CARCINOMA COLON SIGNET-RING CELL TYPE: A RARE CASE REPORT

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ABSTRACT

Carcinoma colon ranks third among the commonest cancer in the world, while seen less common in the Asian countries however lately an increasing trend has been observed. Majorly the elderly population is at an increased risk of colon cancer however early onset of the disease has also been observed in past few years. Various histologic subtypes with different prognoses are present, but the most common histological type is Adenocarcinoma (1). Signet-ring cell carcinoma (SRCC) is a known rare subtype of colorectal carcinoma (< 1%) officially recognized by the WHO, mucinous types are aggressive with poor outcomes and are most commonly diagnosed around the fourth to fifth decade of life comprising only 1% of colorectal cancers. So we report a case of a 40-year-old male with adenocarcinoma colon of signet ring cell type. The patient presented with localized abdominal discomfort with only generalized weakness and fatigue which was later evaluated to be a near-obstructing right-sided colonic mass. Patient was taken up for surgery and was found to have diffuse carcinomatosis.

KEYWORDS: Colorectal cancer, Adenocarcinoma, Signet ring cell Carcinoma, Colon mass.

INTRODUCTION

The incidence of carcinoma colon in the younger age group (<50 years) has escalated at a rate of >1.5% in males and females in recent years. In young patients, CRC presents with more severe histological features. Mucin-producing adenocarcinoma of Signet-ring cell carcinoma (SRCC) type is encountered commonly in the stomach, breast, and colon in around 90% of cases (2,15). Tumours are considered as signet-cell when the tumour cell nuclei are displaced by intracellular mucin in more than 50% of cells within the tumor (3). Signet ring cell carcinoma (SRCC), a rare variant of CRC contributes less than 1% of the cases of CRC (1,4). There are various histologic types of colorectal cancer and diverse subtypes having different characteristics and prognostic relations. SRCC leads to carcinomatosis due to widespread intramural spread and aggressive behavior as the presentation is usually in the later stage which leads to negative prognostics.

CASE REPORT

A 40-years-old healthy male came to the emergency with the chief complaint of abdominal pain with fatigue and generalized weakness with a few episodes of vomiting. Radiological investigations like abdominal X-rays and other laboratory investigations which were done were found inconclusive. A diagnosis of constipation was made and he was discharged, later he returned after three weeks with complaints of increased abdominal pain, distension of the abdomen, and diarrhea. On examination, the patient had a distended and diffusely tender abdomen.

His haemoglobin level was 9 gm% and leukocyte count of 11,800 /mm^3 (with 86% neutrophil and 12% lymphocytes) other investigations like blood urea and serum creatinine levels were within normal range. The patient underwent a colonoscopic examination and a friable, ulcerated, tumour was detected in the proximal ascending colon. The patient was taken up for right hemicolectomy and the resected segment was sent for histopathological examination.

A right hemicolectomy specimen was received at the department of pathology. Specimen comprising of ascending colon along with cecum and part of transverse colon measuring 14 x4 x 3 cm was received. The cut surface of the colon showed diffuse thickening and growth measuring 2 x 1.6cm at the proximal end of ascending colon. The growth was ulcerated and friable. Representative tissue sections were taken, tissue was processed, and sections were made and stained with Haematoxylin and eosin (18). Sections were examined under the microscope and showed colonic mucosa lined by atypical cells with a high nucleocytoplasmic ratio. The underlying lamina propria and muscularis layers were infiltrated by signet ring cells, and the cells that retain

abundant intracytoplasmic mucin nuclei were pushed to one side. All the surgical cut margins were negative. No lymph vascular and perineural invasion was seen. Eight lymph nodes were identified and examined all were negative for tumor. Diagnosis signet ring cell carcinoma, poorly-differentiated type, stage T2N0 was made.

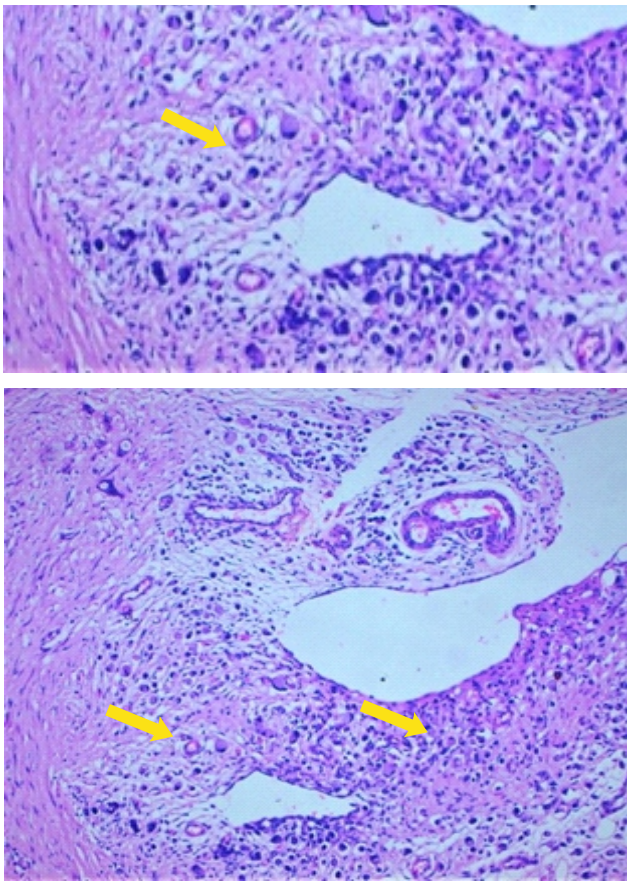
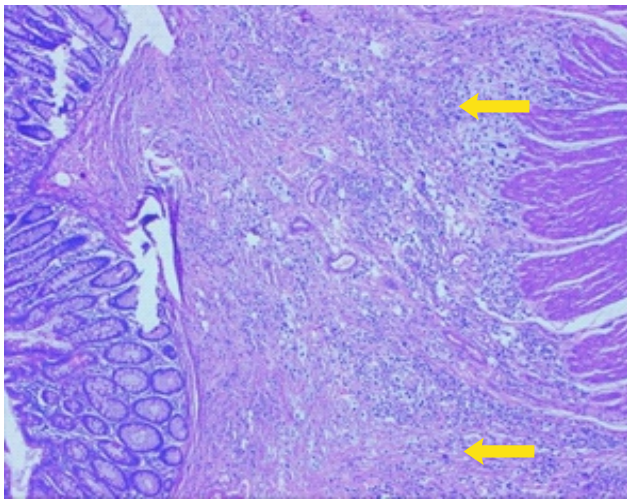


Fig. 1 a:10x H&E photomicrograph shows signet ring cells morphology (marked with an arrow) Fig. 1 b: 10x H&E photomicrograph shows signet ring cell carcinoma colon with infiltration of signet ring cells in muscle layer & serosa of colon. (Marked with an arrow head).



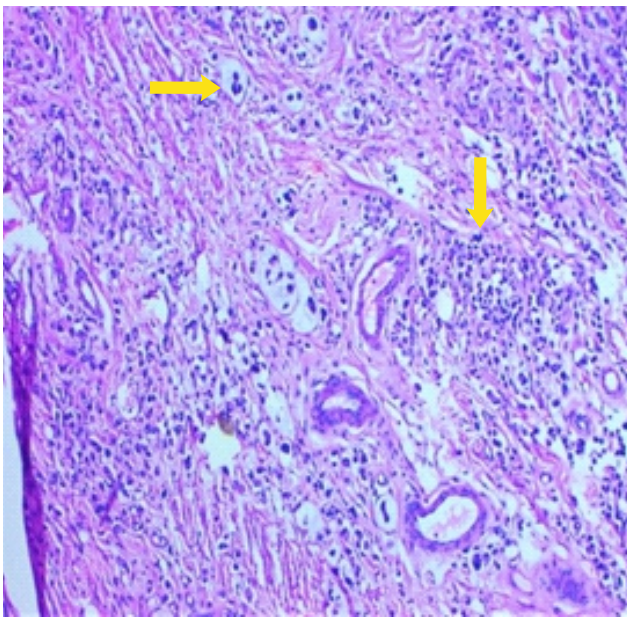


Fig. 2: (10x) & Fig. 3: 40x images: H & E Photomicrograph show >50% tumour composed of signet cells having intra cytoplasmic mucin with displacement of nucleus to one side. (marked with arrow)

DISCUSSION

CRC in the younger age group has a poor outcome due to delay in diagnosis as patients usually present with localized pain abdomen with other non-specific symptoms, uncommonness of disease, and no general screening protocols are present which contributes to delay in detection (5). SRCC arising from the stomach accounts for more than 95% of cases, and the rest are seen less commonly in organs like the pancreas, urinary bladder, colon, rectum, gallbladder, and breast (2,6). SRCC is an unusual and fatal rapidly progressing malignancy affecting the gastrointestinal lining with signet ring cells detected on microscopic examination (7-9). Primary SRCC of the colon and rectum follows a virulent progression of the disease with poor outcomes amounting to delayed diagnosis and presentation at the later stage of the disease (19). The patient usually presents with acute intestinal obstruction with minimal to no prior signs and symptoms or sometimes they may even present with perforation. Often the patient gives a history of long-standing abdominal discomfort, constipation, diarrhea, or may even present with a mass, other specific complaints of mild-moderate jaundice, weight loss, and gross ascites, are suggestive of metastatic disease (17). SRCC is differentiated from characteristic adenocarcinoma due to a large amount of mucin within the tumor and at times this mucin aggregates intracellularly in these tumors and pushes the

nucleus towards the periphery, which gives a signet-ring appearance to the cells. So it is called signet-ring cell carcinoma based on these characteristic morphologic features when present in more than 50% of the tumor. Various studies have documented an association of the disease incidence with younger age group, advanced tumor stage at the time of presentation, and distant lymph node metastasis. SRCC shows distinctive molecular patterns like high rate of microsatellite instability, and presence of chromosomal mutations like BRAF, and a low rate of KRAS mutations (10-14,17). Predominantly, poorly differentiated mucinous adenocarcinoma is more prevalent in adolescents and has a very poor outcome and this histopathological type is extremely aggressive with a tendency for metastatic spread. CRC occurring in the second to third decade is more commonly seen on the left side of the colon, only a few cases present with right colonic involvement (8). In contrast to young adults, cases above 40 years of age show a predisposition for right-side colonic involvement with a poorer prognosis, reduced 5-year survival rate, and 5-year disease-free survival rate (9). Combination chemotherapy is considered even in the most severe cases. The outcome of SRCC is often poor. Majority of patients succumb to death within the initial year of diagnosis and in most cases, the 5-year survival rate is dismal at 12% (18-20).

CONCLUSION

The case we are reporting is an extremely rare, aggressive subtype of early-onset adenocarcinoma colon signet ring cell typePalliative resection is beneficial to relieve the obstruction and it also reduces the tumor load in a few cases. Surgery is the treatment of choice to adjuvant chemotherapy followed by radiotherapy. Tests like digital rectal examination of the patient which should be followed by sigmoidoscopy and/or colonoscopy can render better results ultimately providing effective therapy. A timely diagnosis of this neoplasm can change the entire course of the disease as well as it can provide a better prognosis. Proper screening and molecular profiling will also improve patient care.

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