Case Report

Leiomyosarcoma of the rectosigmoid junction, a case report✩,✩✩

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ABSTRACT

Leiomyosarcomas are a rare type of neoplasm of the digestive tract with an estimated presentation of 0.1% predominantly between the fifth and sixth decade of life [1]. We report the case of a 61 years old male patient without previous medical conditions, with melena and constitutional symptoms and an extensive rectal mass with a final diagnosis of diagnosis leiomyosarcoma of the large intestine, grade II (FNCLCC). Given the rarity of this pathology and the fact that it does not present specific clinical, endoscopic, and imaging features made it difficult to differentiate from other entities of greater frequency such as rectal adenocarcinoma and GIST, representing a real diagnostic challenge.

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Introduction

Leiomyosarcoma (lei-: smooth; -myo-: muscle; sarcoma: malignant connective tissue tumor) is a malignant neoplasm derived from smooth muscle cells. Of all soft tissue sarcomas, approximately 5%-10% are leiomyosarcomas originating from smooth muscle cells in the walls of small blood vessels, the gastrointestinal tract, and the uterus [2].

Gastrointestinal leiomyosarcoma is a rare tumor. Its most common location is the stomach, followed by the small intestine, only 11% of cases occur in the rectum [3] constituting 0.1% of all malignant colorectal tumors [1].

Case report

A 61-year-old male patient, smoker, with a history of longstanding melena of several years, associated with fever, weight loss, and pelvic and lumbar pain of 3 months of evolution. On general physical examination, mucocutaneous pallor was noticed, normal heart sounds, ventilated lung fields, and no evident abdominal alterations were also noted. On proctologic examination, a mass with an irregular surface was detected, located 3 cm from the anal margin. The laboratory tests carried out showed anemia (Hemoglobin 9 gr/dL and Hematocrit 29.4%), Leukocytosis (14,800 per mm³), increased Alkaline phosphatase (186.4 mg/dL) and AST and ALT formerly are called serum glutamic oxaloacetic transaminase (GOT) and serum glutamic pyruvic transaminase (GPT), respec-
tively. Both were increased AST/GOT: 56.8 IU/L and ALT/GPT: 67.9 IU/L.

An abdominal ultrasound was performed, although no mass was detected, it revealed heterogeneous mural thickening of the rectosigmoid junction with an average thickness of 30 mm. The liver and remaining abdominal structures were normal. However, this sonographic examination had limitations due to a large amount of bowel gas and the location of the mass, further investigations were suggested.

Suspecting a possible rectal adenocarcinoma, a colonoscopy was performed which confirmed the presence of a tumor with irregular edges that extended from the sigmoid to the lower rectum with an approximate measurement of 25 cm, obstructing 80% of its lumen. A biopsy sample was taken, the results revealed a malignant neoplasm of a sarcomatoid pattern.

Abdominal computed tomography (CT) revealed an extensive and well-defined solid mass (29 HU) on the anterior wall of the rectosigmoid junction located 20 cm from the anal ring, heterogeneous with gas at its periphery. With a slight increase during contrast administration (46 HU), it showed dilatation and lumen obstruction. This lesion measured 11 × 9 × 10 cm in its main axes. Some paraaortic lymph nodes were noted as well as a left kidney cyst (Figs 1, 2 and 3). The preoperative metastatic workup did not report abnormalities.

Low anterior resection plus end-to-end colorectal anastomosis was performed. Among the intraoperative findings are a large pediced tumor dependent on the anterior wall of the rectosigmoid junction with a measure of 15 × 15 cm; inside the mass, there was purulent material. Fifteen mesorectum lymph nodes were resected, other structures were undamaged.

The biopsy of the surgical specimen concluded leiomyosarcoma versus gastrointestinal stromal tumor (middle grade). It also revealed that one of the 15 resected lymph nodes was infiltrated by the tumor. The immunohistochemical study reported a tumor negative for CD117 (CKit), CD34, and S-100 (polyclonal) and positive for smooth muscle actin (1A4), with 25 mitoses in 10 high-power fields; being the definitive diagnosis leiomyosarcoma of the large intestine, grade II (FNCLCC) (Figs 4 and 5). The postoperative period elapsed without incident and the patient was discharged and referred for radiotherapy.
Fig. 4 – Histological image with H&E (Increase 100 x) showing primary leiomyosarcoma of rectosigmoid, fascicular growth pattern. 400 x presenting fusiform cells with a variable degree of cytological atypia and atypical mitosis.

Fig. 5 – The immunohistochemical study reported a tumor negative for CD117 (CKit), CD34 and S-100 (polyclonal) and positive for smooth muscle actin (1A4), with 25 mitoses in 10 high-power fields.

Discussion

Leiomyosarcoma of the rectum is extremely uncommon, it represents approximately 0.1% of malignant neoplasms of the colon [1]. It usually occurs in patients older than 50 years. Prognosis and treatment vary depending on location, stage, and grade of the primary tumor, as well as the presence of metastases; it usually behaves aggressively, worsened by an early presentation in patients under 50 [4] with a low 5-year survival rate between 20% and 40% [5].

The most common route of spread of rectal leiomyosarcoma is the hematogenous, being liver the organ most frequently affected. The lymphatic route and direct spread to adjacent organs have been rarely reported [3]. Our patient had one regional lymph node infiltrated by the tumor when the low anterior resection was performed and multiple metastatic hepatic nodules, pleural effusion and ascites were detected in the follow-up studies, there were no signs of other solid organs involvement.

The diagnosis of gastrointestinal leiomyosarcoma is a challenge, in addition to its rarity, there are no pathognomonic radiologic features and it does not present specific clinical and endoscopic findings. CT scan features of leiomyosarcoma are nonspecific; they are usually solid heterogeneous masses, with necrosis [6]. Therefore, the importance of imaging studies lies in evaluating the extent of the disease, as well as the feasibility of its resection, and to make decisions about its management.
Due to the lack of pathognomonic radiological features is initially confused with other entities of greater frequency such as rectal adenocarcinoma and GIST, as occurred in this case, and the fact that initial biopsies are often negative or frequently inconclusive reiterates the need to study the surgical specimen and immunohistochemistry for its definitive diagnosis [1,6]. In our patient the final diagnosis was established based on immunohistochemical findings.

Immunohistochemical tests, unlike gastrointestinal stromal tumors, in leiomyosarcoma, the determination of c-Kit (CD 117), and S100 is negative and positive for actin and desmin. Associated with clinical behavior, the presence of nuclear atypia and the presence of 2 or more mitoses per field with the objective of 100 x determine the criteria for malignancy of this neoplasm [7,8]. In our case, the immunohistochemical study revealed 25 mitoses in 10 high-power fields. Early and extensive surgery continues to be the most accepted management [6,8].

Conclusion

The case presented here was an extremely rare entity, with few cases published in medical literature and no data about its prevalence in our setting, which represented a diagnostic challenge, but thanks to immunohistochemical techniques, a definitive diagnosis was reached.

Fig. 8 – Axial images of abdominal CT without contrast that show metastatic hepatic nodules and ascites (white arrow).

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