

## Case Report

# Mucinous adenocarcinoma within an anal fistula in a patient with Crohn's disease

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**Received:** 09 September 2019

**Accepted:** 16 October 2019

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## ABSTRACT

Anal mucinous adenocarcinoma is a rare entity, representing approximately 5-11% of all anal cancers. Sometimes it is associated with chronic inflammation of long-standing anal fistula as it may occur in patients with Crohn's disease. In this case, cancer usually goes unnoticed owing to the inflammatory disease; therefore, a degree of uncertainty surrounds initially the diagnosis. We report a case of a 62-year-old man with A2 L3 B3p Crohn's disease who also presents a perianal mucinous pseudocyst that is removed. Histological examination reveals a mucinous pseudocyst. At the two-year follow-up, we observe the recurrence of the cyst within an anal fistula. Scheduled non-oncologic surgery is performed with resection of the cyst and tutoring of the fistula with seton. Histological examination reveals mucinous adenocarcinoma. With no evidence of metastasis in the extension study, the patient is subjected to radical surgery with abdominoperineal resection. After 3 years and 11 months, a follow-up is carried out and there is no evidence of recurrence or distant metastasis. Mucinous adenocarcinoma within an anal fistula is a challenge in its diagnosis and its treatment due to the anecdotal nature of the cases. Given the high local risk of recurrence, radical surgery is recommended, occasionally associating neoadjuvant chemoradiotherapy if the neoplasm is locally advanced.

**Keywords:** Mucinous adenocarcinoma, Crohn disease, Anal fistula, Neoadjuvant therapy

## INTRODUCTION

Crohn's disease is a chronic inflammatory disease with transmural and patchy involvement of the gastrointestinal tract. Between 17-50% of the time, it presents itself with perianal involvement, mainly in the form of a fistula.<sup>1</sup> The development of mucinous adenocarcinoma within these fistulas is highly unlikely and its diagnosis is difficult as it can be masked by the inflammatory disease.<sup>1-4</sup> This tumor is usually locally aggressive, and it has poor prognosis, especially if there is a local recurrence.

## CASE REPORT

We report the case of a 62-year-old man with ileocolic and perianal involvement fistulizing pattern Crohn's disease since 1980, Montreal score A2 L3 B3p. Until now, he has suffered from outbreaks treated with salazopyrin, corticosteroids or mesalazine, being the last one in 2010 and standing since 2011 without immunologic treatment.

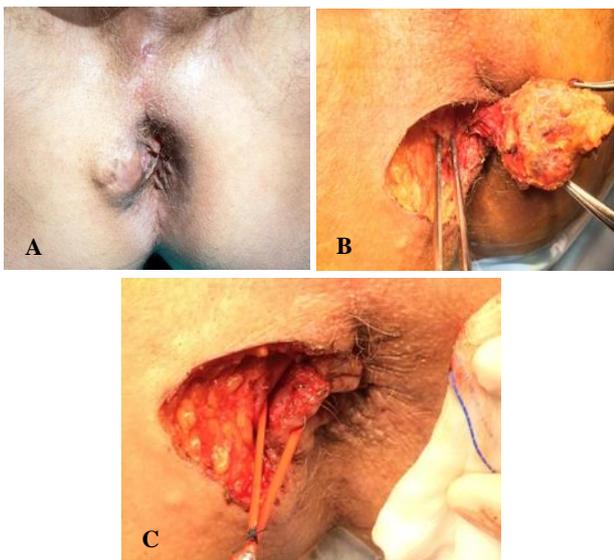
In 2013, he is examined in the Emergency department for two-years perianal swelling with acute super infection; therefore, urgent surgical intervention is performed in

which a mucinous cyst with an abscess is observed and extirpated. Histological examination reveals mucinous pseudocyst.



**Figure 1: Abdominopelvic MR. (A) Sagittal plane; (B) axial plane.**

Multiseptated cyst lesion of 4 cm located in the right posterolateral perianal region which is hyperintense in T2.



**Figure 2 (A-C): Mucinous cyst of 4 cm in the right posterolateral perianal region.**

After a follow-up in consultation, in 2015, recurrence of the tumor is observed, so a magnetic resonance (MR) is requested. A complex transsphincteric fistula with a multiseptated cyst of 4×4 cm (Figure 1) is identified. Scheduled non-oncologic surgery is performed with resection of the cyst and tutoring of the fistula with seton (Figure 2). Histological examination reveals mucinous adenocarcinoma pTxR0. The immunohistochemical technique reports positivity for CK-20, Ck-7 and CDX-2, and negativity for GCDFP-15. These findings point to a mucinous adenocarcinoma within an anal fistula (with a substantial amount of mucin) isolating intestinal mucosa (Ck-20 + and CDX-2 +) and anal glands cells (CK-7+) with pagetoid extension (CK- 20+ y - GCDFP-15).

In the extensive study with abdominopelvic computed tomography, colonoscopy and pelvic MR Crohn's disease with ileitis and ileo-sigmoid-bladder fistula is observed. There is not reported anorectal cancer or distant dissemination data.

On account of the locally aggressive nature of these types of tumors and after evaluating the different therapeutic options, we proceed to direct surgery in 2015. A monobloc abdominoperineal removal was carried out including intestinal and partial bladder resection. A bladder raffia was necessary to close the defect. The anatomopathological result is pT0, N0 (0/28), PN-, L/Vi-, MRC-, R0. The patient has a good post-operative evolution being discharged on the 7th postoperative day.

In 2017, he suffered several urological tract infections secondary to new ileo-bladder fistula. The MR described terminal ileitis with a fistula between the bladder and the previous intestinal anastomosis. Ileocectomy with anastomotic resection was performed and treatment with Azathioprine 150 mg per day was established after the surgery. He is asymptomatic at present. No local or distant neoplastic disease is observed at 3 years and 11 months of follow up after the abdominoperineal, intestinal and bladder resection.

## DISCUSSION

Anal cancer represents a minority of 2-3% among neoplasms of the gastrointestinal tract.<sup>2,5</sup> Three types of anal canal cancer have been described: intestinal adenocarcinoma, squamous cell carcinoma, and mucinous adenocarcinoma. Only 5-11% of anal cancers correspond to anal mucinous adenocarcinoma.<sup>2,5</sup> There is no consensus about its origin, pathogenesis or biological behavior. Nevertheless, there is an increase in its incidence when there are chronic concomitant inflammatory conditions such as chronic anal fistula in a patient with Crohn's disease. In 1934, Rosser et al, was a pioneer in the description of perianal carcinoma arising from chronic fistula.<sup>3,5</sup> Some authors, like Traube et al, have postulated that this association is due to the need for constant regeneration of the mucosa.

It is estimated that mucinous adenocarcinoma of the anal canal has an incidence of 0.7% in patients with Crohn's disease.<sup>2,5,6</sup> If it is associated with chronic anal fistula, the average age at which it occurs is between 43 and 53 years old. The mean duration of the fistula in a patient with Crohn's disease before the diagnosis of cancer is 10-20 years.<sup>5</sup> The risk factors for malignization of the fistula are the degree of associated inflammation, the location, and of least importance the time of evolution.<sup>1</sup> Cancer often goes unnoticed owing to the inflammatory disease itself so a high diagnostic suspicion is necessary, with biopsy being the gold standard for diagnosis.

Given the anecdotal nature of the cases, there is no standardized treatment for this specific type of tumor. On

one side, direct surgery is suggested in local-early stage tumors; on the other side, if the tumor is locally advanced, some case series like Gaertner et al, recommend neoadjuvant chemoradiotherapy before surgery. Occasionally, on account of the extended resection to the levator ani muscle, it is advisable to perform a pelvic floor reconstruction.<sup>3</sup> Radiotherapy as the only treatment is an alternative in fragile patients who cannot undergo radical surgery.

In this case, it is a localized tumor that could have been treated with the extirpation of the tumor and active surveillance. Nonetheless, in consensus with the patient and given their life expectancy and the local recurrent nature of the tumor, it was decided to perform a more aggressive treatment with oncologic direct radical surgery.

## CONCLUSION

In conclusion, mucinous adenocarcinoma of the anal canal is a rare type of tumor in which a high diagnostic suspicion is necessary, especially in patients with concomitant diseases such as Crohn's disease because it may be masked. Given its local aggressiveness, it is essential to accomplish surgery with negative margins being possible to use chemoradiotherapy in a neoadjuvant way.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Riaza Montes M, De Andrés Olabarria U, Badiola Bergara I, Portugal Porrás V, Ibañez Aguirre FJ. Mucinous adenocarcinoma within an anal fistula in a patient with Crohn's disease. *Int Surg J* 2019;6:4121-3.