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Case Report

Surgery Section

Case Report: Collision Tumour of Colon Leiomyosarcoma and Adenocarcinoma

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ABSTRACT

The colon is an exceedingly rare site of primary leiomyosarcoma and only a few cases have been published to date. Of the reported cases of collision tumours, collision tumours that specifically occurred in the colon have consisted of combinations of adenoma or adenocarcinoma with lymphomas or neuroendocrine tumours. Here, not only do we report a case of colon leiomyosarcoma, but we report, what is to our knowledge, the first case of collision tumour consisting of colon leiomyosarcoma and adenocarcinoma. Cause, prognosis, and treatment of colon collision tumours vary and are yet to be understood.

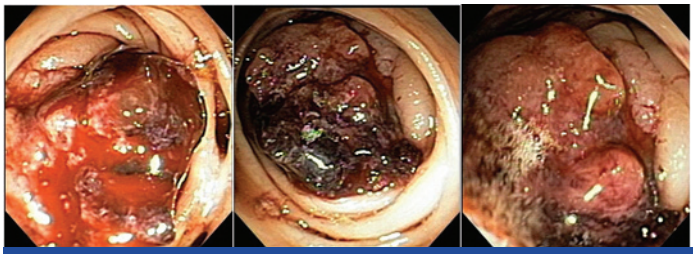
Keywords: Carcinoid, Colonic neoplasms, Independent tumours

CASE REPORT

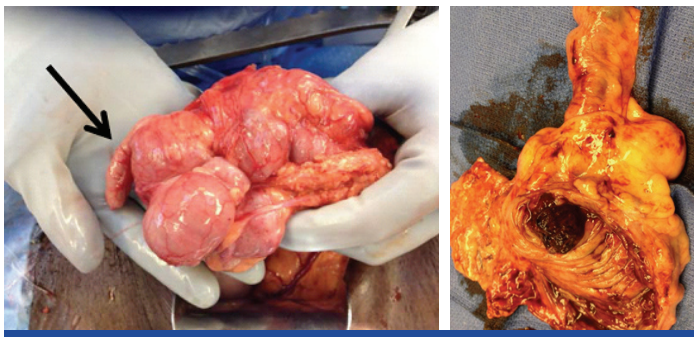
An 82-year-old man with a history of left sigmoid colectomy in 2005 for a malignant sigmoid polyp presented to our Emergency Department in June 2013 after five days of painless, melenic stools. The patient had a history of smoking 1 pack/week for 50 years but had quit in 1981. The polyp in 2005 had been an infiltrating well-differentiated adenocarcinoma arising in tubulovillous adenoma. The surgical pathology specimen from the sigmoid colectomy had been negative for carcinoma or polyps and all lymph nodes were considered benign in nature. One year prior to his presentation to us, he had been admitted to an outside institution with a complaint of three days of melena. At that time upper endoscopy (EGD) was unrevealing, and colonoscopy revealed diverticulosis of the right colon. The source of bleed was found as two nodular polypoid lesions in the proximal small bowel without evidence for malignancy. On presentation to our institution, labwork revealed a CEA level of 3.0 ng/ml (preoperative CEA in 2005 was 1.9 ng/ml) and an admitting haemoglobin level of 9.0 g/dL. Upper endoscopy (EGD) and colonoscopy were performed. EGD was unremarkable, but colonoscopy revealed a nearly obstructing 5 cm friable mass at the ileocecal valve and extending distally [Table/Fig-1]. Histopathology report of the biopsy demonstrated leiomyosarcoma, Grade 2, desmin+, KIT- . Fluorodeoxyglucose (FDG) PET/CT scan of chest, abdomen, and pelvis were obtained and demonstrated an intensely FDG-avid cecal lesion consistent with the known leiomyosarcoma as well as a 0.9 cm right upper lung lobe opacity and hilar nodes with FDG uptake concerning for metastatic disease. Despite the possible metastasis, the decision was made to proceed with surgery due to a near complete obstruction seen on colonoscopy and anemia requiring blood transfusions from the mass (gradual haemoglobin decrease of > 1 g/dL over 24 hours).

The patient underwent exploratory laparotomy and right hemicolectomy [Table/Fig-2a&b]. Histopathologic examination from the right hemicolectomy specimen demonstrated a 2.2 cm tumour composed of two distinct superimposed tumours: 1) high-grade adenocarcinoma arising from a tubular adenoma; and 2) high-grade (Grade 2) leiomyosarcoma. The tumours were intimately admixed but retained their distinct morphologies, a so called "collision tumour". The adenocarcinoma consisted of infiltrating epithelial cells arranged in glands or solid nests with marked nuclear pleomorphism, increased nuclear to cytoplasmic ratio, and nuclear hyperchromasia. The leiomyosarcoma was composed of malignant spindle cells arranged in broad fascicles with elongated, blunt-ended nuclei, nuclear pleomorphism, nuclear hyperchromasia, and numerous mitotic figures (up to 20 per high power field). Immunohistochemically, the malignant spindle cells stained positive for desminand negative for CD117 [Table/Fig-3a-c]. All 12 lymph nodes in the specimen were negative for tumour.

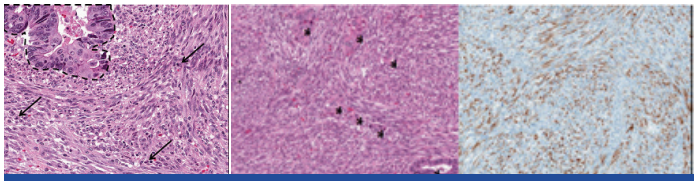
Our patient's tumour's AJCC classification was T4aN0M0. His postoperative course was unremarkable except for a persistent oxygen requirement due to postoperative atelectasis and volume



[Table/Fig-1]: Endoscopic images. Colonoscopy revealed a 5 cm friable mass immediately at the ileocecal valve and extending distally.



[Table/Fig-2a,b]: Intraoperative Images. a) View of the external wall of the cecum. Appendix is visualized on the cecum, as indicated by black arrow. b) View of the tumour in gross specimen.



[Table/Fig-3a-c]: Pathology Images. a) Collision tumour: adenocarcinoma and leiomyosarcoma. Note the intimate association between the cytologically malignant epithelial cells (dotted line) and mesenchymal cells. Cellular pleomorphism is prominent in both cell populations; additionally, the spindle cell proliferation demonstrates numerous mitotic figures (arrows). 20X magnification. b&c b) Collision tumour: adenocarcinoma and leiomyosarcoma. This picture displays the intimate relationship between the two discrete cell populations, (adenocarcinoma islands marked with *) and c) the arrangement of the sarcoma cells into 90 degree intersecting fascicles, highlighted by the desmin stain (10X magnification).

overload superimposed on significant COPD parenchymal disease. He received aggressive pulmonary toilet and was discharged home 9 days after surgery without other issues. An endobronchial ultrasound-guided fine needle aspiration of the right upper lobe lesion and hilar nodes demonstrated benign tissue. Our patient was last seen in clinic 14 months after surgery and was found to be in good health. The most recent CEA level was stable at a postoperative baseline of 1.9 ng/ml.

DISCUSSION

A "collision tumour" is an uncommon co-existence of two completely distinct, independent tumours at the same site but with sharp demarcations. The pathobiology of collision tumours is yet to be understood [1]. Of the reported cases of collision tumours, collision tumours that specifically occurred in the colon have consisted of combinations of adenoma or adenocarcinoma with lymphomas or carcinoid (and related neuroendocrine) tumours [2]. To our knowledge, this is the first reported or known case in the literature of a collision tumour composed of colon adenocarcinoma and leiomyosarcoma.

The uniqueness of a colon adenocarcinoma-leiomyosarcoma collision tumour is likely due to colon leiomyosarcomas themselves being exceedingly rare. Since the establishment of the WHO definition of GI leiomyosarcomas in the late 1990's, there have only been 11 reported cases of colonic leiomyosarcoma versus 26 cases of small bowel leiomyosarcomas [3]. Histopathology particularly differentiates this case from other reported colon collision tumours. Neuron-Specific Enolase (NSE) positivity has been reported for colon adenocarcinoma-carcinoid collision tumours [4]. In this case, the morphology of malignant spindle cell tumours clearly characterized the component tumour. NSE staining was not performed. The differential diagnosis for malignant spindle cell tumours in the colon includes leiomyosarcoma, malignant Gastrointestinal Stromal Tumour (GIST), and Malignant Peripheral Nerve Sheath Tumour (MPNST). An MPNST is desmin negative; a GIST, CD117 positive. In this case, the component tumour was desmin positive and CD117 negative.

Because of their rarity, colon collision tumours do not have a largely, serially studied and delineated standard of treatment and prognosis. However, we can infer from the treatments for the component tumours of a given collision tumour and from the prognosis of the more aggressive or poorly differentiated of the component diseases. A study reviewing 80 cases of colorectal Glandular-Neuroendocrine Mixed Tumours (GNMT), where 58% of the cases were collision tumours (versus amphicrine or composite), concluded that both the glandular and neuroendocrine components of mixed tumours can metastasize alone, combined, or independently. It was also noted that the 2 component tumours can vary in differentiation, in which case the poorly differentiated component is more likely to metastasize and affect prognosis [5].

As this is the first reported case of a collision tumour composed of colon adenocarcinoma and leiomyosarcoma, there is no series to infer treatment and prognosis. Furthermore, the more aggressive of the component tumours, colon leiomyosarcomas, are in themselves rare, poorly studied, and poorly responsive to treatment. Leiomyosarcomas as a whole are poorly responsive to chemotherapy and should a patient be diagnosed with colon leiomyosarcoma, resection remains the best therapeutic option to date. Broader studies of GIST/and gastrointestinal leiomyosarcomas and sarcomas suggest that hepatic resection of metastatic disease and chemotherapy with docetaxel and gemcitabine may prolong survival [6,7]; however, results specific to colon leiomyosarcomas, let alone colon adenocarcinoma-leiomyosarcoma collision tumours, are yet to be established. If our patient's tumour were simply stage IIB adenocarcinoma, the correlating 5-year survival would be 76.3% [8]. However, colon leiomyosarcomas appear to be aggressive, regardless of tumour size and mitotic activity. An analysis of 11 cases estimated an overall survival of 20 months after diagnosis of colon leiomyosarcoma [3]. Of the 8 patients with survival data, 7 died of the cancer. Only 2 of 11 reported patients survived 5 years. In contrast, colorectal GNMTs demonstrated a tumour-related death rate of 68% at 20 months [5].

CONCLUSION

In summary, we report a unique case of a colon adenocarcinoma-leiomyosarcoma collision tumour. Cause, prognosis, and treatment of colon collision tumours vary and are yet to be fully understood, but studies suggest that one component tumour should not be overlooked for another in determining treatment and prognosis and that the more aggressive, poorly differentiated of the components affects prognosis.

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