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Case report

Primary leiomyosarcoma of the sigmoid colon: Case report and review of literature [☆]

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ABSTRACT

Leiomyosarcomas of the sigmoid colon are aggressive and extremely rare tumors representing less than 0.1% of all colorectal malignancies. Few cases have been reported in the literature and their imaging features need to be more detailed. We report the case of a 70 year old male patient, smoker, with a history of arterial hypertension, admitted for abdominal pain and hypogastric mass. Computed tomography has shown a voluminous heterogeneously enhancing tumor process with a necrotic center, attached to the sigmoid wall, with multiple secondary peritoneal and hepatic masses. The anatomopathological examination of the biopsy sample with immunohistochemistry allowed the diagnosis of leiomyosarcoma of the sigmoid colon. Clinical presentation and radiological features of leiomyosarcomas are non-specific, and the definitive diagnosis is only established after an anatomopathological examination.

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Introduction

Primary gastrointestinal leiomyosarcomas are extremely rare, and the colorectal localization is exceptional. They represent less than 0.1% of all colorectal malignancies[1].

These mesenchymal tumors originate from smooth muscle cells in the wall of the digestive tract.

Due to their rarity and the minimal number of confirmed cases reported in the literature after the discovery of the entity

of gastrointestinal stromal tumors (GIST), these tumors can present both a diagnostic and therapeutic problem. The role of the imaging is to guide the diagnosis and to assess the tumor extension.

Case report

A 70-year-old male patient, smoker, with a history of arterial hypertension, admitted for diffuse chronic abdominal pain

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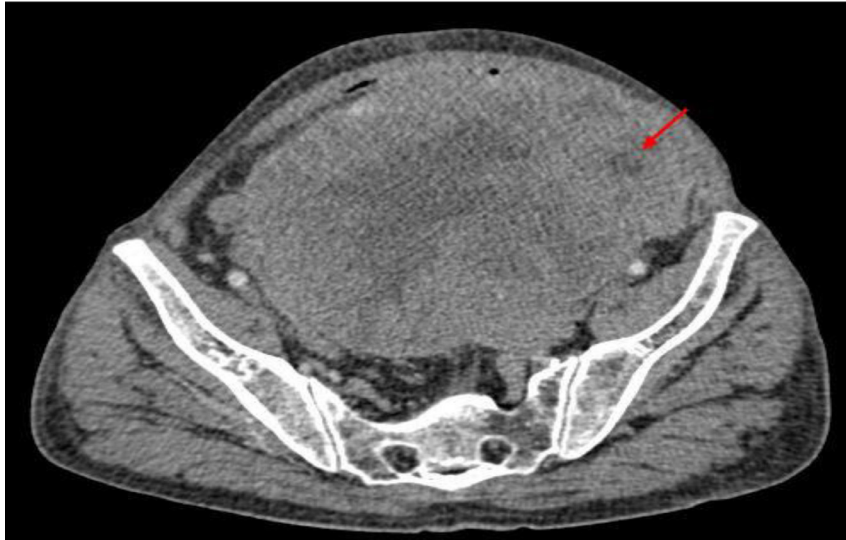


Fig.1 – CT scan of the abdomen showing a voluminous tumor process with a necrotic center attached to the sigmoid wall, with exophytic development, irregular contours, enhancing heterogeneously after injection of the iodinated contrast agent

with progressive abdominal distention and deterioration of general condition.

Clinical examination revealed a distended abdomen with a fixed hypogastric mass and sensitivity to palpation of the hypogastric and iliac fossae.

The biological balance was without abnormality. Even tumor markers including carcinoembryonic antigen (CEA) and carbohydrate antigen 19–9 (CA 19–9) were negative.

An abdomino-pelvic CT initially performed has shown a voluminous tumor process with a necrotic center, attached to the sigmoid wall, with exophytic development, irregular contours, enhancing heterogeneously after injection of the iodinated contrast agent. (Fig. 1)

The CT scan also showed the presence of multiple secondary peritoneal (Fig. 2) and hepatic masses (Fig. 3).

A biopsy sample was taken and the pathological study has shown a tumor proliferation of fusocellular architecture with focal cytonuclear atypia and large foci of tumor necrosis (Figs 4, 5, 6). An immunohistochemical analysis allowed the diagnosis of leiomyosarcoma of the sigmoid colon showing that tumoral cells are positive for SMA, Desmine and H-caldesmon and negative for CD117 and DOG1. (Fig. 7)

Given the presence of metastasis, surgical treatment could not be performed, then chemotherapy was started.

The patient still presents a deterioration of the general condition with abdominal pain and vomiting.

Discussion

Sarcomas are malignant tumors that arise in connective tissue in the body, they account for less than 1 % of all adult cancers. These tumors arise mainly in the soft tissues in at least 70 % of cases[2].

The most common site of digestive leiomyosarcomas is the small intestine (45 %) and the colon (38 %)[3].

Leiomyosarcomas are often diagnosed in middle age patients (50–60 years) with a slight male predominance[3].

These tumors, which remain asymptomatic in the beginning may present with gastrointestinal bleeding, pain, abdominal mass, intestinal obstruction and / or deterioration of general condition at a later stage.

Leiomyosarcomas can have an exophytic development without any endoluminal component, thus they are in certain cases inaccessible to endoscopic examinations, and the diagnosis can be more delayed.

Ultrasonography as an accessible and non-invasive imaging modality can be used initially to detect the mass, its location, size and depth, with a low specificity for characterizing the nature of the tumor. Leiomyosarcoma appears as a heterogeneous hypoechoic mass with central anechoic areas of necrosis. Intra-tumor bleeding can be detected as a hypo, iso or hyperechogenic areas[4].

The CT scan allows the detection of the tumor, the assessment of locoregional tumor extension at the same time with the search for distant metastatic localization.

Leiomyosarcomas appear in the form of a large heterogeneous mass, hypodense, eccentric, with heterogeneous or homogeneous enhancement, which may contain central necrosis or calcifications, with significant local extension. Lymph node involvement is not common[4,5].

Magnetic resonance imaging (MRI) shows a voluminous mass hypointense on T1, hyperintense on T2, with central T2 hyperintense areas of necrosis, and heterogeneous enhancement[4].

The definitive diagnosis is only established after an anatomicopathological examination of the biopsy sample or the resected specimen.

As for immunohistochemical analysis, leiomyosarcomas are characterized by the positivity of desmin, alpha-SMA, vimentin and h-caldesmon, and the negativity of GIST markers, in particular KIT, CD34, CD117 and DOG1 [6].

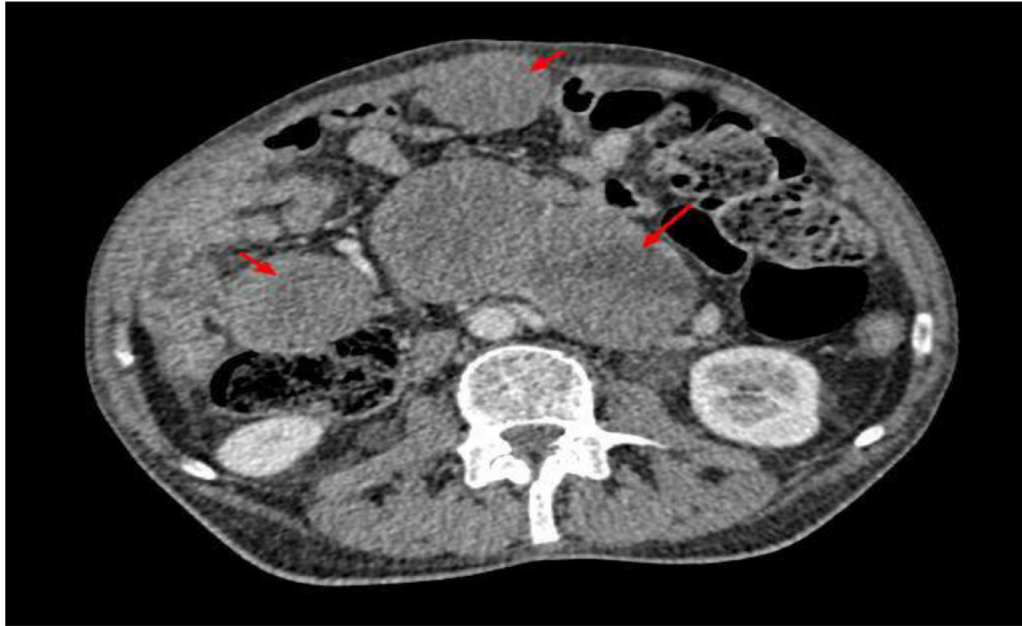


Fig. 2 – CT scan of the abdomen showing the secondary peritoneal masses

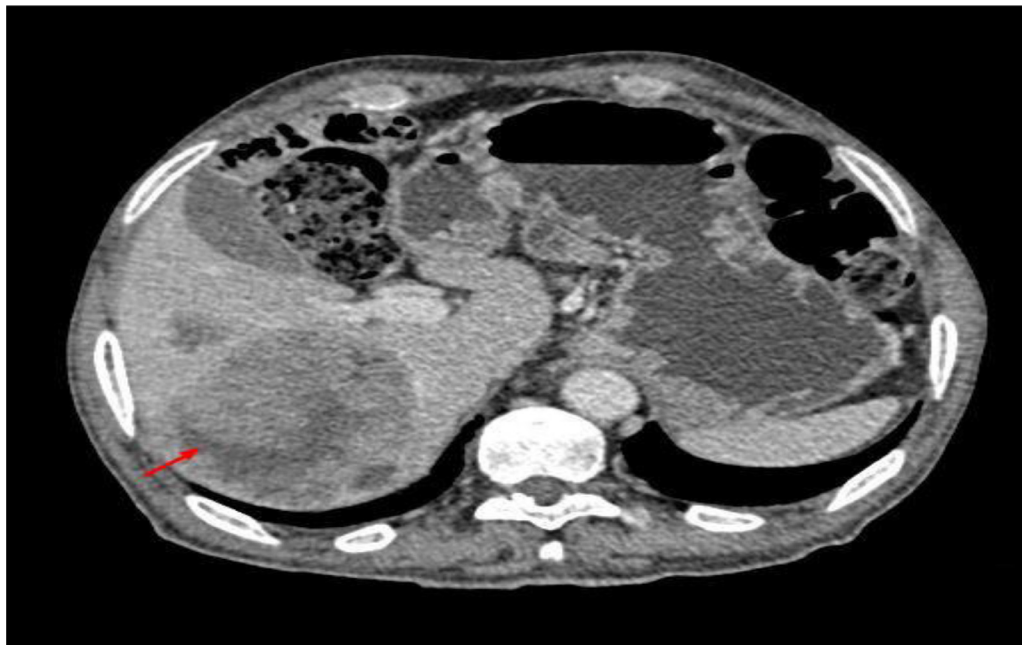


Fig. 3 – CT scan of the abdomen showing a secondary hepatic mass

Treatment remains focused on radical surgical resection. However, given the proportion of cases with late presentations, curative surgery is not always possible [6].

Radiotherapy and chemotherapy for sarcomas do not affect the course of aggressive tumors. The standard chemotherapies for advanced soft tissue and visceral sarcomas are first-

line anthracyclines, with doxorubicin-plus-dacarbazine as an alternative[7].

Leiomyosarcomas are aggressive tumors, and prognostic factors are not well studied due to their rarity, but survival is worse compared to adenocarcinoma patients[8]. They have significant metastatic potential with 50 % distant metastatic cases in 5 years[9].

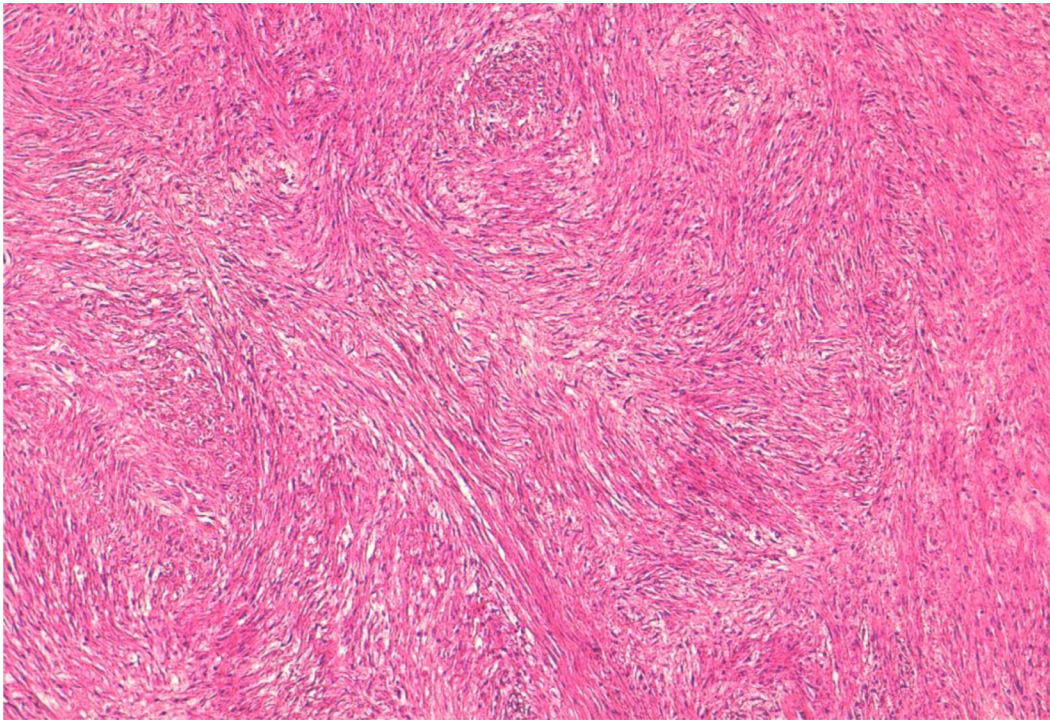


Fig. 4 – Tumoral proliferation composed of fascicles of spindle cells, exhibiting smooth muscle differentiation

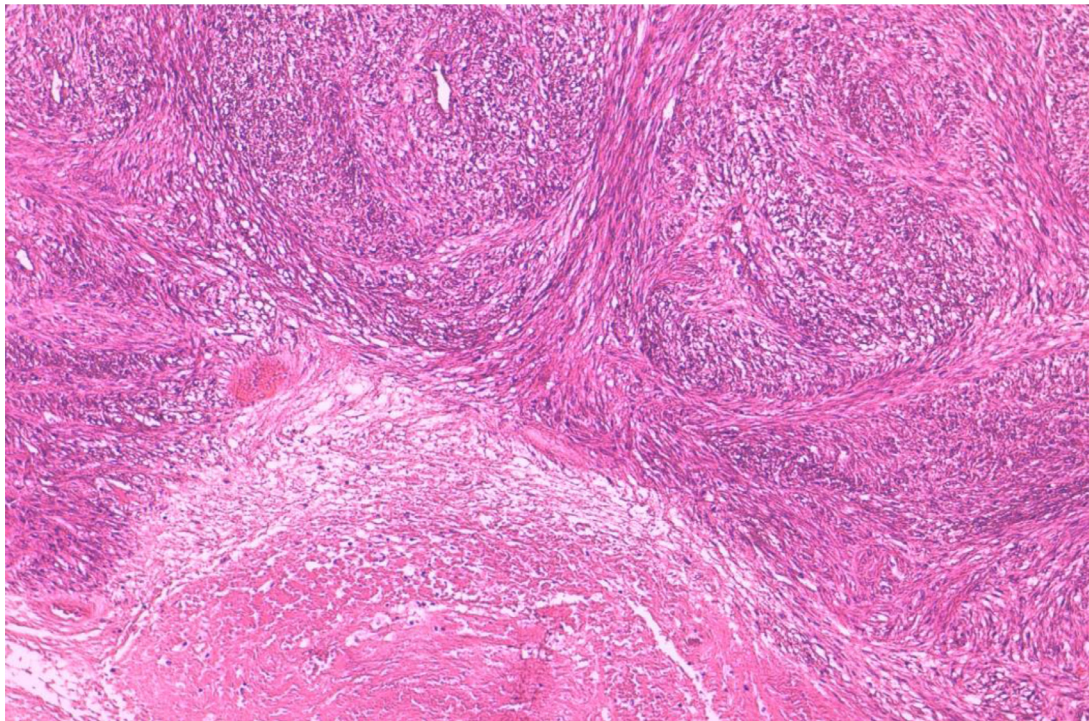


Fig. 5 – Areas of tumoral necrosis

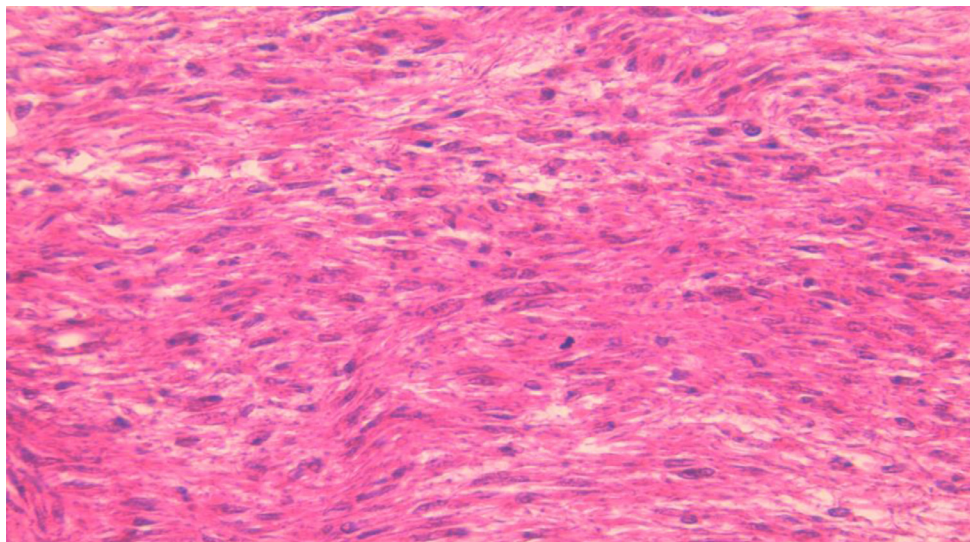


Fig. 6 – The tumoral cells are provided with cigar-shaped nuclei, showing moderate to severe atypia, and an abundant eosinophilic cytoplasmic. Numerous mitotic figures can be identified

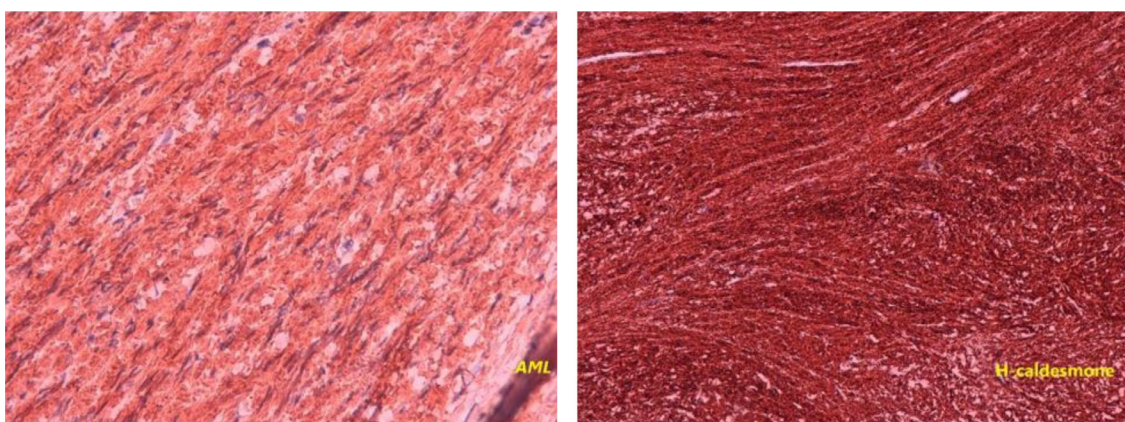


Fig. 7 – The tumoral cells are positive for SMA, Desmine and H-caldesmon, They are negative for CD117 and DOG1

Conclusion

Leiomyosarcomas of the sigmoid colon are not well studied due to their rarity, and their imaging findings are not well known. The case we presented here is an aggressive leiomyosarcoma with intra-abdominal metastasis. The radiological presentation allowed suspecting a malignant origin of the mass, and the diagnosis was established after anatomopathological examination. More cases need to be published to have enough data available to study these rare and aggressive tumors.

Authors' contributions

All authors have made a significant contribution to this manuscript. All the authors have read and approved the final version of the manuscript.

Patient consent

An informed consent was obtained from the patient.

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