CASE REPORT

Primary colonic signet ring cell carcinoma in a young patient

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SUMMARY

A 28-year-old woman presented with colicky abdominal pain for 3 months. Pain was associated with episodes of vomiting, abdominal distension and constipation. She also had loss of weight for this duration. General physical examination was unremarkable and the abdomen was soft, with no palpable organomegaly. A CT of the abdomen showed small bowel and ascending colon dilation with multiple air fluid levels. There was also a short segment of circumferential bowel wall thickening and luminal narrowing in the hepatic flexure with sudden transition of bowel diameter. She underwent a right hemicolectomy after necessary preoperative investigations. Histopathology revealed signet ring cell carcinoma (SRCC). This case highlights the importance of detecting such a lesion in a young, otherwise fit woman. The challenge lies in early diagnosis and awareness of general practitioners about this aggressive form of colonic tumours.

BACKGROUND

Colorectal cancer (CRC) is the fourth most common cancer to cause death accounting for 8% of cancer deaths worldwide.¹ Typically CRC occurs past the fifth or sixth decades of life and only about 20% of the cases are found in patients before 50 years of age. This figure is location dependent and it is necessary to highlight that countries with an ageing population are more likely to see incidences past the 50-year mark, mostly in the West. In younger patients, signet ring cell carcinoma (SRCC) is much more common than adenocarcinoma of the colon,² which is otherwise the most commonly encountered colorectal neoplasm. SRCC is an aggressive form of adenocarcinoma and is poorly differentiated. Histopathology of SRCC shows peripheral accumulation of the mucinous component thereby giving it the characteristic shape of a signet ring. Diagnosis of these cancers remains challenging due to their rapid spread and a lack of awareness in the younger population. The main problem is the late presentation in all age groups as the tumour takes about a decade to transform into a carcinoma from an adenoma. Hence there is a need for awareness to raise clinical suspicions early in such cases.

CASE PRESENTATION

A 28-year-old housewife presented with abdominal pain for 3 months. The nature of the pain was intermittent and colicky with associated episodes of vomiting, abdominal distension and constipation. Her vitals were all stable apart from a history of

intermittent low-grade fever. She also had loss of weight for this duration. The patient was passing stools and flatus and had no bladder disturbances. There was also no history of haematemesis or melaena.

Her past history was unremarkable except for two normal vaginal deliveries. She had regular menstrual cycles. General physical examination was unremarkable and the abdomen was soft, with no palpable organomegaly. There was no free fluid in the abdomen and bowel sounds were present. A subsequent per rectum examination was normal.

INVESTIGATIONS

Full blood count, random blood glucose and renal function tests were all within normal parameters. However, liver function tests showed decreased liver functioning.

An erect X-ray of the abdomen showed multiple air fluid levels in the bowel.

A contrast enhanced CT (CECT) scan of the abdomen showed small bowel and ascending colon dilation with multiple air fluid levels. There was also a short segment of circumferential bowel wall thickening and luminal narrowing in the hepatic flexure with sudden transition of bowel diameter here. The transverse and descending colon had collapsed ([figure 1](#)).

Colonoscopy showed a stricturous growth in the hepatic flexure and the scope could not be advanced further. Stenting could have been considered as a bridge to more elective surgery, but was not offered to our patient. There were no synchronous lesions and the histology was reported as primary SRCC of the colon.

The final histopathology showed an ulcerated tumour with malignant cells having abundant cytoplasm, eccentric and hyperchromatic indented nucleus (signet ring cells) with some prominent nucleoli infiltrating through the muscularis layer and extending upto the serosal surface ([figure 2](#)). There was no vascular invasion or perforation in the tumour. The resected margins, appendix and nine isolated lymph nodes were all free of tumour. Hence, the lesion was classified as being T3 N0 M0.

DIFFERENTIAL DIAGNOSIS

Tuberculosis (TB) of the abdomen was considered due to a high incidence of TB in the Indian subcontinent and its unusual similar presentation. Other working diagnoses were thought to be either inflammatory bowel disease or irritable bowel syndrome (due to the repeated admissions).

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Figure 1 Contrast enhanced CT scan of the abdomen which showed small bowel and ascending colon dilation with multiple air fluid levels. There was also a short segment of circumferential bowel wall thickening and luminal narrowing in the hepatic flexure with sudden transition of bowel diameter.

The patient was admitted at her local hospital repeatedly for similar symptoms and was diagnosed as having typhoid, which was subsequently managed conservatively. The reports suggested low titres.

TREATMENT

A right radical hemicolectomy was performed (figure 3). All the isolated lymphnodes were free of tumour. The liver was normal and there was no ascites or peritoneal spread.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and she recovered completely of her symptoms. She has been recommended chemotherapy by the oncologist.

Figure 2 Histopathology showing an ulcerated tumour with malignant cells having abundant cytoplasm, eccentric, hyperchromatic indented nucleus (signet ring cells).

Figure 3 Surgical specimen showing ascending colon with the lesion.

DISCUSSION

SRCC presents a diagnostic challenge for most clinicians. This is largely attributed to its predominance in the younger population, subsequent diagnosis at an advanced stage and poor prognosis.³ An association has also been found in patients with hereditary non-polyposis colorectal cancer and this can be explained by origination of SRCC from an adenoma.⁴ Stomach is predominantly affected in over 96% of the cases, with the rest occurring in the colon, rectum, pancreas, bladder and breast.⁵ Laufman and Saphir were the first to describe this as a primary neoplasm in 1951.⁶ SRCC has an incidence of 0.1–0.8% and accounts for only 2% of all colon cancers in comparison to conventional adenocarcinomas which have an incidence rate of 0.8–2.6%.⁷ The average age of occurrence is reported to be 23–53 years.⁵

Peritoneal spread as well as ovarian metastases is commonly found with fewer reported cases of liver metastases and this distinguishes SRCC from non-SRCC.⁸ In our patient, the liver and peritoneum was tumour free and no other metastases were found intraoperatively. Histologically these cells have large amounts of intracytoplasmic mucin with peripherally pushed nuclei. Lesions limited to the mucosa or submucosa are rarely found, as these patients are generally asymptomatic and do not often present early. Staging these lesions is also challenging due to the infiltrating character of SRCC tumours. In our case, the lesion was seen extending through muscularis layer, advanced

due to late appearance of symptoms. Patients usually report experiencing altered bowel habits, weight loss and blood and mucus in the stools. This is very similar to features of the irritable bowel disease (IBS) and an association between ulcerative colitis and SRCC has also been reported in the past (about 30%).⁶ In our case, CECT helped prompt colonoscopy to visualise if the lesion fits with a differential diagnosis of either inflammatory bowel disease or IBS.

Messerini *et al*⁹ report an affliction of SRCC for right colon compared to the left like our case. There seems to be considerable variation in the reports of SRCC and this can be attributed to its histological similarity with Linitus Plastica (spheroidal cells, mucin secretion, small and miniature new gland formations).⁶ The two have often been misinterpreted during histological reporting due to these similarities, primarily due to the huge variations in SRCCs.

The 5-year survival of primary SRCC is reported to be between 10–50%.^{9, 10} This is largely owing to the late presentation of younger patients and an initial diagnosis, in most cases, of irritable bowel disease. There have been a few reports of SRCCs being located in areas of difficult visualisation radiographically or endoscopically,¹¹ although in our case, the location was identified with CECT of the abdomen. Surgical resection is the only current modality of cure and adjuvant chemotherapy (5-fluorouracil based for colonic lesions) seems to be of some benefit; this is dependent on the histology of the lesion. The WHO classification for a lesion to be SRCC is the presence of >50% signet ring cells in a sample.¹² The present

studies to determine the efficacy of chemotherapy have included lesions with less than 50% signet ring cells and the benefit of chemotherapy is still to be ascertained. Signet ring cell type tumours are more likely to have serosal infiltration, peritoneal dissemination, lymph node involvement, organ infiltration and poorer overall and stage-specific survivals.¹³ Although our patient is not a candidate for adjuvant chemotherapy based on the tumour status of T3 N0 M0, the classification of the tumour type as SRCC means the patient has to have combined standard chemotherapy.¹⁴ This is known to have an impact on the prognosis and overall patient survival.¹⁵

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Contributors RP was involved in the concept and design of the case report and also the guarantor. NK was involved in manuscript preparation and SK in manuscript editing. RS was involved in defining the intellectual content and literature search.

Competing interests None.

Patient consent Obtained.

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Learning points

- Colorectal signet ring cell carcinoma (SRCC) is a common histopathological neoplasm seen in younger people, despite adenocarcinomas of the colon being the most common among adults overall.
- SRCC has an incidence of 0.1–0.8% and accounts for only 2% of all colon cancers.
- These tumours often present at an advanced stage due to their insidious onset and matters are further complicated due to diagnostic challenges.
- The 5-year survival of primary SRCC is reported to be between 10 and 50%. This is largely dependent on the stage of the lesion and its spread.
- Surgical resection is the only current modality of cure.

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