Mucinous adenocarcinoma arising from a complex perianal fistula: a diagnostic and therapeutic challenge

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DOI: 10.17235/reed.2023.9892/2023
Link: PubMed (Epub ahead of print)

Please cite this article as:

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Dear Editor,

We present the case of a 64-year-old male with a history of recurrent perianal abscesses, which had been surgically treated ten times. The most recent drainage of a perianal abscess occurred two years ago, revealing a swollen area in the left ischiorectal fossa, with purulent discharge and mucus from two external fistula orifice (EFO) at the 3 and 5 o’clock positions in the gynecological position. Drainage was performed, revealing a chronic-looking cavity filled with mucoid material, and an internal fistula orifice (IFO) was identified at the 6 o’clock position along with a trans-
sphincteric mid-level fistula. A biopsy was taken, a loose rubber seton was placed, and the area was irrigated. Pathological findings indicated a possible intestinal mucinous adenocarcinoma (CK20+, CDX2+, TTF1-, CK7+).

The study was completed with colonoscopy, magnetic resonance imaging (MRI), computed tomography (CT) scan (Fig. 1) and positron emission tomography (PET) scan, and the diagnosis was a rectal neoplasia T4N1M0. A diverting colostomy was performed to subsequently initiate neoadjuvant therapy with concurrent chemoradiotherapy using capecitabine. Following neoadjuvant therapy with a partial response, laparoscopic extended abdominoperineal amputation was performed. Pathological findings indicated residual adenocarcinoma (NOS) in the proper muscular layer, ypT2ypN0, N0V0L0, R0.

Discussion
Perianal mucinous adenocarcinoma represents 2% of malignant anorectal tumors (1,2). Currently, its etiology remains unclear, although the literature refers to a possible association with chronic inflammation caused by perianal fistula, inflammatory bowel disease, Lynch syndrome, poorly controlled diabetes mellitus, or tuberculosis (1).

Three diagnostic criteria are required: the fistula should generally precede the carcinoma by ten years, any tumor involvement of the rectoanal canal should only be a secondary extension of the primary tumor, and the internal fistula opening should not be involved (1-3). In our case, the fistula was not present for more than ten years, but upon reviewing the literature, there are multiple reported cases that do not meet this criterion, making it somewhat arbitrary. It is characterized by the presence of neoplastic cells forming mucinous material, and immunohistochemistry highlights CK20+ and sometimes CK7+ (2).

Regarding treatment, it is recommended to perform abdominoperineal amputation with total mesorectal excision and wide resection of perianal lesions (1-5). In this case, the patient is stage III, which involves neoadjuvant radiotherapy and chemotherapy followed by surgical treatment. However, the precise choice of treatment should be made on an individualized basis and discussed by a multidisciplinary tumor committee.
In conclusion, it is crucial to have clinical suspicion of mucinous adenocarcinoma in patients with a chronic anal fistula of several years‘ duration or with the discharge of mucoid material. Furthermore, early biopsies should be performed, initiating surgical treatment +/- radiotherapy and chemotherapy, depending on the stage, leading to a better prognosis and increased survival.

References
Fig. 1. A. Magnetic resonance imaging (MRI) with T2-weighted sequence, where rectal neoplasia with an associated fistulous tract is identified. B. Pelvic computed tomography (CT) scan where an increase in volume is observed in the left lateral wall of the rectum, consistent with a neoplastic process. C. Colonoscopy image where an internal fistula orifice (IFO) is identified 3 cm from the anal margin.