

Letters to the Editor

Severe perianal disease degenerated to adenocarcinoma. Is close monitoring of long-term perianal disease necessary?

Key words: Crohn's disease. Perianal disease. Perineal adenocarcinoma.

Dear Editor,

Although perianal fistulas are common in the natural history of Crohn's disease (CD), accounting for around 30-50 % of cases, its malignant transformation is extremely rare, from 0.004 to 0.7 % (1).

Case report

We present the case of a 50-year-old male diagnosed with ileal CD in 1978, with long-lasting and difficult-to-manage perianal disease (PAD). Due to the seriousness of PAD, an MRI was performed showing an abscessification of the intraabdominal adenopathic mass (Fig. 1A). Exploratory laparotomy was performed with intraoperative biopsy with histology of low-grade mucinous adenocarcinoma, followed by extensive abdominoperineal amputation with radical cystoprostatectomy by bladder infiltration. Resection of the obturator area, also infiltrated by the tumor, was not possible. The histological piece (Fig. 1B) showed moderately differentiated rectal and perianal adenocarcinoma, implanted in a deep perianal fistula.

The possibility of radiation therapy was ruled out, and the patient is currently receiving cycles of chemotherapy.

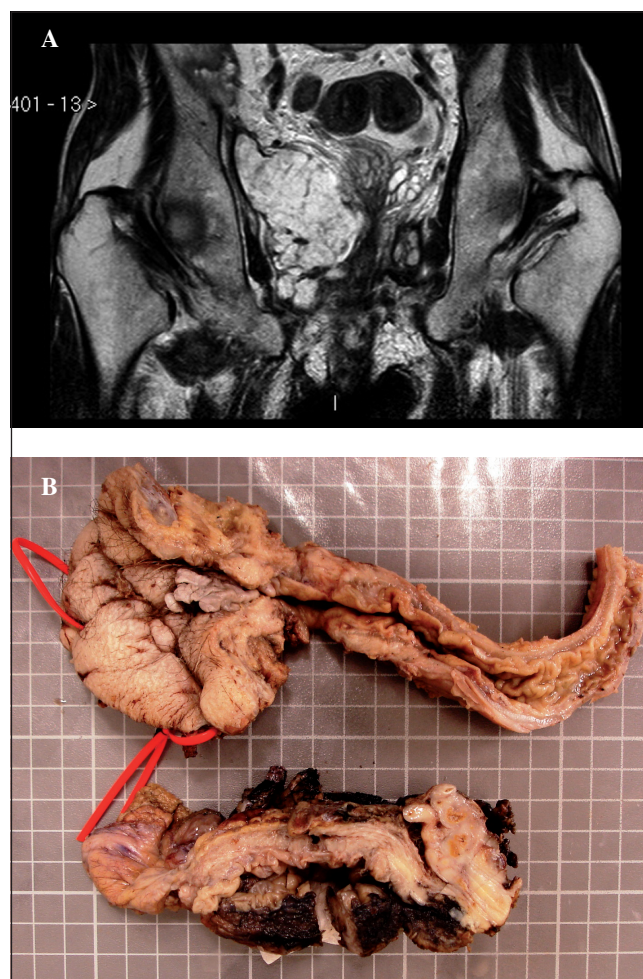


Fig. 1. A. Magnetic resonance imaging. Pararectal mass fistulized to rectum with multiple foci of abscessification and infiltration of the bladder. Its heterogeneity and internal septa suggest a superinfected mucosecretory adenocarcinoma. B. Photo of surgical specimen. Neoplasm located in perianal region related to long-term fistulas.

Discussion

Although the underlying etiopathogenesis is unknown, the most solid causal hypothesis is the constant mucosal regeneration due to chronic inflammation, leading to the malignant transformation of the fistulas or "scar tissue carcinoma" (2). This mainly occurs in patients with long-lasting disease (3), although the duration is shorter in women (4). In our case, and in most of those which have been reported, we do not know the degree of disease activity at the time of diagnosis although our patient did have long-lasting and difficult-to-manage PAD, so constant inflammatory activity could have played an important role in the development of the tumor. Also very important is the long-term immunosuppression to which these patients are generally subject in order to control the disease.

Other possibly related factors are smoking, related to increased mortality (3), and infection by human papillomavirus, which has been associated with the development of an adenocarcinoma in a chronic perianal fistula. (5)

The diagnosis is difficult and generally late, since the clinical manifestations are nonspecific or attributed to the typical symptoms of PAD (1,3).

Some authors (3) recommend annual routine examinations in patients with PAD of more than 10 years duration due to the rapid growth of the adenocarcinoma in a perianal fistula. Due to the inconvenience of the test and also its low prevalence, we believe that it would be indicated in cases of long-term, difficult-to-manage PAD or with clinical worsening.

Results after surgery are poor with high postoperative relapse. There is no clear evidence on the role of adjuvant chemoradiotherapy, although mucinous tumors have a worse response to this therapy and poorer survival (6).

In conclusion, adenocarcinoma implanted on a perianal fistula is an aggressive tumor, with a difficult and late diagnosis, con-

ferring a poor prognosis. Although we believe that routine examination is not indicated, it is important to monitor long-lasting PAD of difficult management or with changes in behavior, because although uncommon, the possibility exists of malignant transformation of perianal fistulas in CD.

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