OPEN ACCESS ISSN: 2837-3642 Kong WK, et al. (2024): 172
Doi: 10.47991/2837-3642/100124

Case Report Annals of Clinical Reviews & Case Reports

Leiomyosarcoma of Sigmoid Colon with Acute Closed Loop Intestinal Obstruction and Pulmonary Embolism: A Diagnosis and Treatment Dilemma

Wai-Keong Kong[1*], Aini Natasha Abdullah[1], Nil-Amri Mohamed Kamil[1]

[1]Colorectal Surgical Department. Hospital Sultanah Bahiyah, Kedah, Malaysia, Km 6, Jalan Langgar, Bandar, 05460, Alor Setar, Kedah, Malaysia.

[*]**Corresponding author:** Wai-Keong Kong, Colorectal Surgical Department. Hospital Sultanah Bahiyah, Kedah, Malaysia, Km 6, Jalan Langgar, Bandar, 05460, Alor Setar, Kedah, Malaysia. Tel: +60 4-740 6233.

Citation: Kong WK, Abdullah AN, Kamil NAM (2024) Leiomyosarcoma of Sigmoid Colon with Acute Closed Loop Intestinal Obstruction and Pulmonary Embolism: A Diagnosis and Treatment Dilemma. Anna Clin Rev Cas Rep: ACRCR-124.

Received Date: 14 April, 2024; **Accepted Date:** 22 April, 2024; **Published Date:** 26 April, 2024

Abstract

Background: *Leiomyosarcoma is a rare colorectal cancer. The soft tissue tumour is less than 0.1 percents of all reported colorectal malignancy. They frequently presented with symptoms mimicking epithelial colon malignancy with preoperative diagnosis and surgical approach dilemma. The patient fitness and operability status leading additional difficulty in the surgical management.*

Purpose: *we report a case with colonic leiomyosarcoma closed loop intestinal obstruction and concurrent pulmonary embolism successful underwent radical curative resection.*

Conclusion: *Well-planned staged surgery for emergency emergencies setting with Multidisciplinary team work perioperative optimization demonstrated better surgical outcome*

Keywords: *Leiomyosarcoma, Colorectal cancer, soft tissue sarcoma.*

Introduction

Leiomyosarcoma (LMS) is a rare type of soft tissue sarcoma. Primary colonic leiomyosarcoma in general is a very rare entity, accounting for 1% of gastrointestinal malignancies and less than 0.1% of all colorectal malignancy. Surgery remains the best curative option. However, the overall prognosis remains poor. Due to its rarity entity, there are no consensus guidelines available for the optimum treatment approach. Operability and fitness of higher age group patients contributed additionally burden to the dilemma.

Case Report

We hereby reported a 64-years-old gentleman was presented to our Surgical Department with acute intestinal obstruction. The patient had a four-month history of altered bowel habit followed by gradually worsening intestinal obstruction symptoms for two weeks. He has underlying comorbidity of diabetes mellitus and hypertension with stable hemodynamic condition. General physical examination identified a distended abdomen with tenderness over lower abdomen, no mass on digital rectal examination. Serum Carcinoembryonic antigen 3 Ug/mL, carbohydrate antigen (CA19-9) 5 U/mL and cancer antigen (CA125) 34.6 U/ml were within the normal limits. Computed tomography (CT) Abdomen revealed circumferential distal sigmoid colon mass with closed loop large bowel dilatation. Complete occlusion of rectal contrast. (Figure 1 and Figure 2).

Figure 1: Computerize Topography Abdomen and Pelvis of the patient showing the long segment mass of distal sigmoid colon with complete luminal occlusion.

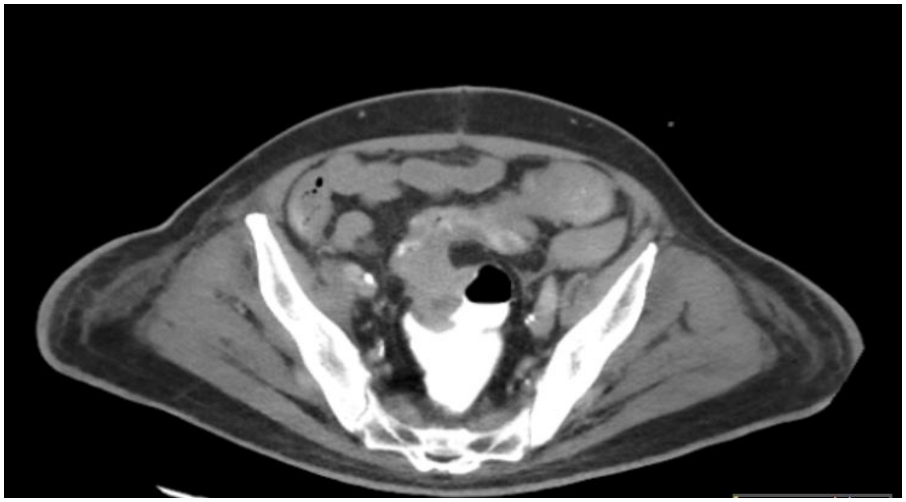
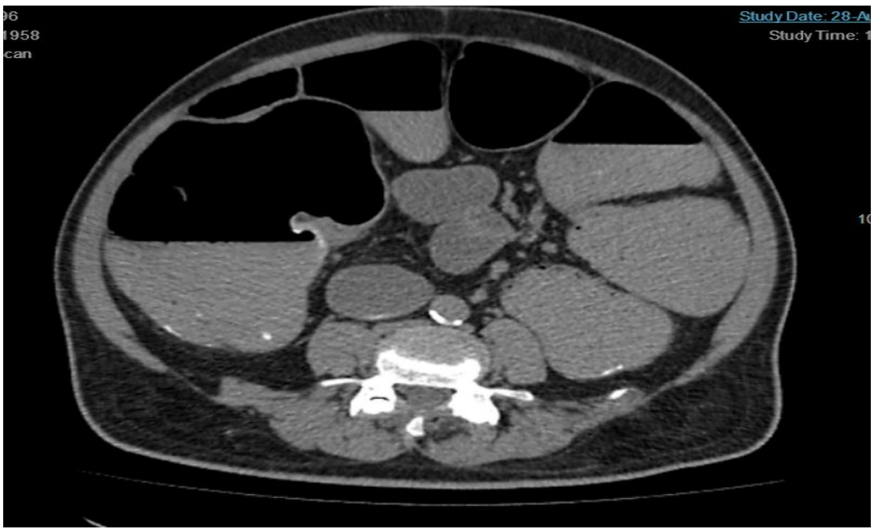


Figure 2: Computerize Topography Abdomen showing closed loop obstruction with gross proximal bowel dilatation.



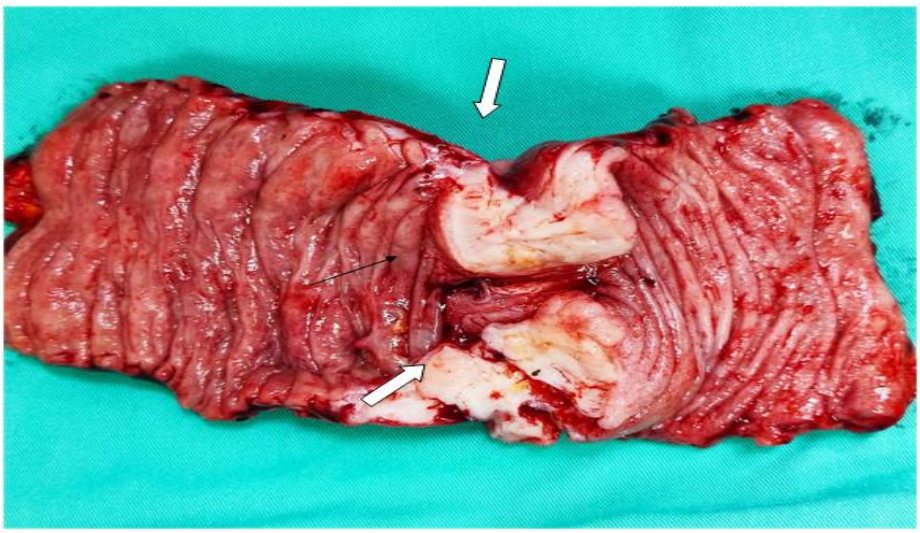
He was initially assigned to emergency resection, however intraoperatively developed hemodynamic instability with severe acidosis. There was a closed loop obstruction by constricting tumour at distal sigmoid without perforation. Decision made for damage control bowel decompression and transverse colostomy proceed by staged surgery. Postoperatively he required prolonged ventilation and intensive care for total 26 days secondary to pulmonary embolism and right Internal Jugular Vein thrombosis, severe Klebsiella pneumonia infection and COVID 19 infection.

Colonoscopy performed showed a complete occlusion large tumour 20 cm from anal verge. Biopsy was done twice with finding consistent with leiomyoma, unable to totally exclude the possibility of high-grade lesion or malignancy. Immunohistochemically, the spindle cells are positive to SMA, Desmin and h-Caldesmon. They are negative to Myogenin, EMA, S100, CD117 and CD34, DOG-1, CD117 and beta-catenin.

Multidisciplinary meeting with family conference for discussion for his acute condition had decided for upfront tumour resection. Decision made to defer his surgery for two months after completion of anti-coagulation treatment for his embolism for risk reduction and optimization. For preoperative nutrition and cardiopulmonary and neurological rehabilitation.

He then successfully underwent second stage laparotomy and sigmoid colectomy. On gross examination, there was mobile distal sigmoid colon tumour, circumferential submucosal hard tumour with serosal puckering (Figure 3).

Figure 3: Sigmoid colectomy specimen with long segment circumferential intramural pale-yellow tumour lesion with puckering at outer serosa. (white arrow) The bowel lumen complete occlusion with visible normal appearance colonic musoca. (thin black arrow)



Unanticipatedly his histopathology demonstrated the mass composed of malignant spindle cells, increase mitosis seen up in most areas up to 18/10 high power field, focal necrosis areas and Immunohistochemical staining show positivity for desmin, H-caldesmon and negative for CD 117 and DOG1. All of 12 lymph nodes retrieved shows reactive changes and no evidence of nodal metastasis. The features are suggestive of leiomyosarcoma of sigmoid colon, early stage pT1 organ confined malignant tumour with complete resection at all surgical margins. He did not require adjuvant therapy and proceeded with surveillance follow up shown no recurrence.

Discussion

Leiomyosarcoma of colon is a rare colorectal malignancy. The incidence of leiomyosarcoma is less than 0.1% of all colorectal malignancy. Only minority of cases reported over the century and remain no standard guideline available in the treatment due

to its rare entity. Most of the cases encounter a difficulty in preoperative confirmative diagnosis, surgical approach and adjuvant therapy.

Wang Y et al conducted a pool analysis in 2020 found that only 41 cases of colonic leiomyosarcoma report over a century. Gender has equivalent predisposing risk with mean age reported at 58 years. Majority presented with symptoms of pain 36%, abdominal mass 12% and intestinal occlusion in only 4.9%. Morphology with polypoidal lesion reported in 34.8% and one fifth with intramural lesion. A systemic review by Alfredo and colleague also mention that majority are asymptomatic. Only 29% of cases presented with obstructive symptoms. The morphological feature reported as polypoidal in 58% follow by intramural and ulceration. Leiomyosarcoma rarely presented as circumferential lesion.

Our case had a rare presentation of leiomyosarcoma with long segment circumferential obstructive intramural lesion on imaging which mimic common colorectal adenocarcinoma malignancy. The age group and constitutional symptoms also point towards the more common adenocarcinoma malignancy group. The specimen visualizes a long segment yellow pale intramural circumferential lesion with a normal mucosal layer which is a rare occasion.

Preoperative confirmative diagnosis is difficult due to the difficult differentiations with Gastrointestinal stromal tumor and benign leiomyoma.

Devriendt and colleagues reported a case in 2019 with similar obstructive presentation encounter an inconclusive biopsy result. Only post-surgery pathology able to determine the nature or tumor.

A case report by Otto et al suggested a Contrasted computer topography with MRI could assist in diagnosis. Leiomyosarcoma tumor display better density on T2-weighted imaging and heterogenous lesion with necrosis in computerized topography.

As describe by Nassif et al in their systemic review in 2019, the therapeutic dilemma is a major problem as Leiomyosarcoma carries a worse prognosis as compare to common gastrointestinal stromal tumor. Leiomyosarcoma immunohistology staining will demonstrate negative marker for KIT, CD 34 and DOG-1 whereas GIST marked positive result. The determination between the benign nature leiomyoma with malignancy leiomyosarcoma is determine by evidence of cellular atypia with high mitotic activity in high power field pathology. The diagnostic procedure as mentioned can only be obtain with a complete resection specimen. Annicchiarico et al in the recent 2024 article also describe similar endoscopic and histology diagnostic dilemma in their systemic review. 35% of cases underwent surgery with no confirmative preoperative histology. Most of the cases reviewed required sort of emergency setting intervention which add further difficulty for more detailed elective workup.

This deems difficulty in our case as presentation in emergency setting and intramural lesion. We successfully convert his surgery into a three-stage surgery and prevented a Hartmann's procedure. Furthermore, we obtain the opportunity for endoscopic assessment and optimisation. Despite repeated endoscopic and biopsy remain inconclusive due to its deeply located intramural prevent a confirmative sample. The inconclusiveness of nature of tumor also limited the option of neoadjuvant therapy.

Currently there are no protocol guideline and consensus for its treatment. Surgical resection remains the curative option for leiomyosarcoma. Yun Wan et al analysis showed cases with age less than 60 and tumor less than 8 cm size has better in prognosis. The survival is associated with the extensa of mitotic changes and completeness of R0 resection. They demonstrated a 25 months longer overall survival. Nassif et al and Annicchiarico et al in their recent studies has proven the lower rate of 20% with radical resection. Unresectable tumor carries poor prognosis with liver and lung metastasis. Nodal metastasis is rare in leiomyosarcoma. However, Beauchamp et al reported a case with mesenteric lymph nodes metastasis. Tago et al from Japan reported a case with malignant transformation from leiomyoma to malignant leiomyosarcoma after 3 years of diagnosis. The transformation accompanies with nodal metastasis at regional drainage even with the tumor was confined only in the proper muscular layer. The pathophysiology distant metastasis for leiomyosarcoma remain incomprehensive due to the rarity of cases.

Our patient had an acceptable performance status and comorbid however with the recent pulmonary embolism stratified him under the high-risk surgical candidate. The severe respiratory illness could have been contributed by infection and the intestinal obstruction. Majority of authority would have opted for a palliative care for his disease with predictable short survival duration. Nevertheless, we successfully prepared our patient for a curative definitive surgery after a well-organized multidisciplinary perioperative care. Our patient final histology reveals an early-stage tumor with organ confined and no nodal involvement. He successfully underwent a R0 radical resection. Exception of his age factor, shown to have a curative good prognosis outcome.

Conclusion

Leiomyosarcoma is a rare colorectal malignancy with unpredictable presentation and preoperative diagnostic dilemma. Well, planned staged surgery for emergency emergencies setting with Multidisciplinary team work perioperative optimization demonstrated better surgical outcome.

Acknowledgement: Appreciation to Dato Dr Wan Khamizah and Dr Nil-Amri Mohamed Kamil for the advice in the management process.

Conflict of interest: There is no conflict of interest to declared.

Reference

1. Nassif MO, Habib RA, Almarzouki LZ, Trabulsi NH. Systematic review of anorectal leiomyosarcoma: Current challenges and recent advances. World J Gastrointestinal Surgery 2019;11(8): 334-341. DOI: https://dx.doi.org/10.4240/wjgs.v11.i8.334
2. Wang Y, Wang H, Yuan ZL, Zhao JF, Dong DB, Gao Q, A pooled analysis of risk factors of surgically treated leiomyosarcoma of the colon in adults. World Journal of Surgical Oncology (2020) 18:61.
3. Annicchiarico A, Montali F, Baldinu M, Casali L, Virgilio E, Costi R. Leiomyosarcoma of the rectum: A systematic review of recent literature. Journal of Surgical Oncology 2024; 129:365–380.

4. Devriendt S, Leman G, Vanrykel F. (2019): Primary leiomyosarcoma of the colon: a case report and review of the literature, Acta Chirurgica Belgica. DOI: 10.1080/00015458.2019.1589185
5. Otto P.O, Loft M.K, Grimm P, Rafaelsen S.R, Pedersen M.R.V. Ultrasound and contrast enhanced CT imaging of a colon mesentery leiomyosarcoma. European Journal of Radiology Open 8 (2021) 100376. https://doi.org/10.1016/j.ejro.2021.100376.
6. Parmar V, Mehta B, Shah S, Yadav R, Rao M, Primary Leiomyosarcoma Of Sigmoid Colon and Rectum-Report of Two Cases. International Journal of Medical Reviews and Case Reports (2021) 5(1):36-38.
7. Korkmaz C, Zamani A, Dogan P, Esen H.H. Colon Leiomyosarcoma Diagnosed by Pleural Biopsy: A Case Study Pakistan Journal of Medical Research, 2021 (July-September) Vol. 60, No. 3, 2021.
8. Avendano TLM, Flores JAC. Leiomyosarcoma of the rectosigmoid junction, a case report. Radiology Case Reports 15(2020)1887-1890.
9. Tago T, Suzuki S, Kuroda J, Udou R, Nishida K, Oshiro Y, Shimazaki J, Kou K, Morishita Y, Shimoda M. Leiomyosarcoma of the transverse colon with lymph node metastasis and malignant transformation: a case report. surg case rep (2020) 6:256. doi.org/10.1186/s40792-020-00998-4.
10. Beauchamp A, Hajjar R, Khullar S, Latour M, Schwenter F, Sebajang H. Mesenteric Lymph Node Recurrence of a Primary Colorectal Leiomyosarcoma. Case Reports in Surgery Volume 2020, Article ID 6935834, 5 pages. https://doi.org/10.1155/2020/6935834.