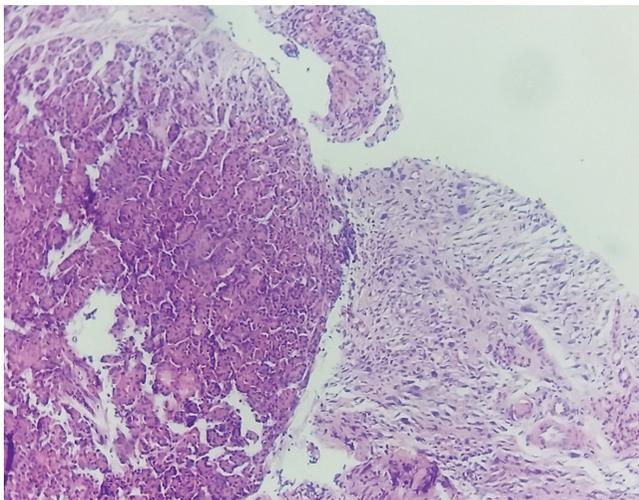
Duodenal leiomyosarcomas most frequently occur in the sixth decade of life and are slightly more common among men than among women. The most common location is in the second portion of the duodenum (16). Like other sar-



**Figure 1.** Lobed mass with mixed density which captures the contrast medium in a heterogeneous way. It has displaced vascular structures and caused severe narrowing through extrinsic compression of the splenic vein. Peritoneal lymphadenopathy can be seen with alterations of fat around the mass. The mass is about 94 x 72 x 48 mm. There is free liquid in the anterior pararenal space.



**Figure 2.** Photographs of duodenal wall. **A.** Duodenal wall infiltrated by malignant tumor formed by spindle cells with epithelioid areas. **B.** Markedly atypical cells with frequent mitosis seen at higher power.



**Figure 3.** Pancreatic parenchyma compromised by malignant tumor

comas, these tumors can disseminate through the blood, especially to the lungs and liver. Lymphatic dissemination can also occur and has been described primarily to the pancreas, mesocolon and transverse colon (17). Metastases to the skin and skeletal muscles have also been reported (6, 18). Patients with this disease have poor prognoses: the five year survival rate is 5% while the ten year survival rate is 0.0% (19, 20). One case of reoccurrence six years after treatment and two years free of tumors has been reported (10).

The clinical diagnosis is complex because the symptoms are not specific. Symptoms include abdominal pain, gastrointestinal tract bleeding, intestinal obstruction, weight loss, jaundice, emesis, fever, diarrhea, anemia, palpable masses,

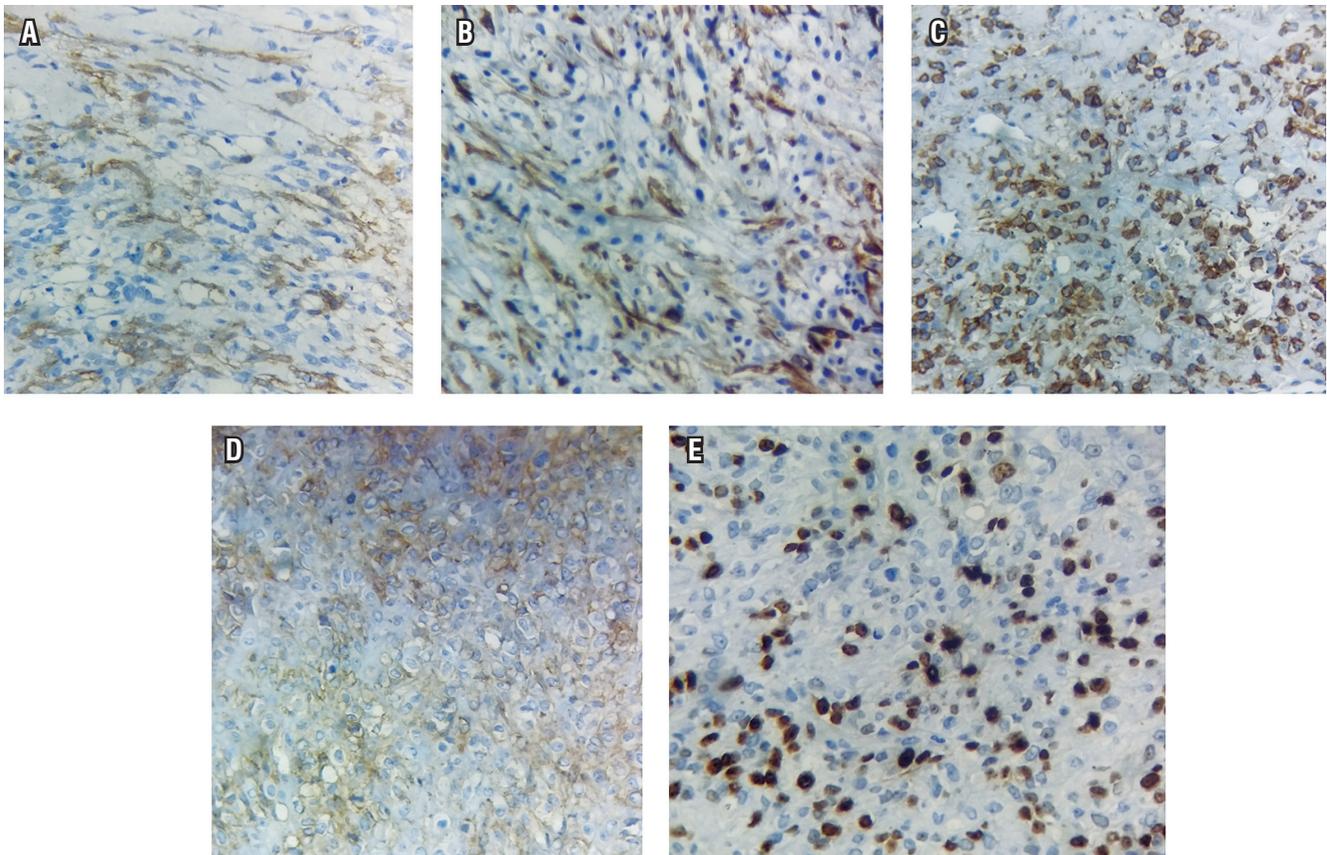
and fistulas (10, 16). Diagnoses using the various endoscopic techniques are not always conclusive (5), and even though CT scans and MRIs may show masses in the duodenum, a definitive diagnosis must be made through histopathological examination, specifically by immunohistochemistry (2).

Macroscopically, leiomyosarcomas are characterized by their irregular appearances that are lobed with sectors of necrosis (21). Microscopically these tumors are clearly distinguished from benign analogs by high cellularity and their cytological features (3). The morphologic spectrum ranges from fusiform to epithelioid variants (1). Tumor cells are characterized by eosinophilic cytoplasm and hyperchromatic elongated nuclei and nuclear pleomorphism (19). Mitotic activity is often greater than 50 mitoses per 10 high power fields. Mitotic counts over 10 in 10 high power fields are associated with tumor size. Compromised tissue at deeper levels and metastases indicate poor prognoses (19, 20).

The immunohistochemistry of leiomyosarcomas is positive for smooth muscle actin, desmin, muscle specific actin, caldesmon, calponin and CD34. In both epithelioid and spindle cell areas, fifty to sixty percent are positive for CD 138, twenty-five percent are positive for calretinin (25%), and thirty percent are positive for CD 99. They rarely test positive for positive for CD117 or HMB45, but this can be seen in epithelioid areas (22, 23). Some cases also test positive for cytokeratins (23).

Differential diagnosis is primarily concerned with GISTs since they are the most common mesenchymal tumors. This requires immunohistochemistry in which over 95% of these tumors positively express CD 117 and DOG1 (24).

In 1935 Whipple described a pancreatoduodenectomy for treatment of duodenal neoplasms (6). Nevertheless, there is



**Figure 4.** Positive immunohistochemical studies of tumor cells. **A** Smooth muscle actin. **B.** Desmin. **C.** CD99. **D.** CD138. **E.** Ki67 cell proliferation index of 60%.

no current consensus on appropriate surgical treatment for these tumors. Treatment options include tumor excision, segmental resection of the duodenum and pancreatoduodenectomy. Management with radiation therapy has demonstrated therapeutic utility locally, but has not been proven to have an impact on long-term survival (17, 19).

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