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Case report
Olgu sunumu

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Primary rectal linitis plastica: Report of two cases and a review of the literature

Primer rektal linitis plastika: İki olgu sunumu ve literatürün gözden geçirilmesi

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Abstract
Primary rectal linitis is a rare tumor which mostly affects young patients. The definite diagnosis is based on the histologic examination of deep biopsy samples. It is mostly diagnosed in advanced stages, which worsens the prognosis. In this report, two cases of primary rectal linitis plastica will be presented and discussed in light of the literature.
Keywords: Plastic linitis, Primary rectal tumor

Öz
Primer rektal linit, çoğunlukla genç hastaları etkileyen nadir bir tümördür. Kesin tanı derin biyopsi örneklerinin histolojik incelemesine dayanır. Çoğunlukla ileri aşamalarda teşhis edildiğinden, prognozu kötüdür. Bu raporda, iki primer rektal linitis plastica olgusu literatür eşliğinde sunulacak ve tartışılacaktır.
Anahtar kelimeler: Plastik linit, Primer rektal tümör

Introduction

The conventional definition of plastic linitis, a.k.a. “scirrhous carcinoma”, is a tumor that massively infiltrates the entire thickness of the wall of a hollow organ. Histologically, it corresponds to an adenocarcinoma of independent cells with a signet ring appearance, and usually affects the stomach. The involvement of another digestive organ is often secondary. In fact, the primary rectal linitis is a rare entity [1].

Below are presented two patients diagnosed with primary rectal linitis.

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Case presentation

Case 1

A 31-year-old female patient, who was operated for an ovarian cyst in 2015, was admitted with progressive hematochezia lasting for two months and severe weight loss. At 2 cm from the anal margin (AM), we palpated a circumferential, stenosing and obstructive mass fixed to the anterior and posterior planes. On examination, the posterior wall of the vagina was thick and rough. Biopsies revealed rectal carcinoma with independent cells in a signet-ring appearance (Figure 1). The thoraco-abdomino-pelvic computerized tomography (CT) revealed a thickening 3 cm proximal to the AM, extending for 10 cm and infiltrating the perilesional fat, along with adjacent lymphadenopathy (Figure 2). The patient underwent an esophagogastroduodenoscopy (EGD) and a colonoscopy to search for a primary source of tumor, which both turned out normal, hence the diagnosis of primary rectal linitis. The patient received a long-term radiotherapy protocol with concomitant chemotherapy. The disease progressed, as marked by local extension of the tumor which was deemed unresectable, and the patient was referred for palliative chemotherapy.

Case 2

A 38-year-old male patient with no medical history was admitted for rectal bleeding, proctalgia, and transit disorder with a weight loss of 10 kg evolving for six months. The abdominal examination was normal, and digital rectal examination was very painful. The patient underwent a rectoscopy on sedation, which revealed an infiltrated, stenotic mucosa 4 cm proximal from the AM (Figure 3). The biopsies were reported as undifferentiated and infiltrative carcinoma with signet ring cells. A thoraco-abdomino-pelvic CT scan showed a tumoral thickening of the lower rectum with secondary iliac lymphadenopathies. The patient underwent an EGD which was normal, then received radiotherapy concurrent with chemotherapy. Magnetic resonance imaging (MRI) scan revealed a posterior hemicircumferential lower rectum mass, infiltrating the rectal mucosa along with the internal and external sphincters, without prostatic or bladder involvement (Figure 4). Intraoperative exploration revealed an unresectable tumor. A sigmoidostomy was performed, after which the patient was referred for palliative chemotherapy.

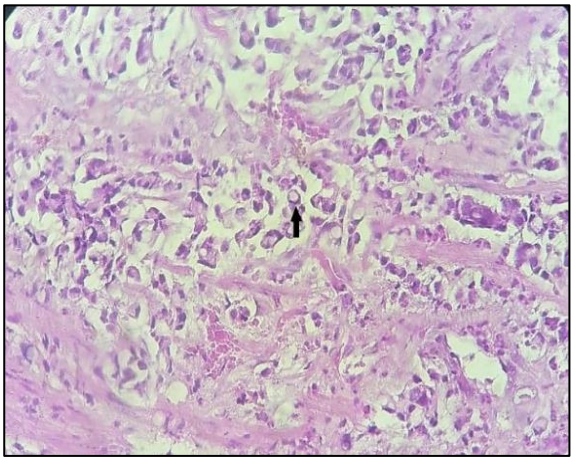

Figure 1: Microscopic image shows the histological appearance of rectal linitis: rectal carcinoma with independent cells

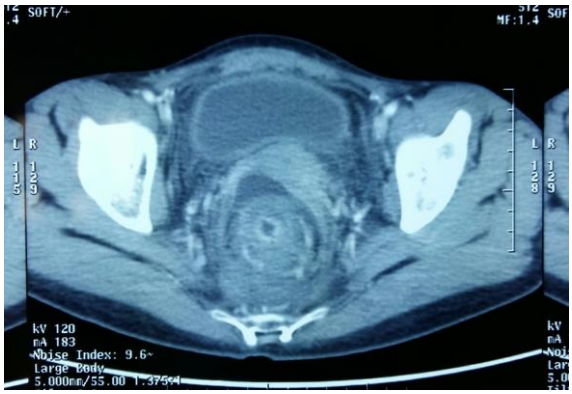

Figure 2: CT image shows the local extension of rectal linitis

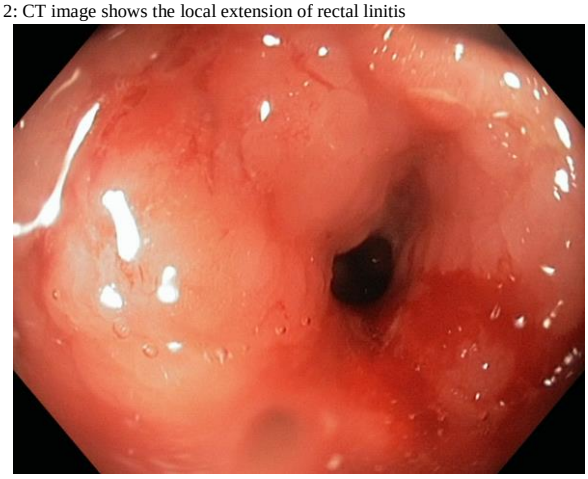

Figure 3: Rectoscopy shows the infiltrating and stenosing aspect of rectal linitis

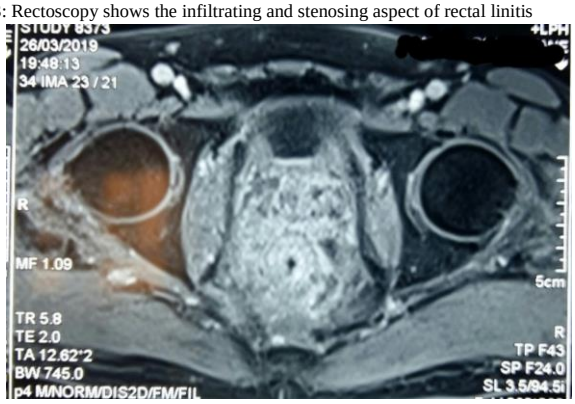

Figure 4: Image of MRI shows a rectal process with internal and external sphincter invasion

Discussion

Rectal primary plastic linitis, accounting for 1/1,000 for colonic and rectal cancers, is rare [1], and more common in men (sex ratio M/F: 1,3). Most authors insist on the occurrence of linitis at an early age [1,2], which is the case of our two patients. It does not share the same characteristics and etiopathogenic factors with conventional adenocarcinomas. Indeed, genetic predisposition or malign transformation of an adenomatous polyp have not been established for this kind of tumor. A relationship between colorectal linitis and inflammatory bowel disease, particularly Crohn's disease, has been reported [2, 6]. The revealing symptoms, such as transit disorders, bowel obstruction, abdominal pain, weight loss or hematochezia, are not specific [2-4]. The diagnosis is often late. In most of cases, digital rectal examination reveals in infiltrating mass and a fixed rectal ampulla [4]. Upon endoscopic exploration, the mucosa is seen intact with a narrowed or obstructed lumen. The biopsies results are negative in 50% of the cases, which necessitates the need for deep or surgical biopsies [2]. Endoscopic ultrasonography assists in the diagnosis by guiding biopsies and

revealing a circumferential thickening reaching the mucosa and the muscular mucosa in a concentric, hypoechoic sleeve around the ultrasound probe. It plays a significant role in not only preoperative assessment and staging, but also in follow-up by measuring the regression of lesions under radiotherapy and chemotherapy [5,6]. Computed tomography and magnetic resonance imaging show locoregional and distant invasion [7]. Definite diagnosis is based on histological examination: Macroscopically, a thickening of the wall and circumferential involvement of the rectum, engulfing the mesorectum, with a most often normal mucosa [2-1] is seen. Microscopically, we find abundant (50%) signet ring cells within a dense fibrous stroma infiltrating the rectal wall [1].

The search for a primary extra-rectal tumor, especially gastric, must be systematic, because only its negativity affirms the primitive nature of the lesion. Secondary rectal linitis is more frequent, most often corresponding to the metastasis of a gastric cancer, and more rarely, to cancers of gall bladder, breast, or prostate [8,9]. Linitis has a predominantly locoregional extension to the lymphatic (lymphadenopathy: 86%), pelvic (ovary, trunk, and uterus: 58%) and peritoneal (47%) areas. Hepatic metastases are exceptional (17%). Metastases to the bone are present in 12% of patients and other sites, 6% of patients. Aggressive treatment with ganglion dissection is performed if the extension allows. Combination of radiotherapy with a chemotherapy combining 5-fluorouracil and cisplatin appears to provide better outcomes in long term survival [1]

The prognosis of primary rectal linitis is poor, since lymphatic metastases are often found immediately at diagnosis. Survival is variable from one month to two years [2-8].

Conclusion

Primary rectal linitis is a rare and an extremely aggressive tumor, diagnosed most often at advanced stages, which worsens prognosis. The definite diagnosis is based on histological examination of deep biopsy samples.

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