

# Signet Ring Cell Carcinoma of Caecum presenting as Subacute Intestinal Obstruction: A Rare Case Report

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**Abstract:** Colorectal cancer (CRC) is the third most prevalent cancer globally, with adenocarcinomas being the most frequent type. Signet ring cell carcinoma (SRCC) is a rare subtype of conventional colorectal adenocarcinoma. Most common location of SRCC is stomach, however, it can occur in other sites such as rectum, colon, lung, prostate, breast and gallbladder. We report a case of primary caecal Signet Ring Cell Carcinoma in a young male patient, presented with colicky abdominal pain and distention for one month, associated with vomiting, constipation for 5 days, significant weight loss and weakness. The clinical examination revealed abdominal tenderness and sluggish bowel sounds. Laboratory tests showed an elevated ESR at 38 mm/hr (normal value: 0-9 mm/hr). The abdominal scan showed abnormally dilated caecum and ascending colon with abnormal wall thickening. Additionally, mild free fluid was noted in abdomen and pelvis. Further histologic examination confirmed the presence of signet-ring cell carcinoma. Primary Caecal Signet Ring Cell Carcinoma is a rare entity and presents at an advanced stage with a poor prognosis.

**Keywords:** Case report, Carcinoma, Signet-ring, Colorectal, Ileo-colectomy, SRCC

## 1. Introduction

Colorectal cancer (CRC) is the third most prevalent cancer in males and the second in females, according to the World Health Organization. <sup>1</sup>Adenocarcinomas is the most frequent type of colorectal cancer. Signet ring cell carcinoma (SRCC) is one of the rare histologic variants of colorectal carcinomas, occurring in less than 1% of all CRCs. <sup>2</sup> It is associated with presentation at an earlier age, advanced tumor stage, and poor prognosis as compared to conventional adenocarcinoma. Here, we report a case of young male with primary caecal Signet Ring Cell Carcinoma, due to rarity of its occurrence and an extremely poor prognosis.

## 2. Case Presentation

A 40-year-male patient presented with complains of colicky abdominal pain and abdominal distention for one month,

associated with vomiting, constipation for 5 days, significant weight loss and weakness. The clinical examination revealed abdominal tenderness and sluggish bowel sounds. Laboratory tests showed an elevated ESR at 38 mm/hr (normal value: 0-9 mm/hr) and microcytic hypochromic anemia, haemoglobin of 9 g/dl. The abdominal scan showed abnormally dilated caecum and ascending colon with abnormal wall thickening. Additionally, mild free fluid was noted in abdomen and pelvis. Clinical diagnosis of subacute intestinal obstruction was made and patient was taken for emergency laparotomy. An exploratory laparotomy showed distention of the bowel loops of caecum and ascending colon, with a suspected thickening of the cecum. The patient underwent a right ileo-colectomy and ileocolostomy. On gross examination a circumferential growth at ileocecal junction was seen measuring 6x3 cm. The growth was 22 cm away from ileal resected end and 9 cm away from colonic resected end. Cut section showed a grey white mucoidish growth reaching up to serosa. (Fig 1 and 2)



Microscopic examination revealed primary signet ring cell carcinoma of cecum, poorly differentiated. The tumor invaded through the muscularis propria into pericolic tissues and involved serosa. (Fig 3-6)

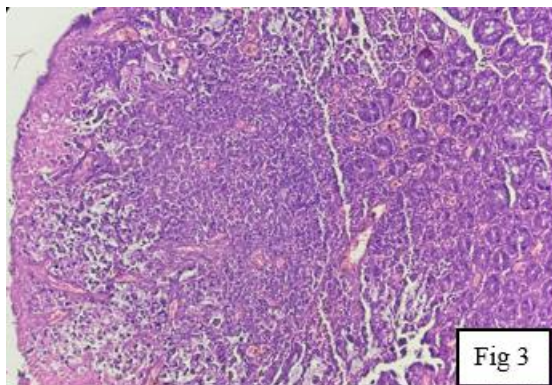


Fig 3

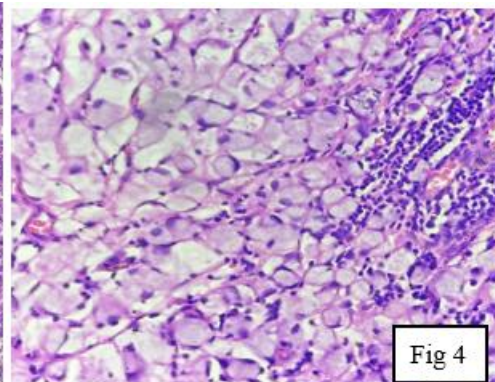


Fig 4

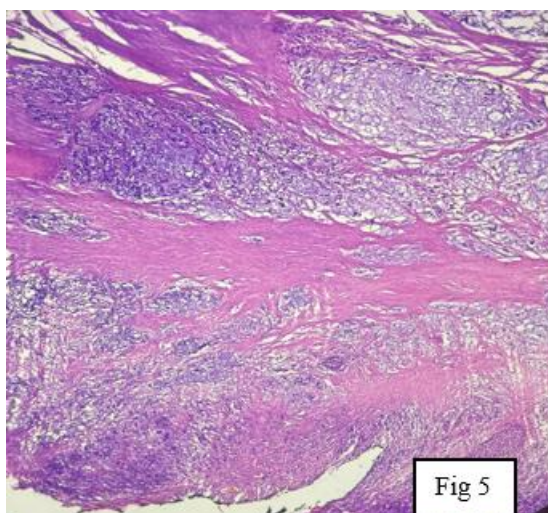


Fig 5

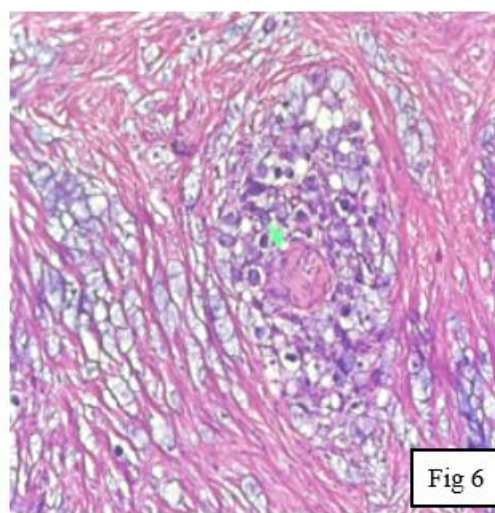


Fig 6

The adjacent colon showed serosal deposits of signet ring cell carcinoma. The ileal resected end, intervening ileal mucosa and appendix were not involved. One of 16 lymph nodes isolated showed tumor deposits. (1/16) Additionally, two tumor deposits were noted without any residual lymph node tissue in the pericolic fat. (Fig 7 and 8)

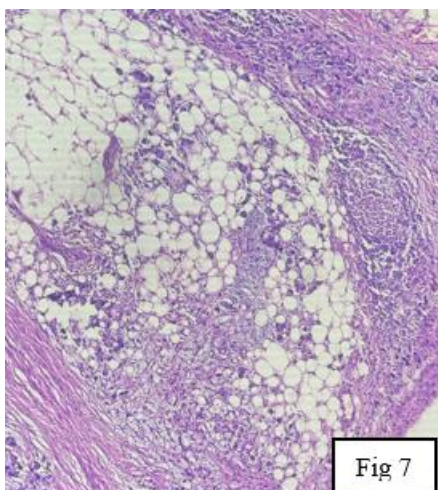


Fig 7

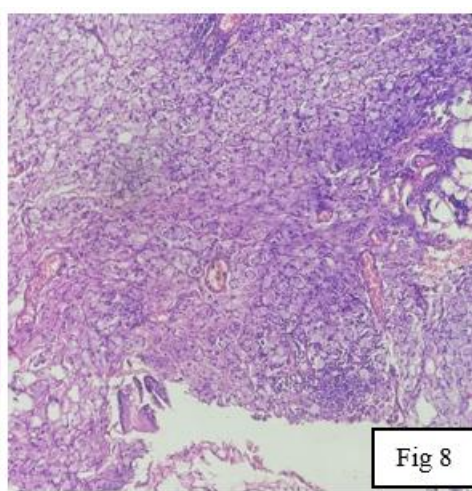


Fig 8

Finally, a diagnosis of Signet ring cell carcinoma, poorly differentiated was rendered and pathological staging as per AJCC TNM (8<sup>th</sup> edition) pT4b N1a was given. The recovery was uneventful, and the patient is in good health after one – month of follow-up.

### 3. Discussion

Signet ring cell carcinoma is rare and aggressive subtype of colorectal adenocarcinoma.<sup>1</sup>Most commonly it arises in stomach. However, it can occur at any site and has been

described in lung, breast, gallbladder, pancreas, bladder, and rarely in colon.<sup>2</sup>The most frequent localization in colon is the right side of colon, whereas 20 % cases have been reported in rectum.<sup>3</sup> Signet ring cell carcinoma is defined by presence of >50% of tumour cells with intracytoplasmic mucin.<sup>4</sup> According to WHO's classification signet ring cell carcinoma is defined as poorly cohesive carcinoma predominantly composed of tumor cells with prominent intracytoplasmic mucin and eccentrically placed crescent-shaped nucleus.<sup>5</sup>As compared to conventional adenocarcinoma, signet ring cell carcinoma has distinct molecular and clinicopathological

characteristics. SRCC is predominately observed in younger group (usually age less than 40 years), associated with right sided colon involvement, advanced stage at presentation, and a dismal prognosis.<sup>6-8</sup> Later and advanced stage of presentation has been attributed to the non-specific clinical manifestations and resemblance of tumor to inflammatory process in radiological and endoscopic evaluation resulting in delay in diagnosis.<sup>9</sup> The aggressivity is due to high rate of lymphovascular invasion, extensive peritoneal seeding, and lower incidence of hepatic metastasis. These cells have lower incidence of KRAS mutation and a higher BRAF mutation rate as compared to conventional adenocarcinoma. Also, microsatellite instability is more prevalent in SRCC.<sup>10</sup> The 5-year survival rate ranges from 0% to 12%, and disease recurrence is more frequent compared to other Adenocarcinomas.<sup>7</sup> Hence early diagnosis and a multidisciplinary approach are essential for appropriate management.

#### 4. Conclusion

Caecal signet ring cell carcinoma is a rare and a distinct entity based on the clinical and pathological features. Presence of signet ring cells is an independent prognostic factor associated with less favourable outcome, hence it is important to consider SRCC in our differential in patients presenting with and vague gastrointestinal symptoms, as signet ring cell carcinoma is an aggressive subtype of colorectal cancer with poorer prognosis.

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