

Primary Rectal Linitis Plastica: Report of Two Cases and Literature Review

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Abstract

Background: Linitis plastica is classically described in the stomach, characterized by a diffuse infiltrative growth pattern and poor prognosis. Primary rectal involvement is extremely rare. Case presentation: We report two cases of primary rectal linitis plastica diagnosed at Hassan II University Hospital of Fez. Both patients presented with rectal syndrome and weight loss. Imaging revealed circumferential rectal wall thickening. Histopathology confirmed poorly differentiated adenocarcinoma with signet-ring cells. Despite neoadjuvant chemoradiotherapy, the tumors were deemed unresectable, and both patients were referred for palliative chemotherapy. Conclusion: Primary rectal linitis plastica is a rare but aggressive malignancy. Diagnosis requires deep biopsies and exclusion of secondary involvement. Prognosis remains poor due to late presentation and limited therapeutic options.

Keywords: Linitis plastica; Rectal syndrom; Adenocarcinoma; Chemotheray; Radiotherapy

1. Introduction

Linitis plastica is a term used to describe tumors that diffusely infiltrate the wall of a hollow viscus, resulting in a rigid, "leather bottle" appearance. Histologically, it corresponds most often to an adenocarcinoma with independent, poorly cohesive cells, frequently with a signet-ring morphology. While gastric linitis plastica is well known, primary rectal localization is extremely rare, representing less than 1% of colorectal cancers. This entity poses diagnostic and therapeutic challenges due to its infiltrative nature, frequent negative superficial biopsies, and advanced stage at presentation.

2. Case Reports

2.1. Case 1

A 31-year-old female, with a history of ovarian cyst surgery, was admitted for rectal bleeding and unquantified weight loss. Abdominal examination was unremarkable. Digital rectal examination revealed a fixed, stenosing mass 2 cm from the anal verge. Vaginal examination found a thickened, rigid posterior vaginal wall. Rectoscopy identified a stenosing lesion at 3 cm from the anal verge, which was impassable. Biopsies showed poorly cohesive carcinoma with signet-ring cells.

Thoraco-abdominal-pelvic CT revealed a circumferential rectal wall thickening extending over 10 cm, with perilesional fat infiltration and regional lymphadenopathy. Upper endoscopy and total colonoscopy excluded another primary tumor. A diagnosis of primary rectal linitis plastica was established. The patient received neoadjuvant chemoradiotherapy but the disease progressed locally and was considered unresectable. She was referred for palliative chemotherapy.

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2.2. Case 2

A 38-year-old male, without significant medical history, presented with rectal syndrome and weight loss. Abdominal examination was normal. Digital rectal examination was painful and revealed poor sphincter tone and a firm pelvic mass. Rectoscopy under sedation showed circumferential rectal mucosal infiltration and stenosis 4 cm from the anal verge. Biopsies revealed a poorly differentiated infiltrating carcinoma with signet-ring cell features. CT demonstrated low rectal wall thickening with iliac lymphadenopathy. Upper endoscopy excluded a gastric or colonic primary. After chemoradiotherapy, pelvic MRI showed persistent posterior circumferential tumor involving both internal and external sphincters. At laparotomy, the tumor was deemed unresectable. A sigmoidostomy was performed and the patient was referred for palliative chemotherapy.

3. Discussion

Primary rectal linitis plastica (PRLP) represents a rare clinicopathological entity, accounting for less than 1% of colorectal malignancies. It is histologically characterized by a poorly cohesive adenocarcinoma, most often with signet-ring cell features, that infiltrates diffusely through the rectal wall, leading to a rigid, circumferential thickening. Unlike classical exophytic or ulcerative adenocarcinomas of the rectum, linitis plastica develops in the submucosa and muscularis propria, explaining the frequent discrepancy between dramatic clinical and radiological findings and the sometimes negative or inconclusive superficial mucosal biopsies.

Diagnostic challenges: Diagnosis is particularly difficult. Clinically, patients usually present with nonspecific symptoms such as rectal bleeding, constipation, pelvic pain, or progressive obstructive syndrome, often in the context of unexplained weight loss. On digital rectal examination, the rectal wall may feel rigid, thickened, and stenotic, sometimes mimicking inflammatory conditions or endometriosis in women. Radiological imaging is crucial. Pelvic MRI is considered the most accurate modality, showing circumferential wall thickening with hypointense signal on T2-weighted images and restricted diffusion, often associated with spiculated extension into perirectal fat and pelvic structures. CT may demonstrate similar features but with less precision in locoregional staging. Endoscopic ultrasound can reveal hypoechoic circumferential thickening involving the entire rectal wall.

Histologically, the infiltrative nature of PRLP results in frequent false negatives on standard rectal biopsies, as the mucosa may appear intact. Deep biopsies, transrectal ultrasound-guided biopsies, or full-thickness surgical specimens may be necessary to confirm the diagnosis. Immunohistochemistry plays an essential role in distinguishing primary rectal origin from secondary linitis due to metastasis of gastric, breast, or prostatic carcinoma. Typically, colorectal origin is supported by CK20 and CDX2 positivity, whereas CK7 positivity suggests another primary site.

Pathogenesis and differential diagnosis: The pathogenesis of PRLP remains poorly understood. Some authors suggest that the predominance of signet-ring cell carcinoma reflects a distinct molecular pathway, possibly involving alterations in cell adhesion molecules such as E-cadherin. Differentiating primary from secondary linitis is critical. Gastric linitis plastica can metastasize to the rectum through peritoneal seeding or lymphatic spread, while breast lobular carcinoma and prostate adenocarcinoma are also known to mimic PRLP. Hence, a complete upper gastrointestinal endoscopy, colonoscopy, and systemic imaging are mandatory before confirming the diagnosis.

Therapeutic management: Management of PRLP is challenging and not yet standardized. Surgery is the only potentially curative treatment but is rarely feasible due to the advanced stage at diagnosis, frequent pelvic extension, and poor response to neoadjuvant therapy. When resection is attempted, it usually requires extended pelvic surgery with high morbidity and questionable survival benefit. Neoadjuvant chemoradiotherapy, standard for locally advanced rectal adenocarcinoma, has been used in PRLP with variable results. Tumor downstaging is often limited because of the diffuse stromal infiltration and relative radioresistance of signet-ring cell carcinomas. Systemic chemotherapy with regimens such as FOLFOX or FOLFIRI has been reported, but responses are generally modest. Targeted therapy and immunotherapy have not been specifically studied in this entity, though extrapolation from colorectal cancer guidelines may be considered in selected cases. In most situations, palliative management, including diversion stoma to relieve obstruction and systemic chemotherapy, remains the mainstay of treatment.

Prognosis: The prognosis of PRLP is extremely poor. Several series report a median survival of less than 12 months from diagnosis. Prognostic factors include the stage at presentation, possibility of complete surgical resection, and response to systemic therapy. Unfortunately, due to its insidious presentation and aggressive biology, PRLP is usually diagnosed at an unresectable stage, as in our two patients.

Contribution of our cases: Our two cases highlight the typical features of PRLP: young patients, nonspecific symptoms, late presentation, imaging suggestive of diffuse infiltrating rectal tumor, histology revealing signet-ring cell carcinoma, and unresectability despite chemoradiotherapy. These cases confirm the aggressive course and therapeutic limitations described in the literature. They also emphasize the importance of considering PRLP in the differential diagnosis of rectal stenosis with inconclusive superficial biopsies.

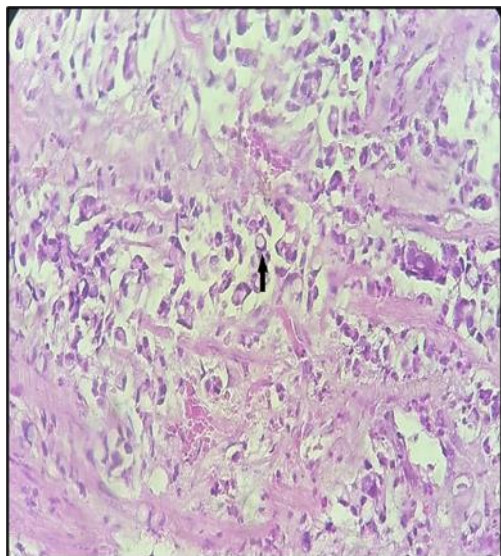


Figure 1



Figure 2



Figure 3

Figure 1 Histological section showing diffuse infiltration of the rectal wall by poorly cohesive adenocarcinoma cells with a desmoplastic reaction

Figure 2 Pelvic MRI demonstrating circumferential thickening of the rectal wall with hypointense signal on T2-weighted sequences, highly suggestive of rectal linitis plastica

Figure 3 Colonoscopy revealing a narrowed rectal lumen with rigid, infiltrated wall and loss of distensibility, typical of rectal linitis plastica

4. Conclusion

Primary rectal linitis plastica is an exceptional entity with aggressive behavior and poor prognosis. Its diagnosis relies on a combination of clinical suspicion, imaging, and histopathology, while excluding metastatic disease. Early

recognition and multidisciplinary management are essential, but most patients present with unresectable tumors, making palliative care the most frequent option.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval was obtained.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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