



Malignant transformation of a chronic anorectal fistula



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Malignant transformation of a chronic anorectal fistula

Up to date, only a small number of carcinomas arising from a chronic anorectal fistula have been described in medical literature, especially in patients without Chron's disease.

A 72-year-old man with a 6-year history of discharging perianal sinus without Crohn's disease arrived at our institution. He had previously undergone three surgical procedures in other institutions for incision and drainage of recurrent perianal abscesses.

Our therapeutical approach was to drain the two abscess cavities, perform a fistulectomy, and biopsy the fistula tissue. Anatomopathological examination of the specimen revealed a mucosocerrnig adenocarcinoma arising from the fistula tract. We decided to perform an abdominal perineal resection. The two-year oncological follow-up is negative.

In conclusion, it is clear that the diagnosis of mucinous adenocarcinoma occurring in perianal fistula is difficult, particularly in patients without any risks or predisposing factors. Wide resection of the tumor with Miles's procedure still represents the surgical treatment of choice and may provide a good long term outcome in localized disease

KEY WORDS: Cronic anorectal fistula, Malignant transformation, Mucinous adenocarcinoma

Introduction

Malignant transformation of a chronic perineal fistula is a rare entity in patients without Crohn's disease; it consists of a specific neoplastic involvement of the fistulous tract in the absence of rectal mucosal carcinoma. Since symptoms are usually attributed to the fistula itself and the growth of tumor is normally slow, the diagnosis of adenocarcinoma within a chronic fistula results difficult and may be delayed.

Up to date, only a small number of carcinomas arising from a chronic anorectal fistula have been described in medical literature, in particular for patients without Chron's disease. Rosser first reported seven cases in 1934, and, more recently, Ong et Yang described small series of 4 and 3 patients, respectively ^{1,2,4}. However, the rarity of this tumor and the lack of sufficient patients for controlled trials have led to a lack of any consensus regarding appropriately diagnostic and treatment strategies.

Even rarer are cases of chronic fistulas hiding mucinous adenocarcinomas, an uncommon cancer that accounts for approximately 3-19% of all anal carcinomas ^{1,2}.

Case Report

A 72-year-old man with a 6-year history of discharging perianal sinus without Crohn's disease arrived at our institution. His past medical history included right

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saphenectomy for varicose veins, atrial fibrillation, and arterial hypertension. He had previously undergone three surgeries at other institutions for incision and drainage of recurrent perianal abscesses. A lot of technique are described for treatment of perianal abscess fistula related¹⁰. A major problem after this procedure, often executed in day surgery regimen, is the stenosis of the anal canal^{11,12}.

On clinical examination of perianal zone in lithotomy position, two external openings were observed in medial posterior and lateral right position. Anorectal digital examination showed chronic inflammatory tissue. The inguinal lymph nodes were not palpable on both sides. Pelvic MRI showed the presence of two abscesses: the bigger one was intersphincteric with a horseshoe shape image of 78x51x28 mm communicating with the anal canal. The other was located in lower lateral right zone 40x12x27 mm with an external communication (Fig. 1). Colonoscopy showed no mass or stenosis, the aspect of colonic mucosa was normal and multiple biopsies confirmed the absence of any inflammatory bowel disease (IBD). Our therapeutical approach was to drain the two abscess cavities, perform a fistulectomy, and biopsy the fistula tissue.

Anatomopathological exam of the specimen revealed a mucoscercnig adenocarcinoma arising from the fistula tract.

An MRI for rectal cancer staging was performed after the drainage of abscesses cavity, showing a localized tumor with negative perirectal lymph nodes. Thoraco-abdominal CT was negative for local invasion and/or metastasis. CEA and CA 19-9 levels were elevated: 12.8 mcg/L and 79.8 U/mL, respectively.

We decided to perform an abdominal perineal resection (Miles's procedure). The postoperative course was uneventful and the patient was discharged 12 days after surgery. Two-year oncological follow-up is negative.



Fig. 1

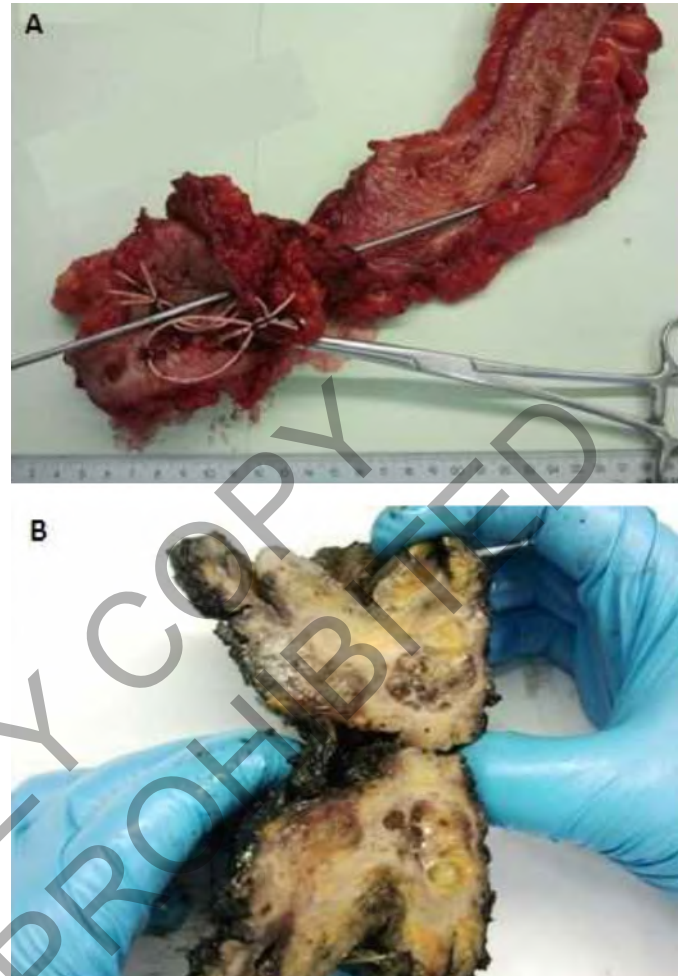


Fig. 2

The specimen showed a mucinous adenocarcinoma stage T3 N0 M0 R0. The pathologists described the surgical specimen as composed by sigma, rectum, and anal canal with a total length of 25 cm; the anal presented a skin lozenge of 9x4 cm. Not significant macroscopic changes were found at opening of the bowels. Instead, the anal canal was difficult to explore and was transformed in a mass of significantly increased consistency of cm 10x7x5. Numerous mucoid-material leaking fistulas paths with rubberized wires were observed on skin. On sectioning, this mass showed multicystic lesion with poorly distinguishable boundaries (Fig. 2).

Microscopically the lesion was constituted by infiltrating mucinous glands, which invaded perianal soft tissues and determining lakes of mucin and granulation tissue in some areas. In addition, the neoplastic glands determined burrowing on perianal skin (Fig. 3).

Immunohistochemical profile of the neoplastic glands was AE1/AE3+, CK7+/-, CK20+, CK8/18+, 34betaE12-/-, E-Cadherin+, CDX2+; anctin and PSA were negative.

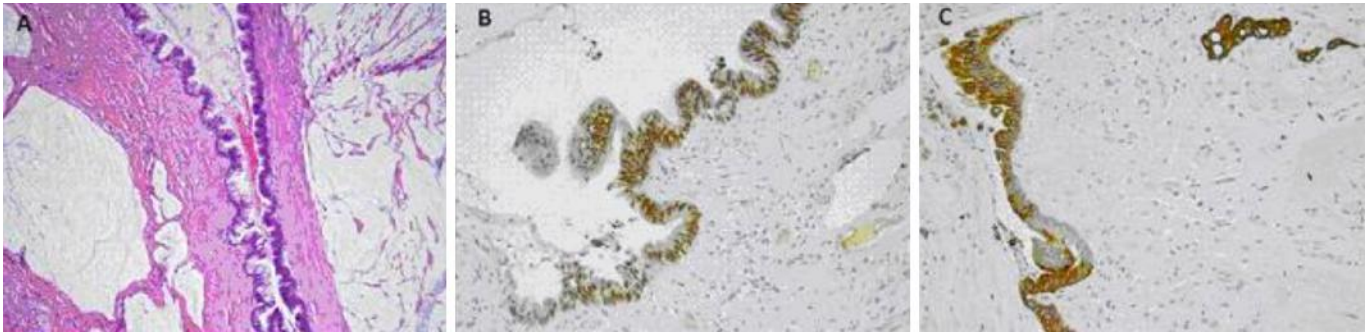


Fig. 3

Discussion

Chronic anal fistula is a common condition in coloproctology; however, perianal adenocarcinoma associated with fistula in-ano is extremely rare. Only a few articles have described this entity in literature; a paper by Bo-Lin Yang et al, reported 10 cases in the last 20 years in the English literature ².

Pathogenesis of this disease remains controversial. Some authors suggest that adenocarcinoma of anal canal may originate from rectum, fistula-in-ano or anal glands. Two major theories have been suggested to explain the coincidental mutation between a chronic anorectal fistula and the development of a carcinoma. Traube et al hypothesized a key role for scar tissue; chronic inflammatory changes due to a long standing fistula-in-ano are probably the origin of the mucinous adenocarcinoma development, triggering the degeneration of scar tissue into cancer ⁶.

Conversely, Church et al suggested that the cancer itself may cause enterocutaneous fistulas ⁷.

Nevertheless, in most cases, the fistula pre-existed long-time before any sign of carcinoma appears, making this theory less acceptable.

Rosser proposed a set of diagnostic criteria for carcinoma originating from the anal fistula: the fistula should usually antedate the carcinoma of at least 10 years; the only tumor present in the rectum or anal canal should be secondary to direct extension from the carcinoma in the fistula; and the internal opening of the fistula should be into the anal canal and not into the tumor itself ⁴. Our patient presented all the above criteria except the first one, since the anal fistula had been present for only 6 years. A further clue to the diagnosis of mucinous aspect is the presence of free globules of mucin, which lie away from areas of granulation tissue within a fistulectomy specimen.

The role of neoadjuvant and adjuvant chemo-radio therapy in the treatment of mucinous adenocarcinoma perianal fistula associated is not clear and, in the recent reports the benefit is not clearly demonstrated ²⁻⁵.

Preoperative staging should include a thoraco-abdominal CT; this is useful not only for detecting distant metas-

tases, but also for assessing the local extension of the disease. MRI has been evaluated for use in the preoperative staging of low rectal tumors and has been shown to be useful in assessing the presence of sphincteric invasion and of surrounding structures ^{8,9}. CEA level assessed preoperatively appeared to be a useful marker of metastases in patients with anal canal adenocarcinoma.

Conclusions

In conclusion, it is clear that the diagnosis of mucinous adenocarcinoma occurring in perianal fistula is difficult, particularly in patients without any risks or predisposing factors. Biopsy of fistulous tracts and perianal abscesses to assess histology are thus paramount in the early diagnosis and subsequent treatment of these tumors. Wide resection of the tumor with Miles's procedure still represents the surgical treatment of choice and may provide a good long term outcomes in localized disease.

Riassunto

La trasformazione maligna di una fistola perineale cronica è un'entità rara in pazienti che non siano affetti da morbo di Crohn. Fino ad oggi, solo un piccolo numero di carcinomi derivanti da una fistola anorettale cronica sono stati descritti in letteratura.

Un uomo di 72 anni con ascesso perineale e una storia di fistola perineale nota da circa 6 anni e senza arriva alla nostra attenzione. In precedenza aveva subito tre interventi chirurgici in altre istituzioni per incisione e drenaggio di ascessi perianali ricorrenti.

Il paziente è stato sottoposto ad intervento di drenaggio, fistulotomia parziale con apposizione di setone lasso e biopsia del tessuto. L'esame anatomopatologico del campione ha rivelato un adenocarcinoma mucosocernente derivante dal tramite fistoloso.

Il paziente è stato sottoposto a ad amputazione addominale perineale. Il follow-up oncologico a due anni è negativo. La diagnosi di adenocarcinoma mucinoso derivante da fistola perianale è rara, soprattutto nei pazien-

ti senza fattori di rischio. Un'ampia resezione del tumore con la procedura di Miles rappresenta ancora il trattamento chirurgico di scelta permettendo di fornire un buon risultato a lungo termine, in termini di recidiva e sopravvivenza, nella malattia localizzata.

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