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CASE REPORT | INFLAMMATORY BOWEL DISEASE

Early Diagnosis of a Small Adenocarcinoma in a Rectovaginal Fistula Related to Crohn's Disease

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Abstract

While adenocarcinomas have occasionally been reported in perianal fistulae, malignant changes occurring in rectovaginal fistulae are rare, with only a handful of reported cases. We report a 73-year-old woman with Crohn's disease who was diagnosed at an early stage with adenocarcinoma in a rectovaginal fistula. This rare disease poses many diagnostic challenges.

Introduction

Crohn's disease is a chronic inflammatory disorder characterized by focal, transmural inflammation commonly involving the ileo-cecum, but it can involve any part of the gastrointestinal tract. There is an increased risk of fistula formation (17–50%) due to the transmural nature of the disease.[1] Two-thirds of these fistulas are external (perianal 55%, enterocutaneous 6%) and one-third are internal.[1] The prevalence of perianal fistulas varies according to disease location, and ranges from 12% in patients with isolated ileal disease to 92% in patients with colonic disease involving the rectum.[2] Rectovaginal fistulas represent 9% of all fistulas in Crohn's disease, and are difficult to treat.[3] Rectovaginal and anovaginal fistulas present with fecal vaginal discharge, vaginal flatus, and infection, but may be asymptomatic if the rectal stump is out-of-circuit in patients undergoing subtotal colectomy with ileostomy.

Case Report

A 73-year-old postmenopausal woman presented after "passing a small piece of tissue" from her vagina after voiding. She had longstanding Crohn's disease with perineal fistula for 23 years, for which she was on long-term oral steroids. She had undergone subtotal colectomy with end ileostomy 19 years prior, with an out-of-circuit rectal stump for which she never had surveillance; she did not return for follow-up after surgery. She had no other complaints, and examination did not reveal any masses in the vagina or rectum. On histopathological examination, the tissue that she passed from her vagina revealed adenocarcinoma with intestinal immunophenotype. Immunohistochemistry revealed neoplastic cells stained with CK7, CK20, CDX2, and villin, but were nonreactive with CA-125 and S-100, consistent with adenocarcinoma of colorectal origin.

Proctoscopic examination revealed a distal rectum stenosis and irregularity of the anterior rectal wall, but the fistulous opening was not clearly visualized. Examination of the vagina revealed granularity of the posterior wall and a fistulous opening. Rectal and vaginal biopsies were negative for malignancy. A magnetic resonance imaging (MRI) study revealed a T2 hyperintense rectovaginal fistula surrounded by a 1.6 x 1.3-cm minimally enhancing soft tissue nodule (Figure 1). Positron emission tomography–computed tomography (PET-CT) showed linear uptake in the area of the fistula without evidence of local or distant spread (Figure 2). The patient underwent posterior exenteration with removal of her rectum, anus, posterior vaginal wall, uterus, fallopian tubes, and

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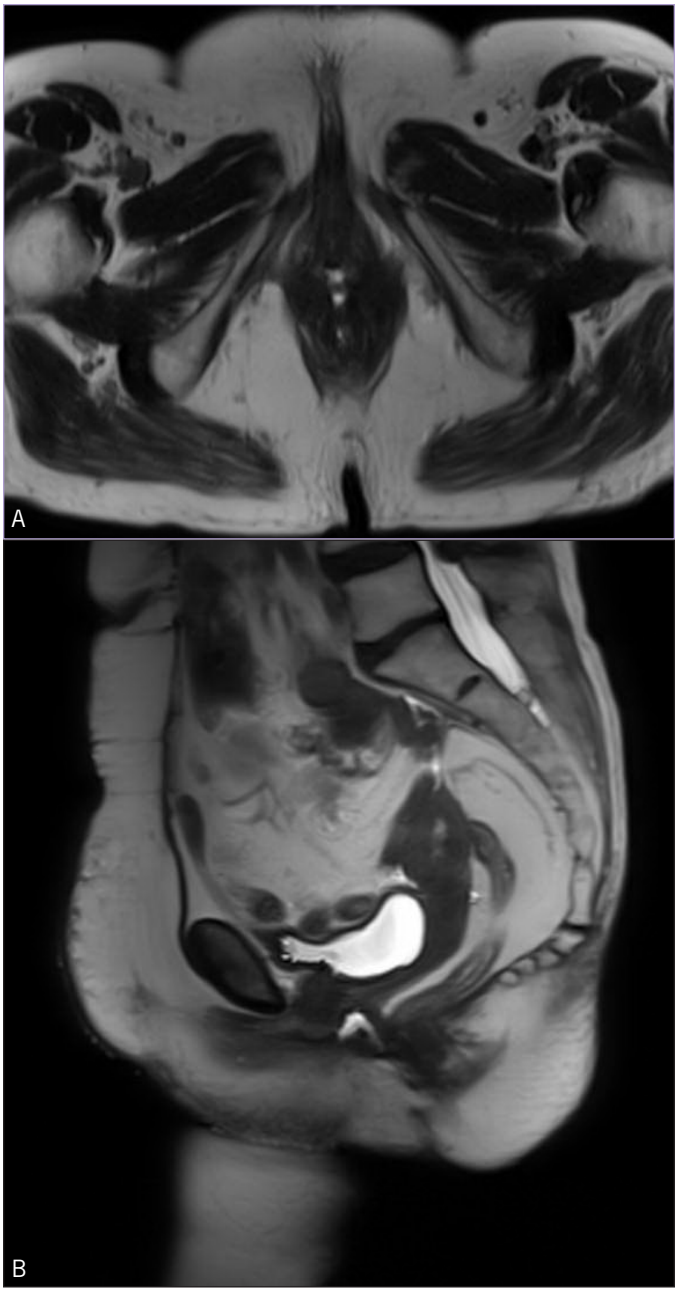

Figure 1. MRI showing (A) the fistula in cross-section between the vagina anteriorly and the anorectum posteriorly, and (B) more clearly as a T2 hyperintense fistula in sagittal plane.

ovaries. Pathology revealed high-grade adenocarcinoma involving the rectovaginal fistula, 1.2 mm in greatest dimension, and 0.8-mm invasion into the perineal body (Figure 3). The vagina, rectum, pelvic lymph nodes, and excision margins were negative for tumor. The patient opted not to receive postoperative chemotherapy given the disease was diagnosed at an early stage and surgical margins were negative. The patient has no evidence of disease at follow-up 10 months later.

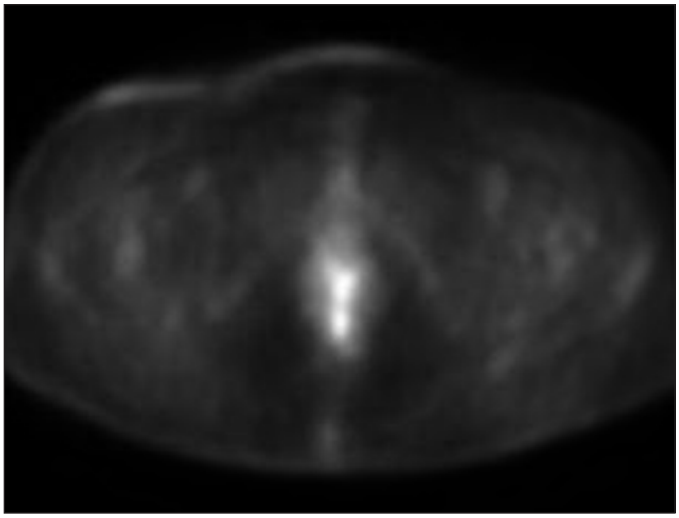

Figure 2. PET-CT showing increased uptake in the area of the fistula.

Discussion

Perianal adenocarcinoma has been reported in association with fistula-in-ano, but is very rare. Chronic inflammation has been suggested as the possible mechanism for malignant transformation in Crohn's-related fistulae.[4] Malignant change may also occur in rectovaginal fistula, but is even rarer.

A review of literature revealed 4 case reports describing 5 patients who developed adenocarcinoma arising from a rectovaginal or anovaginal fistula.[5-8] Four of these patients had longstanding Crohn's disease complicated by non-healing fistulae, while the fifth patient developed adenocarcinoma in a remnant of the rectovaginal fistula tract after surgical repair of an imperforate anus.[8] When described, the tumors

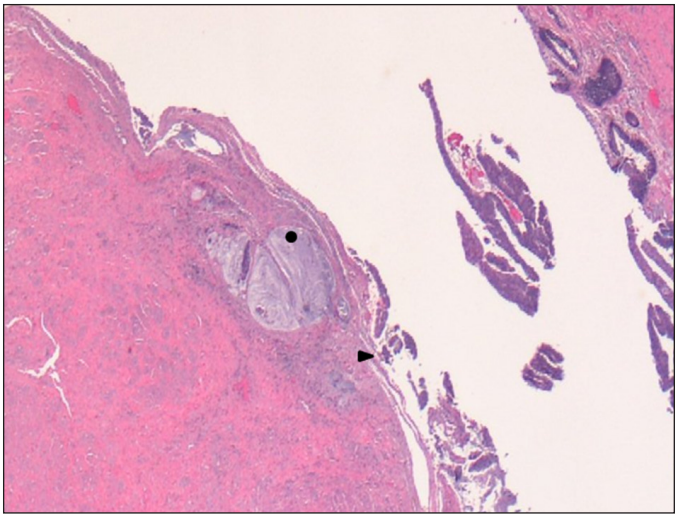

Figure 3. Histopathology with H&E stain (magnification 40x) showing the focus of invasion (circle) and dysplastic cells lining the fistula (arrowhead).

were large, with sizes ranging from 3.5 to 8 cm. Management was described in 3 patients, all of whom were treated with pelvic exenteration, with final pathology showing no rectal extension of tumor. The only patient who received adjuvant chemotherapy and radiation did not have evidence of disease at 18 month follow-up.

Malignant transformation, although rare, should be considered in the context of a non-healing rectovaginal fistulae in patients with longstanding Crohn's disease.[5-8] A high index of suspicion, close follow-up, and biopsies of any masses or the fistulous tracts are necessary for an early diagnosis. Women may present to their gynecologist rather than their gastroenterologist with vaginal bleeding and discharge. If a mass is found, it is usually in the vagina, and the rectum is not involved. MRI is the best modality to evaluate these tumors, which have high signal intensity on T2-weighted images due to their mucin content.[9] Our case illustrates another complication of the retained rectal stump after subtotal colectomy, supporting recent evidence favoring an aggressive approach to early completion proctectomy in such patients.[10]

Disclosures

Author contributions: DM Narasimhulu collected all the information, drafted the manuscript, and is the article guarantor. A. Serur and N. Khulpateea assisted in data collection and edited the manuscript.

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