Case Report

Primary Leiomyosarcoma of left sided colon: a case report

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ABSTRACT

Leiomyosarcomas are rare smooth muscle tumors that represent 0.1% of all colorectal malignancies. The lack of literature available concerning leiomyosarcomas presents a challenge when diagnosing and treating these tumors, thus it is crucial that we differentiate them from gastrointestinal stromal tumors (GIST), the most common type of mesenchymal neoplasms of the gastrointestinal (GI) tract, especially when considering leiomyosarcoma's high prevalence of recurrence and malignancy. In this article, we present a case of a 69-year-old female with a diagnosis of sigmoid colon leiomyosarcoma. We analyze the tumor's CT imaging findings as well as correlation with the patient's pathological findings including immunostains, size, and mitotic activity, as well as the patient's risk for recurrence.

KEYWORDS

Leiomyosarcoma, Gastrointestinal stromal tumors, Smooth muscle tumor, GI tract

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INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most prevalent type of mesenchymal neoplasms in the gastrointestinal (GI) tract [1]. However, it wasn't until the late 1990s when Hirota et al. introduced the presence of KIT-mutations, which allowed for a distinction between GIST and leiomyosarcomas(LMS) to be made [2]. Since its differentiation in 1998, leiomyosarcoma, a true smooth muscle tumor, has accounted for less than 0.1% of all colorectal malignancies, which may account for the dearth of literature discussing the tumor's demographic, radiologic, and clinicopathological features. Proper diagnosis of leiomyosarcoma is critical given the tumor's aggressiveness and potential for recurrence [3,4]. In this article, we report the case of a 69-year-old female with a diagnosis of sigmoid leiomyosarcoma and correlate the clinical presentation, imaging and pathological findings.

CASE PRESENTATION

A 69-year-old female patient presented to our emergency department from Pokhara with complains of lower abdominal pain and bloody diarrhea for 3 weeks. She had a history of fall from standing height as she had to go to bathroom multiple times because of her diarrhea. She had a medical history of total hysterectomy, appendectomy, coronary artery bypass grafting, hypertension and seizure disorder, but with no drug allergy. She denied a history of familial or genetic diseases, and a general physical examination showed no abnormal signs, except for obvious tenderness in the left lower abdomen at palpation.

Hematological and blood biochemical tests revealed no abnormalities but hyponatremia. Carcinoembryonic antigen 2.1 ng/mL and carbohydrate antigen (CA19-9) <2.00 U/mL were within the normal limits.

Dynamic contrast-enhanced computed tomography (CT) revealed a heterogeneously enhancing polypoidal mass measuring about 38.7×31×15.8 mm in size noted in mid part of sigmoid. The mass acted as a lead point and caused telescoping of sigmoid colon into sigmoid colon and rectum. Length of intussusception measured 10.4 cm. Multiple intraluminal air foci were noted around the mass.; this lesion contained a solid area that was found to be hyperenhanced in the arterial phase and hypoenhanced in the portal phase. There were no feature suggestive of distant metastasis on computed tomography.(Fig. (Fig.1).1). These findings led to a suspected diagnosis of malignant lesions of the sigmoid colon. Therefore, resection of the tumor was performed by laparotomy and Hartman's procedure was done.

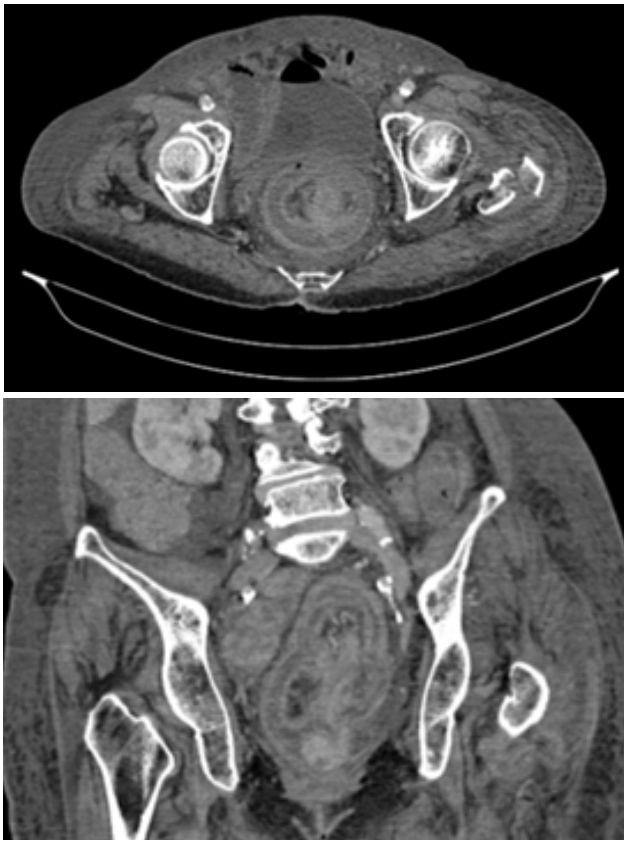

Fig.1: Heterogeneously enhancing polypoidal mass causing telescoping of sigmoid colon into sigmoid colon and rectum.

On gross examination, a polypoidal mass measuring 4 x 3 cm with stalk(1.5 cm)which was 10 cm from proximal margin and 20 cm from distal resected margin. Cut surface of polypoidal mass appeared greyish white with ulcerated mucosal surface and multiple pericolic lymph node were identified. Microscopically, there were spindle neoplasm arranged in fascicles with eosinophilic cytoplasm, moderately pleomorphic nucleus with clumped chromatin and inconspicuous nucleoli (Fig. 3). Area of necrosis and cystic degeneration were noted. Numerous mitosis seen with overlying ulcerated mucosa (Fig.3). Immunohistochemical examination was positive for smooth muscle actin (SMA), Desmin, Ck(focal) and Caldesmon. Stainings were negative for CD117, DOG-1, CD34, S-100, MyoD1 and SS18. The proliferation marker Ki-67 was detected in 45% of all examined tumor cells. There was no regional lymph node metastasis (0/19). These findings were consistent with the diagnosis of an LMS of the sigmoid colon.

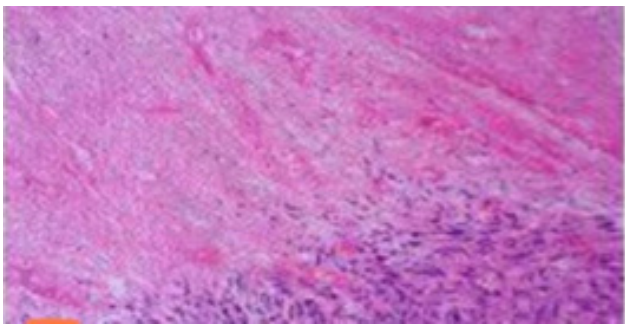

Fig. 2: Area of necrosis and cystic degeneration with numerous mitosis

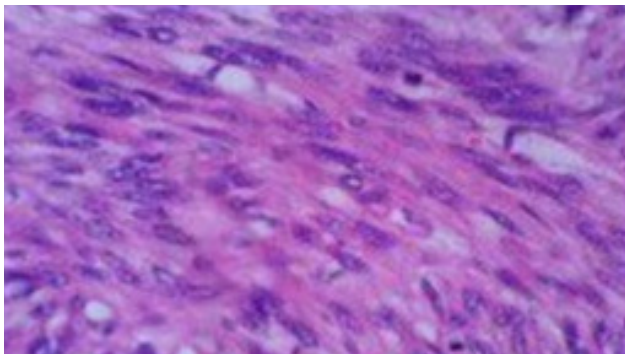

Fig. 3 Histological image showing tumor cells being spindle like and elongated with round nuclei and coarsened chromatine.

DISCUSSION

LMS occurs mainly in the fifth and sixth decade of life, and abdominal pain and gastrointestinal bleeding have been reported to be the most common clinical signs at presentation. Many smooth muscle tumors previously reported as LMS in the era of pre-GISTs were reviewed and proved to be GISTs, since these tumors have similar gross and microscopic appearance.[2] The diagnosis of LMS depends on accurate differential diagnosis from other sarcomas, and especially from GISTs.[5] The vast majority of smooth muscle tumors arising in the gastrointestinal tract are GISTs, defined by immunohistochemical positivity for KIT, CD34, CD117, and DOG1, and sometimes by molecular evaluations of activating mutations in the KIT or PDGFRA genes. Immunohistochemical features of LMS included positivity for desmin, alpha-SMA, vimentin, and h-caldesmon, and negativity for GIST markers-KIT, CD34, CD117, and DOG1.[6] GISTs occur most commonly in the stomach (60%–70%), followed by the small intestine (20%–25%), the rectum, and anus (4%), and are rare in the esophagus (1%) and colon (1%). In contrast, LMSs occur preferentially in the small intestine (45%) and colon (38%), and are extremely rare in the stomach and esophagus.[7] LMS of the colon is a rare malignant entity with unfavorable prognosis, and responds poorly to conventional chemotherapy or radiation therapy. Only a few cases of the LMS of colon have so far been reported. However, LMS accounted for 57.5% of a series of 433 patients with primary colorectal sarcoma reported between 1998 and 2012 in the National Cancer Data Base, USA by Thiels et al.[8] There are a few studies that defined the outcome of primary LMS patients alone. Grade and size are the most important prognostic factors for disease-specific survival and distant recurrence in patients with primary LMS.[9] The prognosis of patients with LMS in general seems to be correlated with the degree of histological grading. Site is not an important independent prognostic factor for local recurrence in this series.[10]

Surgery is the main treatment for primary colonic LMS, as most of the reported cases were diagnosed using surgically resected specimens. While adjuvant chemotherapy and/or radiation are often used in the management of primary colonic LMS, there are conflicting data on their efficacy and impact on overall survival. LMSs of the colon have been previously reported to exhibit a great potential of local recurrence. Effective treatment strategies for LMS are difficult to develop due to lack of data. So far several cytotoxic chemotherapy regimens have been described.[11] However, LMS is relatively insensitive to chemotherapy.[3] Our case did not receive any chemotherapy and radiation therapy after surgery. Long-term follow-up of LMS patients is very important.

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