Sarcomatoid Carcinoma of the Ascending Colon: A Case Report and Literature Review

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Patient: Female, 77-year-old
Final Diagnosis: Sarcomatoid carcinoma
Symptoms: Abdominal pain • abdominal distension • fever • nausea
Medication: —
Clinical Procedure: R2 right hemicolectomy, stapled ileo-colostomy, and partial omentectomy
Specialty: Surgery

Objective: Rare disease
Background: Sarcomatoid carcinoma is a rare tumor that can occur in different organs and anatomical locations. Colonic sarcomatoid carcinoma, also known as carcinosarcoma, is an extremely rare tumor, with only 32 cases reported world-wide. The pathogenesis and guidelines for treatment are poorly understood due to the rarity and invasiveness of the disease.

Case Report: A 77-year-old woman presented with worsening lower abdominal pain and associated fever after having initially been diagnosed with stump appendicitis and associated phlegmon 3 weeks prior, which was treated with antibiotics. Repeat imaging revealed an extraluminal versus perforated colonic mass with associated phlegmon. The patient’s condition continued to worsen, with development of obstructive-like symptoms, resulting in operative intervention involving a R2 right hemicolectomy, stapled ileo-colostomy, and partial omentectomy. The patient had an uneventful remainder of her hospitalization other than continued lower abdominal pain. After initial discharge, the patient presented to an outside hospital due to continued deterioration of health, with findings of an additional mass, likely secondary to the previous lymphadenopathy. Ultimately, goals of care were discussed, and the decision was made to provide palliative care, and the patient died due to her illness 32 days after the initial procedure.

Conclusions: Carcinosarcoma is an extremely rare tumor with scant research guiding treatment guidelines. Current guidelines gathered from previous case reports suggest treating colorectal carcinosarcoma as adenocarcinoma. Additional research and studies are needed to establish appropriate therapeutic guidelines for carcinosarcoma.

Keywords: Carcinosarcoma • Colorectal Neoplasms • Colorectal Surgery • General Surgery

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Background

Sarcomatoid carcinoma, also referred to as carcinosarcoma, is a rare tumor that exhibits 2 distinct tissue origins. The biphasic tumor has properties composed of epithelial and mesenchymal origin. The epithelial component (carcinoma) is composed of high-grade or mid-grade adenocarcinoma [1]. The mesenchymal component (sarcoma) may or may not be differentiated [1]. These tumors can occur in different anatomic locations or different origins, with the first reported case in 1864 by Virchow [2]. Over the course of time, research and clinical evidence has shown a predilection for the female genital tract [3,4] along with the head and neck [5]. Isolated reports of intra-abdominal carcinosarcoma have seen higher reports in the stomach [6] and esophagus [7] compared to the rest of the gastrointestinal tract. Based upon our review, colorectal carcinosarcoma is extremely rare, with only 32 cases reported in the literature. Herein, we present a case of ascending colon carcinosarcoma treated with a R2 en-block right hemicolectomy, which is resection of all possible gross disease with known residual gross disease that was not resected.

Case Report

The patient was a 77-year-old woman with noted history of type 2 diabetes mellitus and hypertension, who initially presented to the hospital due to lower abdominal pain that had been present for approximately 1-1.5 weeks prior to presentation. The pain was worse when she was supine but improved iwhen walking. She reported associated nausea but denied any episodes of emesis. She underwent imaging (Figure 1) with radiographic interruption for stump appendicitis and extensive peri-focal phlegmon. Initially, she was managed with antibiotics and pain medication with discharge home when symptoms resolved. The patient returned to the hospital 18 days later due to continued worsening abdominal pain with associated fever. She had repeat imaging (Figure 2) performed that was concerning for extraluminal versus perforated colonic mass with associated phlegmon. The patient was admitted for medical therapy treatment with antibiotics and pain control, and was initially evaluated by Gastroenterology for possible colonoscopy to obtain a tissue specimen, which was deferred due to risk of perforation or further injury because of the procedure.
Interventional Radiology evaluated and obtained a tissue biopsy along with fluid aspiration. Further imaging was obtained (Figure 3) due to the patient developing obstructive-like symptoms on hospital day 6. The patient was taken to the operating room for an exploratory laparotomy with an R2 (resection of disease with known disease not resected) right hemicolectomy, stapled ileo-colostomy, and partial omentectomy.

Intraoperatively, we found an approximately 7-cm mass that was extraluminal in nature, causing compression on the ascending colon just distal to the cecum. The mass was associated with significant lymphadenopathy involving the mesentery down to the root along the superior mesenteric artery and to the duodenum, in addition to dense lymphadenopathy and fibrosis in the retroperitoneum. The liver appeared grossly normal. Staples were placed in the resection bed for possible need of radiation in the future. The patient had a predominately uncomplicated hospital course with an extended stay for a total of 15 days following the procedure. During that time, she had re-occurrence of lower abdomen pain, which was further evaluated with additional imaging (Figure 4) that showed extensive lymphadenopathy, which was expected due to the inability to resect all diseased tissue. On the day of discharge, she was tolerating diet without complications and was having regular bowel and bladder function. The patient was awaiting pathology results with scheduled appointment for oncology evaluation. She presented to an outside hospital 4 days after discharge from our institution due to persistent lower abdominal pain with associated poor PO intake. She was subsequently found to have an approximately 5-cm necrotic appearing mass with surrounding mesenteric lymphadenopathy, which was likely a part of the known disease not resected at the index procedure, in addition to a 2-cm rim-enhanced fluid collection. While at the outside hospital, the family decided on palliative care due to the patient’s rapidly declining condition. The patient died 32 days after her initial procedure.

**Macroscopic Description**

During the procedure, the mass was approximately 7 cm in diameter, involving the ascending colon to the hepatic flexure, with a gross distal margin of 7 cm and a proximal margin of
10 cm. There was no identified involvement of the cecum. The mass was densely adhered to the abdominal sidewall, mesentery, omentum, and duodenum. An area of contained perforation was noted into the mesentery, without feculent or purulent peritonitis. Significant lymphadenopathy was identified in the mesentery down to the root along the superior mesentery artery.

**Microscopic Description**

On pathology examination, the specimen was noted to be a biphasic tumor with identified areas of single and multi-nucleated cells with pleomorphic nuclei admixed with spindle cells containing anaplastic nuclei (Figure 5) in addition to areas of pleomorphic nuclei, prominent nucleoli, and abundant mitotic figures (Figure 6). Additional immunohistochemistry performed showed reactive tumor cells to calretinin (Figure 7) but other markers for mesothelioma were negative. Additional tumor cells were positive for claudin-4. The specimen was noted to have extensive area of necrosis measuring 5.7 cm, with extension of the tumor to the peri-colonic fat. Nine lymph nodes were positive malignancy out of 19 (9/19).

**Final Diagnosis**

The final diagnosis was stage IIIC T4bN2bM0 sarcomatoid carcinoma of the ascending colon involving extension into the mesenteric root and retroperitoneum with negative margins and 9/19 lymph nodes positive for malignancy.

**Discussion**

Carcinosarcoma of the colon is an extremely rare tumor. The first reported case, by Weidner and Zekan [8], was in 1986; it was located in the sigmoid colon and treated with a left hemicolectomy. Since 1986, a total of 32 cases have been reported (Table 1) [1,8-38]. A review of the patient characteristics reported in the literature (Table 2) revealed a mean age of presentation of 67 years old, with 59% of cases in females. The most common chief concern on presentation remains bright red blood per rectum, while abdominal pain is the second most common. The rectum is the site of highest occurrence in the colon, with our case as the sixth reported episode in the ascending colon.

Sarcomatoid carcinoma has had numerous other aliases throughout literature. In addition to sarcomatoid carcinoma, it has been referred to as carcinoma with mesenchymal stroma, carcinoma with sarcomatous change, spindle cell carcinoma, and pleomorphic anaplastic carcinoma [38]. The term carcinosarcoma was officially coined by the World Health Organization as the classification class [16].

Current knowledge is lacking in understanding the development of the tumor. Current theory favors a metaplastic origin. It is believed that the carcinoma component differentiates into the sarcomatoid component during the development of cancer cells. Other theories have documented possible infection or a p53 gene resulting in conversion of the carcinoma to sarcoma [10,11].

Sarcomatoid carcinoma has many of the same presenting features as traditional colonic adenocarcinoma. Chief concerns on presentation range from rectal bleed [1,17,34] to abdominal pain [29], making it difficult to differentiate without...
| No. | Author                        | Age/Sex | Site        | Treatment | Lymph Node Involvement | Survival               |
|-----|-------------------------------|---------|-------------|-----------|------------------------|------------------------|
| 1   | Weidner and Zekan [8]         | 73/M    | Sigmoid     | LHC       | Negative               | 48 months              |
| 2   | Staroz et al [9]              | 64/M    | Descending  | LHC       | Positive               | 4 months               |
| 3   | Roncaroli et al [10]          | 71/F    | Rectum      | LAR       | Positive               | 6 months               |
| 4   | Isimbaldi et al [11]          | 86/F    | Ascending   | RHC       | Negative               | Alive at 24 month follow up |
| 5   | Gentile and Castellanta [12]  | 40/F    | Cecum       | RHC       |                        | 2 months               |
| 6   | Bertram et al [13]            | 79/F    | Ascending   | RHC       | Positive               | 5 months               |
| 7   | Serio and Aguzzi [14]         | 69/F    | Descending  | LHC       | Negative               | Alive at 6 month follow up |
| 8   | Shoji et al [15]              | 78/M    | Descending  | LHC       | Negative               | Alive at 16 month follow up |
| 9   | Nakao et al [16]              | 68/F    | Transverse  | RHC       | Positive               | Alive at 14 month follow up |
| 10  | Takeyoshi et al [17]          | 82/M    | Rectum      | LAR       | Positive               | 6 months               |
| 11  | Shah et al [18]               | 57/F    | Rectosigmoid| LAR       | Positive               | 5 months               |
| 12  | Di Vizio et al [19]           | 56/F    | Descending  | LHC       | Positive               | 21 months              |
| 13  | Kim et al [20]                | 41/F    | Sigmoid     | LAR       | Positive               | 4 months               |
| 14  | Armendi et al [21]            | 84/M    | Descending  | LHC       | Negative               | 4 days                 |
| 15  | Ishida et al [22]             | 80/F    | Hartmann + RHC |          | Positive               | 6 months               |
| 16  | Macaigne et al [23]           | 67/F    | Rectum      | LHC       |                        | 2 months               |
| 17  | Ekinci et al [24]             | 39/F    | Rectosigmoid| LAR       | Positive               | Alive at 4 month follow up |
| 18  | Kim et al [25]                | 71/M    | Ascending   | Unresectable | Lost to follow up     |
| 19  | Ambrosini-Spaltro et al [26]  | 81/M    | Ascending   | RHC       | Positive               | Alive at 24 month follow up |
| 20  | Tsekouras et al [27]          | 60/M    | Rectum      | APR       | Positive               | 6 months               |
| 21  | Öztürk et al [28]             | 65/F    | Rectum      | APR       | Negative               | Alive at 60 month follow up |
| 22  | Lee et al [1]                 | 52/F    | Rectum      | APR       | Negative               | Alive at 8 month follow up |
| 23  | Jeong et al [29]              | 13/F    | Rectosigmoid| LAR       | Positive               | Alive at 2 month follow up |
| 24  | Patel et al [30]              | 43/F    | Sigmoid     | Hartmann  |                        | Alive at 2 month follow up |
| 25  | Shim et al [31]               | 65/M    | Ascending   | RHC       | Positive               | 1 month                |
| 26  | Mori et al [32]               | 65/M    | Sigmoid     | LAR       | Negative               | 10 months              |
| 27  | Ryu et al [33]                | 72/F    | Cecum       | RHC       | Negative               | Alive at 20 month follow up |
| 28  | Kolodziejzak et al [34]       | 83/M    | Rectum      | Unresectable |                        | 1 month                |
Table 1 continued. Colorectal carcinosarcoma cases in current literature.

| No. | Author                  | Age/Sex | Site        | Treatment | Lymph Node Involvement | Survival  |
|-----|-------------------------|---------|-------------|-----------|------------------------|-----------|
| 29  | Peris Tomas et al [35]  | 59/?    | Sigmoid     | Hartmann  |                        | 6 months  |
| 30  | Sudlow et al [36]       | 80/F    | Low rectum  | APR       | Negative               | 25 months |
| 31  | Osholowu et al [37]     | 68/M    | Rectum      | Unresectable |                        | Positive  |
| 32  | Saad et al [38]         | 67/M    | Cecum       | RHC       | Positive               | Lost to follow up |
| 33  | Our Case                | 77/F    | Ascending   | RHC       | Positive               | 32 days   |

LHC – left hemicolectomy; RHC – right hemicolectomy; LAR – low anterior resection; APR – abdomino-perineal resection.

Table 2. Characteristics of current literature for colonic carcinosarcoma.

| Total cases in literature, n | 33 |
|-----------------------------|----|
| Mean age of presentation    | 67 (13-86) |
| Gender, n (%)               |    |
| Male                        | 13 (41) |
| Female                      | 19 (59) |
| Primary presentation        |    |
| Large bowel obstruction     | 2  |
| Melena                      | 2  |
| Fresh blood per rectum      | 11 |
| Abdominal pain              | 10 |
| Diarrhea                    | 3  |
| Site, n (%)                 |    |
| Cecum                       | 3 (9) |
| Ascending                   | 6 (18) |
| Transverse colon            | 1 (3) |
| Descending                  | 5 (15) |
| Sigmoid                     | 5 (15) |
| Recto-sigmoid               | 4 (12) |
| Lymph node status           |    |
| Positive                    | 16  |
| Negative                    | 9   |

tissue-proven diagnosis. Given the current literature and lack of overall treatment direction, many of these patients are being treated as cases of general colonic cancer. The major difference between sarcomatoid carcinoma and colonic adenocarcinoma is the aggressive nature with poor prognosis of the tumor. As seen in our patient and numerous other patients in previous cases, the overall length of survival is short and aggressive therapy is needed to improve patient outcomes. Current therapy involves 5-fluorouracil, leucovorin, doxorubicin, and cisplatin, with additional radiation to treat patients in an adjuvant manner, but no evidence has shown their overall effect on the patient [16,20,28]. Additional research is necessary to determine appropriate chemotherapy regimens and prognostic factors. In addition, it has been noted that the metastasis of these tumors follows along the carcinoma origin, with spread to distant sites or lymph nodes rather than hematogenous spread as with sarcomas.

The rarity of sarcomatoid carcinoma leads to a difficult challenge when treating a newly diagnosed patient. Additional research needs to be performed on determination of optimal prognostic factors, role of adjuvant therapy, and appropriate surgical planning procedures.

In this current setting, the most appropriate management involves treating the patient along a colonic adenocarcinoma algorithm. In the end, due to the severity of the disease and rapid spread, it may be difficult to achieve appropriate studies to develop definitive treatment algorithms.

Conclusions

Carcinosarcoma is an extremely rare tumor, with little research guiding treatment guidelines. It has been identified as a biphasic tumor displaying origins from the epithelium and mesenchyme. Immunohistochemistry remains the standard for diagnosis. Current guidelines gathered from previous case reports suggest the treatment of choice for colorectal carcinosarcoma is the same as for adenocarcinoma. Additional research and prospective studies are needed to establish appropriate therapeutic guidelines and prognostic factors for carcinosarcoma.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.
References:

1. Lee JK, Ghosh P, McWhorter V, Payneau M, et al. Evidence for colorectal sarcomatoid carcinoma arising from tubulovillous adenoma. World J Gastroenterol. 2008;14(27):4389-94

2. Vischow R. Die krankhafte Geschwulste. Berlin: Hirschwald; 1864; 170-384 [in German]

3. Balocchi G, Kanavagh JJ, Wharton JT. Endometrioid stromal sarcomas arising from ovarian and extranovarian endometriosis. Report of two cases and literature review. Gynecol Oncol. 1990;36:147-51

4. Norris H, Taylor HB. Mesenchymal tumors of the uterus: III. A clinical and pathological study of 31 carcinosarcomas. Cancer. 1966;19:1459-65

5. Berthelet E, Shenouda G, Black MJ, et al. Sarcomatoid carcinoma of the head and neck. Am J Surg. 1994;168(5):455-58

6. Tsuneyama K, Sasaki M, Sabit A, et al. A case report of gastric carcinosarcoma with rhabdomyosarcomatous and neuroendocinial differentiation. Pathol Pract. 1999;195(2):93-98

7. Madan AK, Long AE, Weldon CB, Jaffe BM. Esophageal carcinosarcoma. J Gastrointest Surg. 2001;5(4):414-47

8. Weidner N, Zekan P. Carcinosarcoma of the colon. Report of a unique case with light and immunohistochemical studies. Cancer. 1986; 58(5):1126-30

9. Starko F, Botton A, Potet F. Malignant tumors of the colon with two components (carcinosarcoma): Report of a case. Ann Pathol. 1995;15:457-58

10. Roncaroli F, Montironi R, Felicetti F, et al. Sarcomatoid carcinoma of the anorectal junction with neuroendocrine and rhabdomyoblastic features. Am J Surg Pathol. 1995;19(2):217-23

11. Isimbaldi G, Sironi M, Assi A. Sarcomatoid carcinoma of the colon: Report of the second case with immunohistochemical study. Pathol Pract Res. 1996;192(5):483-87

12. Gentile R, Castellana E. Carcinoma carcinosarcoma of the colon, or two tumors? Pathologica. 1997;89:62-68

13. Bertram P, Treutner KH, Tietze L, Schumpelick V. True carcinosarcoma of the colon. Report of a case and review of the literature. J Gastroenterol. 1998;33(7):545-49

14. Shoji M, Dobashi Y, Iwabuch K, et al. Sarcomatoid carcinoma of the descending colon: A histological, immunohistochemical and ultrastructural analysis. Acta Oncol. 1998;37(7-8):765-68

15. Nakao A, Sakagami K, Uda M, et al. Carcinoma carcinosarcoma of the colon: Report of a case and review of the literature. J Gastroenterol. 1998;33(2):276-79

16. Takeyoshi I, Yoshida M, Ohwada S, et al. Skin metastasis from the spindle cell component in rectal carcinoma. Hepatogastroenterology. 2000;47(36):1611-14

17. Shah S, Kim DH, Harster G, Hossain A. Carcinoma carcinosarcoma of the colon and spleen: A fleshy purple mass on colonoscopy. Dig Dis Sci. 2001;46(1):106-8

18. Di Vizio D, Insabato L, Conzo G, et al. Sarcomatoid carcinoma of the colon: A case report with literature review. Tumori. 2001;87(6):431-35

19. Kim JH, Moon WS, Kang MJ, et al. Sarcomatoid carcinoma of the colon: A case report. J Korean Med Sci. 2001;16(5):657-60

20. Aramendia T, Fernandez-Acenero MJ, Villanueva MC. Carcinosarcoma of the colon: Report of a rare tumor. Pathol Res Pract. 2003;199(5):345-48

21. Ishida H, Ohawa T, Nakada H, et al. Carcinoma carcinosarcoma of the rectosigmoid colon: Report of a case. Surg Today. 2003;33(7):545-49

22. Macaigne G, Aouad K, Boivin JF, et al. Sarcomatoid carcinoma of the colon: Report of a case and review of the literature. Gastroenterol Clin Biol. 2004;28:600-4

23. Ekinci N, Ergun SA, Kar H. Sarcomatoid carcinoma of the colon. Turk J Cancer. 2005;35:138-40

24. Kim N, Luchs JS, Halpern D, et al. Radiology-pathology conference: Carcinosarcoma of the colon. Clin Imaging. 2005;29(4):259-62

25. Ambrosini-Spaltro A, Vaira V, Braidotti P, et al. Carcinosarcoma of the colon: Report of a case with morphological, ultrastructural, and molecular analysis. BMC Cancer. 2006;6:185

26. Tsekouras DK, Katsaragakis S, Theodorou D, et al. Rectal carcinosarcoma: A case report and review of literature. World J Gastroenterol. 2006;12(9):1481-84

27. Ozturk E, Yilmazlar T, Yeri C. A rare tumor located in the anorectal junction: Sarcomatoid carcinoma. Turk J Gastroenterol. 2006;17:236-39

28. Jeong YJ, Lee MR, Kim JC, et al. Carcinoma carcinosarcoma of the rectosigmoid colon in a 13 year old girl. Pathol Int. 2008;58(7):445-50

29. Patel DH, Dang S, Bentley FR, et al. Carcinosarcoma of the colon: A rare cause of colovesical fistula. Am Surg. 2009;75(4):335-37

30. Shin H, Hong YK, Kim SS, et al. Carcinoma carcinosarcoma on ascending colon found by bowel perforation: A case report. J Korean Soc Coloproctol. 2010;26(5):368-72

31. Mori Y, Katsumata K, Suzuki S, et al. Carcinoma carcinosarcoma of the sigmoid colon: Report of a case. Case Rep Gastroenterol. 2010;4(3):484-91

32. Ryu Y, Kim A, Kim H, et al. Carcinomasarcoma in the cecum. Gut Liver. 2012;6(3):395-98

33. Kolodziejczak M, Bielecki K, Sudol-Szopinska I, et al. Very rare case of rectal carcinosarcoma. Tech Coloproctol. 2013;17(5):615-17

34. Peris Tomas N, Lozano AG, et al. Very rare case of rectal carcinosarcoma: Report of a case with morphological, ultrastructural, and molecular analysis. BMC Cancer. 2006;6:185

35. Ambrosini-Spaltro A, Vaira V, Braidotti P, et al. Carcinosarcoma of the colon: Report of a case with morphological, ultrastructural, and molecular analysis. BMC Cancer. 2006;6:185

36. Tsekouras DK, Katsaragakis S, Theodorou D, et al. Rectal carcinosarcoma: A case report and review of literature. World J Gastroenterol. 2006;12(9):1481-84

37. Ozturk E, Yilmazlar T, Yeri C. A rare tumor located in the anorectal junction: Sarcomatoid carcinoma. Turk J Gastroenterol. 2006;17:236-39

38. Jeong YJ, Lee MR, Kim JC, et al. Carcinoma carcinosarcoma of the rectosigmoid colon in a 13 year old girl. Pathol Int. 2008;58(7):445-50

39. Patel DH, Dang S, Bentley FR, et al. Carcinosarcoma of the colon: A rare cause of colovesical fistula. Am Surg. 2009;75(4):335-37

40. Shin H, Hong YK, Kim SS, et al. Carcinoma carcinosarcoma on ascending colon found by bowel perforation: A case report. J Korean Soc Coloproctol. 2010;26(5):368-72

41. Mori Y, Katsumata K, Suzuki S, et al. Carcinoma carcinosarcoma of the sigmoid colon: Report of a case. Case Rep Gastroenterol. 2010;4(3):484-91

42. Ryu Y, Kim A, Kim H, et al. Carcinomasarcoma in the cecum. Gut Liver. 2012;6(3):395-98

43. Kolodziejczak M, Bielecki K, Sudol-Szopinska I, et al. Very rare case of rectal carcinosarcoma. Tech Coloproctol. 2013;17(5):615-17

44. Peris Tomas N, Lozano AG, et al. Carcinoma carcinosarcoma of the colon: A report of a case with morphological, ultrastructural, and molecular analysis. Pathol Res Pract. 2003;249(10):1481-84

45. Ambrosini-Spaltro A, Vaira V, Braidotti P, et al. Carcinosarcoma of the colon: Report of a case with morphological, ultrastructural, and molecular analysis. BMC Cancer. 2006;6:185

46. Tsekouras DK, Katsaragakis S, Theodorou D, et al. Rectal carcinosarcoma: A case report and review of literature. World J Gastroenterol. 2006;12(9):1481-84

47. Ozturk E, Yilmazlar T, Yeri C. A rare tumor located in the anorectal junction: Sarcomatoid carcinoma. Turk J Gastroenterol. 2006;17:236-39