

Case Series

Primary linitis plastica of the rectum: Case series with emphasis on “target sign” on MRI

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ABSTRACT

Colorectal cancers are more common in the West than in Asian subcontinent. An increasing trend in the occurrence of colorectal signet cell carcinomas has been observed, exhibiting association with inflammatory bowel disease. Its distinct clinical features, pathognomonic, histologic, and radiologic appearance make it an unmissable entity. We report two such cases in the background of inflammatory bowel disease. We aim to familiarize our readers with its cross-sectional imaging features.

Keywords: Linitis plastica, Target, Signet ring cell, Colon

INTRODUCTION

Signet cell adenocarcinomas are a common occurrence in the stomach. When these occur in the colorectum (with this histologic variant accounting for <1% of the colorectal cancers), they follow a more aggressive course, often presenting at an advanced stage.^[1] Inflammatory bowel disease and familial colorectal cancers related syndromes are strongly associated with colonic signet cell adenocarcinomas. Linitis plastica is synonymous with desmoplastic reaction due to intramural infiltration of tumor cells in a hollow viscus, phenotypically manifested as a rigid, shrunken organ with thickened walls.^[2] This entity, although can rarely present as a primary malignancy, is commonly described as secondary to metastatic disease from carcinoma of gallbladder, breast, bladder, and stomach.^[3]

CASE 1

A 38-year-old female diagnosed with ulcerative colitis 1.5 decades back, taking on and off medications for the same, presented with the complaints of lower abdominal colicky pain for 2 months, which reduced in intensity on passing flatus. She complained of loss of appetite with significant weight loss and increased frequency of stools. No history of rectal bleeding, fever, abdominal distension, hematemesis, melena, bony pain, and jaundice was found. Colonoscopy revealed a tight concentric stricture 7–27 cm from the anal verge with friable and ulcerated mucosa [Figure 1]. Distal colonic biopsy was reported as ulcerative colitis with moderate activity, while the rectal biopsy revealed signet ring cell adenocarcinoma.

A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was performed which revealed circumferential “whorl-like” wall thickening with significant luminal

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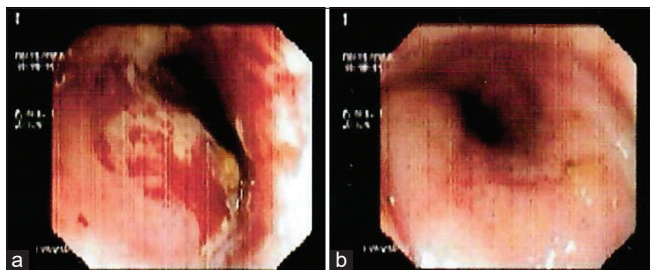


Figure 1: (a and b) Colonoscopy showing concentric tight stricture with overlying friable and ulcerated mucosa and changes of Grade II colitis.

narrowing involving the rectosigmoid. Significant mesorectal and pericolon fat stranding with fascial thickening was also seen. Pelvic magnetic resonance imaging (MRI) was done which confirmed the findings. T2-weighted images and diffusion-weighted images revealed triple-layered thickening, consisting of hyperintense intermediate layer showing diffusion restriction, along with hypointense non-diffusion restricting innermost and outer layers, giving rise to “target sign.” Predominant infiltration into submucosa and muscularis propria was demonstrated with sparing of the mucosa, like other scirrhous carcinomas [Figure 2]. Given the locally advanced disease, the patient is being treated with neoadjuvant chemotherapy and radiotherapy comprising 25 cycles of radiotherapy with concurrent oral capecitabine – 500 mg.

CASE 2

A 21-year-old male with no significant history presented with the complaints of abdominal pain that aggravated on passage of stools, along with increased stool frequency and significant weight loss. Contrast-enhanced CT abdomen and pelvis were performed which revealed long segment circumferential thickening of the rectum, anal canal, and rectosigmoid junction with significant luminal narrowing. MRI was also performed for local staging which again revealed targetoid pattern of thickening [Figure 3]. Punch biopsy from the palpable mass revealed an invasive tumor involving the submucosa with multiple signet ring cells [Figure 4]. The patient is being treated with gap filling chemotherapy and radiotherapy.

DISCUSSION

Linitis plastica or Brinton disease refers to the remarkable resemblance of the shrunken/rigid appearance of the neoplastic infiltrative hollow organs to the entrapped cells in a linen cloth. While historically described in stomach, this term has now extended to the colorectum.^[4] A meager <0.1% of primary colorectal malignancies are accounted for by signet ring cell adenocarcinomas.^[5] Early diagnosis

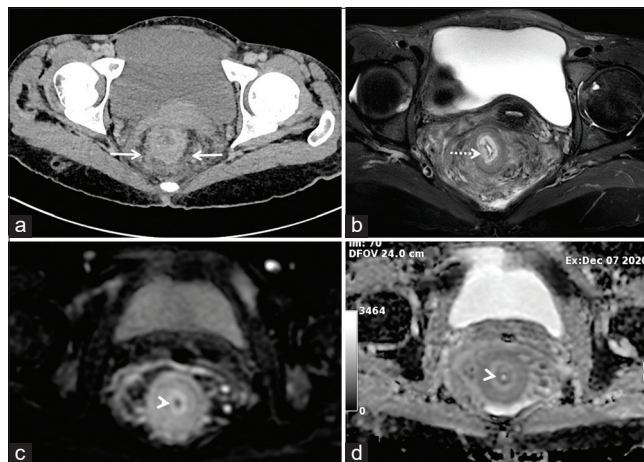


Figure 2: (a) Contrast-enhanced computed tomography showing enhancing circumferential “whorl-like” thickening of the rectal wall with florid perirectal fat stranding (arrows). (b) T2WI depicting the “target sign” with hyperintense intermediate zone (dashed arrow) representing the infiltrated submucosa, sandwiched between the hypointense inner and outer zones. (c and d) Diffusion-weighted and ADC maps showing diffusion restriction in the intermediate zone corresponding to tumor infiltration into the submucosa (arrowhead).

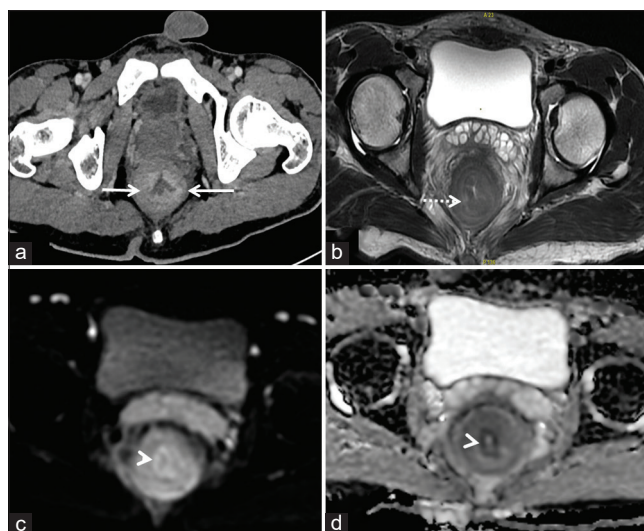


Figure 3: (a) Contrast-enhanced computed tomography showing circumferential thickening of the rectal wall with hyperenhancement of the submucosa (arrows). (b) T2WI depicting the targetoid thickening with hyperintensity (dashed arrow) in the submucosa. (c and d) Diffusion-weighted and ADC maps showing diffusion restriction in the submucosa (arrowhead).

and a high index of suspicion, when encountered with the classic radiologic signs, may negate major surgery and aid in extending patient survival time.

Case 1 had been under the purview of ulcerative colitis for longer than a decade, which abides with the estimated risk of

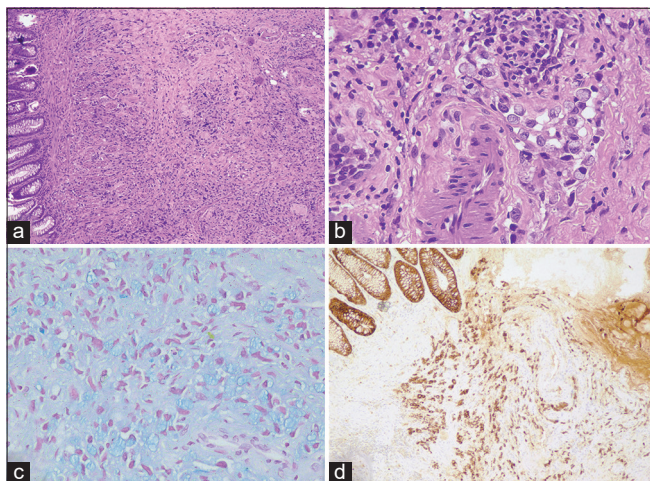


Figure 4: (a) Low-power view showing poorly differentiated tumor composed of singly infiltrating malignant cells (H&E, $\times 100$). (b) High-power view showing signet ring cells with eccentric hyperchromatic nuclei and intracytoplasmic mucin (H&E, $\times 400$). (c) Alcian blue stain highlighting the intracytoplasmic mucin of the signet ring cells ($\times 400$). (d) Immunohistochemistry for pancytokeratin (IHC, panCK, $\times 100$).

occurrence of colorectal cancer of 8% at 20 years of disease duration.^[6] On the other hand, Case 2, while still being evaluated for inflammatory bowel disease, graduated to advanced stages of the disease within a span of 2 months of the onset of symptoms, essentially serving as a telltale of the aggressive, unpredictable course of the disease.

Both our cases showed the characteristic concentric ring pattern as described by Rudralingam *et al.*^[7] The specificity of this sign for linitis is debatable as it is also found in ischemic, inflammatory, and infectious pathologies of the colon.^[8]

As was elucidated by Ha *et al.*,^[9] both our cases demonstrated the target sign with CT showing hypoattenuating intermediate zone sandwiched between hyperattenuating inner and outer zones with long segment circumferential thickening of the involved segment of large bowel.^[9] The histopathological correlation of this sign, that is, sparing of mucosa with infiltration of the submucosa and muscularis propria, was confirmed in both cases. Important sequences to acquire on pelvis MRI are two-dimensional FSE-T2 without fat suppression (TR 9060, TE 107) in sagittal, coronal, and axial planes for the evaluation of tumor characteristics, along with diffusion weighted imaging (DWI) (TR 8800, TE 84) to assess the cellularity of the tumor. T2-weighted images reveal a hyperintense intermediate layer due to the proliferation of mucin-containing cells in the submucosa and muscularis propria. This layer shows restricted diffusion on DWI due to densely packed cells, with intracellular edema and elevated viscosity. Hydrophobic cell membranes contribute to elevated density.^[9] The innermost and outmost layers of the three-

layered thickening show a T2 hypointense appearance with no restricted diffusion. Thus, these three layers cumulatively give rise to a concentric appearance, resulting in a “target sign.” This sign could also occur secondary to stratification brought about by preferential zonal tumor infiltration.

CONCLUSION

We conclude with the suggestion that the appearance of target sign in the occurrence of long segment circumferential thickening of colorectum in young/middle aged and irrespective of history of inflammatory bowel disease should warrant the exclusion of colorectal linitis plastica by biopsy. Positive family history, the clinical background of IBD, and primary cancers in other organs would point toward secondary colorectal linitis plastica.

Teaching points

1. Target sign on MRI and CT in the occurrence of long segment circumferential thickening of colorectum irrespective of history of inflammatory bowel disease, in young/middle-aged, should raise suspicion of linitis plastica, and must be confirmed/excluded via biopsy.
2. Preferential zonal infiltration of the intermediate zone with sparing of mucosa that gives rise to alternate layers of attenuation is seen in signet ring cell carcinoma.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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