Case report

The double challenge (preoperative diagnosis and surgical approach) of primary leiomyosarcoma of the sigmoid colon

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Abstract. *Background and aim:* Primary colonic leiomyosarcoma (cLMS) is a rare malignancy of *muscularis mucosae* or *muscularis propria* showing highly aggressive behaviour and poor prognosis. To date, making a preoperative diagnosis and performing the most appropriate treatment represent laborious tasks for the clinicians. On the one hand, in fact, cLMS diagnosis is often difficult to achieve preoperatively because of the low specificity of clinical, radiological and bioptical features: for these motives, the diagnosis is usually obtained at postoperative histology/immune-histochemistry. On the other hand, although surgery represents the mainstay of multi-modal treatment, in the current era of minimally invasive surgery the optimal approach to cLMS is debated: in the absence of a standardized and unanimous algorithm, in fact, laparoscopy is usually proposed for small tumors, whereas laparotomy for masses exceeding 4 cm in diameter. Our aim was to elucidate such two aspects by reporting our experience. *Methods:* We present the case of a 51-year-old man affected with a 6-cm LMS of the sigmoid colon. *Results:* Preoperative diagnosis was achieved through a preoperative echo-endoscopic biopsy. The lesion was successfully and safely managed by laparoscopic surgery. *Conclusions:* Our case suggests that a preoperative diagnosis of cLMS is possible in an appropriate setting. Moreover, laparoscopy seems to be a safe and successful approach to resect cancers even larger than the common 4 centimetres proposed by the current literature. (www.actabiomedica.it)

Key words: Leiomyosarcoma; large bowel; laparoscopy; surgery

Introduction

Primary leiomyosarcoma of the colon (cLMS) is a rare cancer arising from muscularis mucosae or muscularis propria of the large bowel wall having a highly aggressive behaviour and poor prognosis (1). Diagnosis of cLMS is seldom obtained in the preoperative setting because of the lack of specificity of symptoms, imaging and biopsy; most times, in fact, the nature of this soft-tissue sarcoma is elucidated only with conclusive histology and immunostaining (2). As of 2021, after reviewing the available relevant literature, surgery appears the treatment of choice (3). However, the type of surgical approach (open versus minimally invasive procedure) still is a matter of study (3). Hereafter we present a case of a 6-cm-sigmoid LMS detected by means of a preoperative echo-endoscopic histology and successfully managed by laparoscopic surgery.

Case Presentation

A 51-year-old caucasian male presented with a three-month-history of aspecific abdominal pain and

bloating. US showed a 6-cm-mass located in left lower abdomen, near the sigmoid colon. His past medical history was negative except for smoking addiction. Physical examination of the abdomen was negative. Colonoscopy showed a granular lesion in medium sigma mucosa (histology on bioptic sample was negative for neoplastic tissue). Tumor markers (CA 19-9, CEA) serum level were within normal limits. Abdominal CT-scan confirmed the presence of a 6x5.5x6.5 cm solid mass connected to the posterior wall of medium sigmoid colon (Fig. 1a). Endoscopic ultrasound showed a roundish, extra-mucous inhomogeneous sigmoid mass, predominantly hypo-ecogenic with non-echogenic areas and hyper-echogenic spots (Fig. 1b). Histology examination on fine needle biopsy reported a low-grade mesenchymal neoplasia with leiomuscular phenotype. Staging CT scan excluded liver/pulmonary metastases.

Following multidisciplinary meeting discussion, the patient underwent laparoscopic left hemicolectomy. A traditional 5-trocar-laparoscopic approach was used. The first trocar (12 mm) was placed in the umbilicus according to the "open" technique, followed by CO2 insufflation (12 mmHg pneumoperitoneum). A second 12 mm trocar was located in the right lower abdomen and three additional 5-mm trocars were placed in patient left side, right hypocondrium and suprapubic area. At abdominal exploration, a bulging mass, indiscernible from the sigmoid colon, apparently reaching the peritoneal layer, was identified in the absence of distant metastasis and carcinosis. The inferior mesenteric vessels were dissected by harmonic scalpel, clipped and severed. The left colonic flexure was completely mobilized by transverse mesocolon root section at the anterior aspect of the pancreas. Colonic recto-sigmoidal junction was dissected and the proximal rectum was sectioned by linear stapler 10 cm distally to the neoplasia, and pulled out through a Pfannenstiel incision. Proximal colon section was performed after evaluating its vascularization via indocyanine green test. An end-to-end mechanical anastomosis by double stapling according to Knight-Griffen technique was performed. A para-anastomotic Jackson-Pratt drain was left in place. Histolopatological examination recorded a 6x5.7x5.5 cm grey, nodular neoplasia (Fig 1c). Macroscopically, a heterogeneous mass, containing both cystic and solid areas, was described. Microscopically, the tumor presented a thin peripheral layer characterized by spindle cells with aspects of leiomuscular differentiation and a prevalent central part consisting of great, atypical, pleomorphic and, sometimes, multinucleated cells. Scattered areas of necrosis and myxoid stroma were reported. Immunohistochemical pattern tested positive for smooth muscle actin (SMA), caldesmin, desmin and EMA, instead, it was negative for DOG1, c-kit, S100. The Ki67 index was 30-40% (Fig 2a-b).

The neoplasia was arising and limited to the muscolaris propria, without lymphovascular invasion and no lymph node metastasis (0/12 lymph nodes examined) was observed. Final diagnosis was poorly differentiated - G3 - cLMS according to the French Federation of Cancer Centers Sarcoma Group (FNCLCC). The patient had flatus on postoperative day (POD) 1, but suddenly presented hyperpyrexia (38°C) on POD 3. Hemocolture was negative and abdominal CT scan was negative for anastomotic dehiscence or deep collection. On POD 4, a superficial, incisional collection was evacuated. On POD 5 the patient had bowel movements, on POD 6 the drain was removed

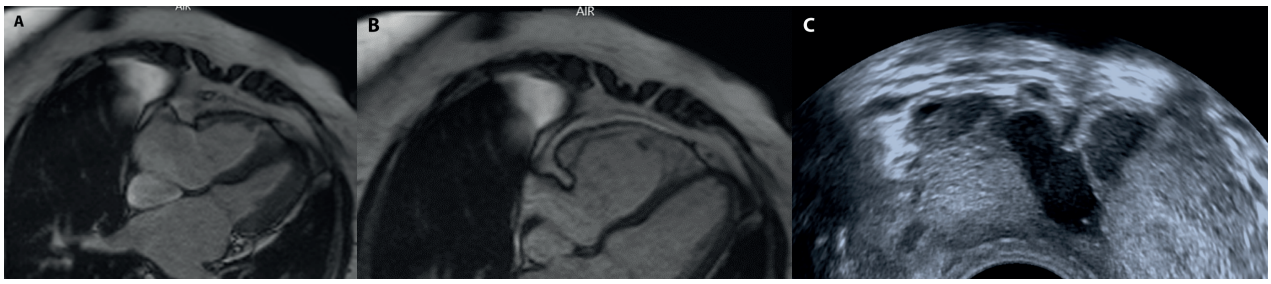

Figure 1. 1a. Contrast-enhanced computed tomography scan showing the lesion and its relationship with the sigmoid colon wall; 1b. Endoscopic ultrasound feature of the lesion; 1c. Macroscopic view of the surgical specimen showing the neoplastic lesion.

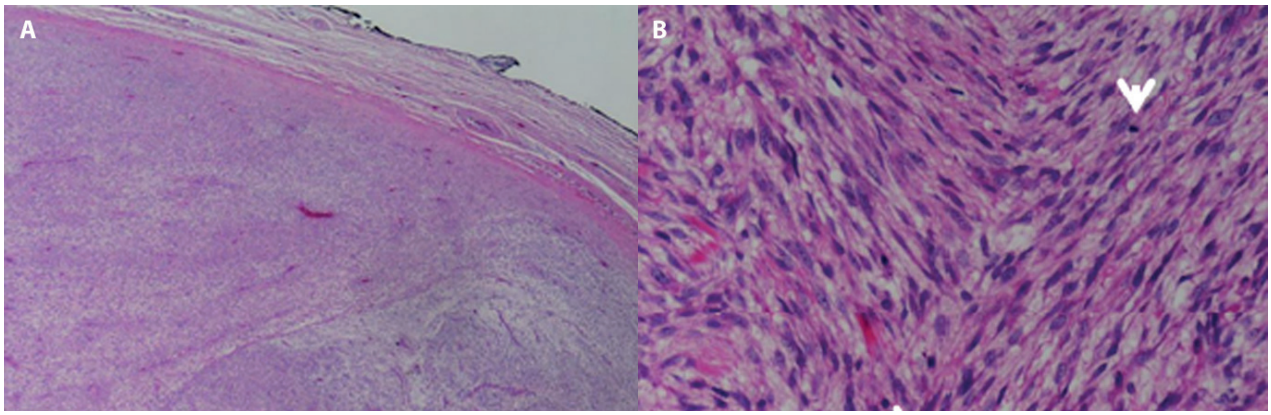

Figure 2. 2a. Microscopic aspect: hematoxylin-eosin staining revealed spindle-shaped cells with aspects of leiomuscular differentiation and great, atypical, pleomorphic and, sometimes, multinucleated cells. 2b. At immunohistochemistry, tumor stained positive for α-SMA, caldesmin and desmin.

and, on POD 10, the patient was discharged. The patient is doing well 12 months after surgery.

Discussion

Leiomyosarcoma (LMS) is a rare malignant tumor of smooth muscle representing only 0.1 to 3% of all gastrointestinal malignancies (1). LMSs reside preferentially the small and large intestine (45% and 38%, respectively) while stomach and esophagus are rarely affected (4). Colorectal LMSs (cLMS) represent only 0.12% of all colorectal malignancies: the sigmoid and transverse colon comprise the most frequent locations of the large bowel (5,6). Historically, before the discovery of the oncogenic role of KIT in gastrointestinal stromal tumors (GISTs), many smooth muscle neoplasms, such as LMSs, were misdiagnosed (2); nowadays, on the contrary, diagnosis of LMS is easily performed by immunohistochemistry. Unfortunately, several important factors affect the achievement of a preoperative diagnosis of cLMS. First, cLMS origins from the muscular layer of the large bowel wall; hence, from an intraluminal-endoscopic point of view, the colonic mucosa appears usually intact. Second, radiological features (especially computed tomography) of cLMS features are non-specific as well. Third, whenever a stromal tumor is suspected at imaging, endoscopic ultrasound could confirm the diagnosis of LMS but, unfortunately, adequateness of endoscopic sampling is far from being optimal reaching, for example, an impressively poor 29%-accuracy in the case of rectal LMSs (7). Moreover, biopsy may underestimate the grading of malignancy (8). Owing to these reasons, final diagnosis is mostly achieved postoperatively on specimen studied with definitive histology; this is especially the case of cLMS resected during emergency procedures. cLMS are considered aggressive bowel tumors with an estimated 5-year tumor-specific overall survival rate of 51.6% (9). Typically, they present a high local recurrence rate and significant hematogenous spread to the liver and lung at the time of diagnosis, whereas lymph node involvement is rare (1,10). Differentiation and size are the stronger predictors for disease specific survival and distant recurrence (11). The French Federation of Comprehensive Cancer Centers (FNCLCC - Fédération Nationale des Centres de Lutte Contre Le Cancer) grading system, which divides LMSs into three categories based on their differentiation, necrosis and mitotic rate, is the classification mostly frequently used (12). Literature does not propose a unique protocol for cLMS treatment. According to the ESMO-EURACAN clinical practice guidelines, radical surgery with complete tumor resection is considered the treatment of choice, possibly followed by adjuvant chemotherapy in the case of high-risk patients (8). In the relevant literature, surgical resection of cLMS is usually described through laparotomy. Interestingly, in 2019, Yahagi et al. reported a case of LMS of the sigmoid colon resected by laparoscopic surgery, claiming

that this approach should be used only for lesions <5 cm in diameter (3).

Conclusion

We successfully obtained a pre-operative histological diagnosis of cLMS thanks to endoscopic ultrasound biopsy and we made a completely safe laparoscopic resection. In elective surgery of cLMS the pre-operative histological diagnosis could play an important role in order to obtain an oncologically correct resection, which can be totally performed through a laparoscopic approach also in situation of large masses.

Conflict of interest: Each author declares that she or he has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

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