Case Report

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Mucinous adenocarcinoma arising from chronic perianal crohn's disease: A case report

Abstract

Background: Carcinoma associated with chronic fistulous Crohn's disease (CD) is considered as a rare event. Malignant transformation of perineal fistula in CD has rarely been reported in literature.

Case presentation: We report a case of a 44-year-old patient with a 20-year medical history of CD. He was referred to our hospital for recurrent and refractory perineal complex fistula. The digital rectal examination did not find any mass. Colonoscopy showed left congestive and ulcerative recto colitis with inflammatory pseudo-polyps. Pelvic MRI revealed a trans-sphincteric complex fistula associated with a perianal abcess without any signs of malignancy. Examination under anaesthesia was performed, abcess was drained and multiple biopsies were taken. Histopathology concluding to a mucinous adenocarcinoma developed on anal fistula.

Conclusion: The diagnosis of carcinoma arising from a perianal fistula in patients with Crohn's disease stills a big challenge. The incidence is estimated to be 0.3-0.7% of all patients with Crohn's disease. Given the difficulty in diagnosis of carcinoma arising in complicated perianal fistula in patients with Crohn's disease, it is important that the clinician perform systematic biopsy or curettage at examination under anesthesia to avoid delaying the diagnosis of a low rectal malignancy and the radiologist be aware of the findings that may distinguish cancer from inflammatory changes.

Keywords: Crohn's disease, anal fistula, biopsy, adenocarcinoma

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Abbreviations: CD, Crohn's disease; MRI, Magnetic resonance imaging; CT, computerized tomography

Background

It is widely acknowledged that Crohn's disease (CD) increases the risk of small bowel carcinoma/lymphoma and colorectal cancer, however, the occurrence of carcinoma related to chronic fistulous CD is deemed uncommon. Malignant changes in perineal fistulas associated with CD are seldom reported in literature, with the risk of this transformation possibly being less than 5%. The relationship between Crohn's disease (CD) and small bowel carcinoma/lymphoma and colorectal cancer is well established. However, carcinoma associated with chronic fistulous CD is considered as a rare event. Malignant transformation of perineal fistula in CD has rarely been reported in literature. The risk of this transformation may be even lower than 5%.[1]

Case presentation

We report a case of a 44-year-old patient with a 20-year medical history of CD. He underwent ileocecal resection at age of 22 years. He had poor treatment adherence and a lapse in clinical follow-ups in recent years. He was referred to our hospital for recurrent and refractory perineal complex fistula. The examination of the perineum showed 4 productive external fistulas with purulent secretion and an ulceration of 4 cm (Figure1). The digital rectal examination revealed no mass. Colonoscopy showed signs of left-sided congestive and ulcerative colitis, along with inflammatory pseudo-polyps. On pelvic MRI, a complex trans-sphincteric fistula with four primary orifices was observed, along with a 4 cm perianal abscess extending into both ischioanal fossae (Figure 2). Examination under anaesthesia was performed, abcess was drained and multiple biopsies were taken. The histopathological analysis concluded to a mucinous adenocarcinoma originating from the anal fistula (Figure 3). Thoraco-abdomino pelvic CT scan showed no metastases. Abdominoperineal resection was indicated but the patient refused surgery.

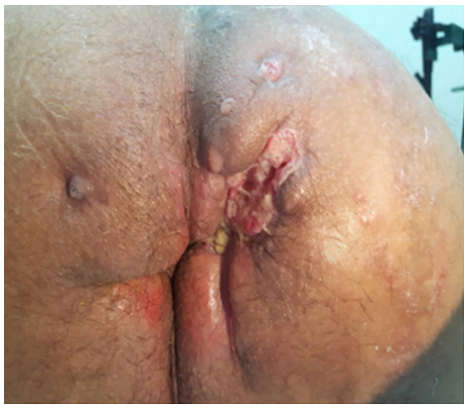

Figure 1 Examination of the patient: external fistula with ulceration of 4 cm.

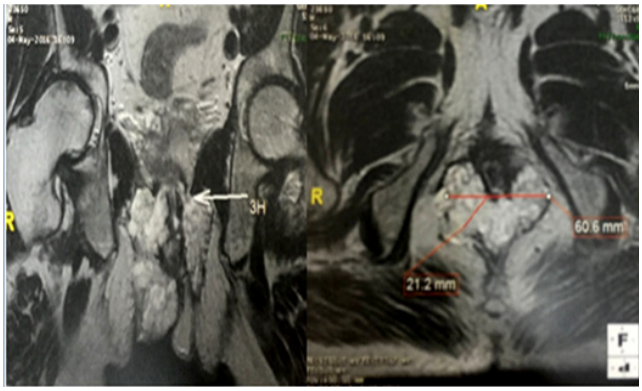

Figure 2 Pelvic MRI: trans-sphincteric complex fistula with 4 primary orifices associated with a perianal abcess measuring 4 cm extended to the 2 ischio-anal fossae.

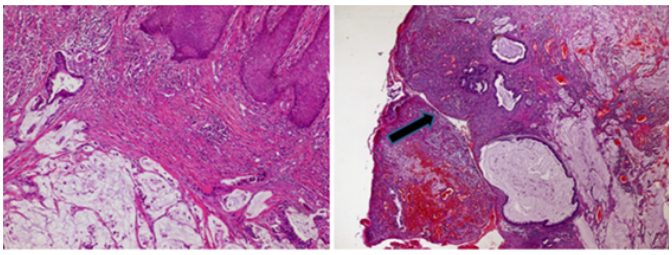

Figure 3 Histological Examination: infiltration of the anal mucosa by adenocarcinoma, the fistula is infiltrated by the adenocarcinoma (arrow).

Discussion and conclusions

The diagnosis of carcinoma originating from a perianal fistula in patients with Crohn's disease remains a significant challenge. While the estimated incidence of carcinoma in patients with Crohn's disease is between 0.3% and 0.7%,[2] the actual occurrence of cancer in this population is reported to be 0.2 /1000 person-years.[3] Both adenocarcinomas and squamous cell carcinomas can occur in patients with CD. As a rare tumor entity, squamous adenocarcinomas are most frequently found in the right-sided colon followed by the rectum and anal duct.[4] Risk Factors contributing to the malignant transformation associated with a perianal fistula include: prolonged duration of Crohn's disease, typically exceeding 10 years; disease onset before the age of thirty; and the presence of fecal diversion.[5]

Chronic inflammation and sepsis, usage of immunosuppressive medications, and viral infections are additional factors that may contribute to this association.[2] Osterman's findings suggest that patients receiving a combination of adalimumab and immunomodulators are at higher risk of cancer development compared to those on adalimumab alone.[6] Church proposes that fistula formation could potentially result from the cancer itself, contrary to conventional expectations.[7]

Given the challenges in diagnosing carcinoma arising from complicated perianal fistulas in patients with Crohn's disease, it is crucial that:

a) Clinicians perform systematic biopsies or curettage during examination under anesthesia to prevent delays in diagnosing low rectal malignancies.

b) Radiologists are attentive to distinguishing findings indicative of cancer versus inflammatory changes. Dynamic contrast-enhanced MRI along with T2 imaging is recommended for these patient.[8]

The gold standard treatment of this entity is abdominoperineal resection.

Mucinous tumors usually don't have a good response to the neoadjuvant chemoradiotherapy and poor survival.[9]

Acknowledgments

None.

Conflicts of interest

None.

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