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Mucinous adenocarcinoma associated with chronic suppurative hidradenitis: Report of a case and review of the literature

Natalia Mukai[a], Lílian Vital Pinheiro[a], Maria de Lourdes Setsuko Ayrizono[a], Guilherme Cardinali Barreiro[b], Paulo Kharmandayan[b], Mariana Hanayo Akinaga[b], Adriano Mesquita Bento[b], Carlos Augusto Real Martinez[a], Rita Barbosa de Carvalho[c], Marc Ward[d], Cláudio Saddy Rodrigues Coy[a], Raquel Franco Leal[a,*]

[a] Coloproctology Unit, Department of Surgery, University of Campinas, UNICAMP, Campinas, Sao Paulo, Brazil
[b] Plastic Surgery Unit, Department of Surgery, University of Campinas, UNICAMP, Campinas, Sao Paulo, Brazil
[c] Department of Pathology, Gastrocenter, University of Campinas, UNICAMP, Campinas, Sao Paulo, Brazil
[d] University of Chicago Medical Center, Chicago, IL, United States

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ABSTRACT

INTRODUCTION: Chronic suppurative hidradenitis (CSH) is a benign condition that can affect the perineal region and often leads to the formation of abscesses and fistulas. It is rare for CSH to undergo malignant degeneration into mucinous adenocarcinoma.
PRESENTATION OF CASE: We report a case of a 55-year-old male patient with perineal CSH who suffered worsening long-term pain despite multiple surgical procedures to alleviate his symptoms. Pelvic magnetic resonance imaging (MRI) showed multiloculated cystic lesion on the left side wall of the distal rectum with gluteal extension. Pathological examination revealed mucinous adenocarcinoma. The patient underwent an abdominoperineal resection (APR) of the rectum with cutaneous muscle flap reconstruction. Although histopathological sections showed clear margins, the tumor recurred 6 months following surgery.
DISCUSSION: Perineal mucinous adenocarcinoma arising in a patient with CSH is an extremely rare condition. This diagnosis is often difficult, due to the paucity of signs of malignant degeneration as well as the rarity of the disease itself. Surgical resection of the lesions is a well-established approach. In this case, diagnosing the tumor at such a late stage likely compromised his outcome.
CONCLUSION: Malignant degeneration to mucinous adenocarcinoma must be suspected in patients with a history of long-term CSH. In such cases, local biopsies and a radiological examination, such as MRI can help in the diagnosis.

1. Introduction

Chronic suppurative hidradenitis (CSH) is a condition where there is an infection of apocrine glands and surrounding soft tissues of the skin and adjacent structures. This often leads to the formation of fistulas and abscesses. When there is perianal involvement with extensive infection, there is an indication for wide resection of the affected area with primary or secondary closure. One of the rare complications is malignant degeneration, squamous cell carcinoma being the most common [1–5].

Mucinous adenocarcinoma in the perianal region is an uncommon condition and constitutes only 3% to 11% of all perianal cancers. This type of tumor can arise from anal glands and often presents itself as abscesses and/or fistulas, which compromises its early diagnosis [6–8]. The pathogenesis of those perianal tumors is still unknown and there are no histopathologic methods to demonstrate the primary association between the lesion and apocrine glands or anal glands. There are few reports in the literature of patients with perianal CSH who develop mucinous adenocarcinoma, making it an extremely rare association. Our aim here is to present a patient with perianal CSH who developed mucinous adenocarcinoma and to also review of the cases already published in the literature.

2. Presentation of the case

A 55-year-old male patient was followed at our Coloproctology Outpatient Clinic at the University of Campinas for the past 8 years. He was sent to our Clinic because of multiple perianal fistulas that began at 12 years of age requiring several surgical procedures

* Corresponding author at: João Lopes Vieira Street, n° 108, 61,Campinas, Sao Paulo 13087-734, Brazil.
E-mail address: rafranco.unicamp@gmail.com (R.F. Leal).

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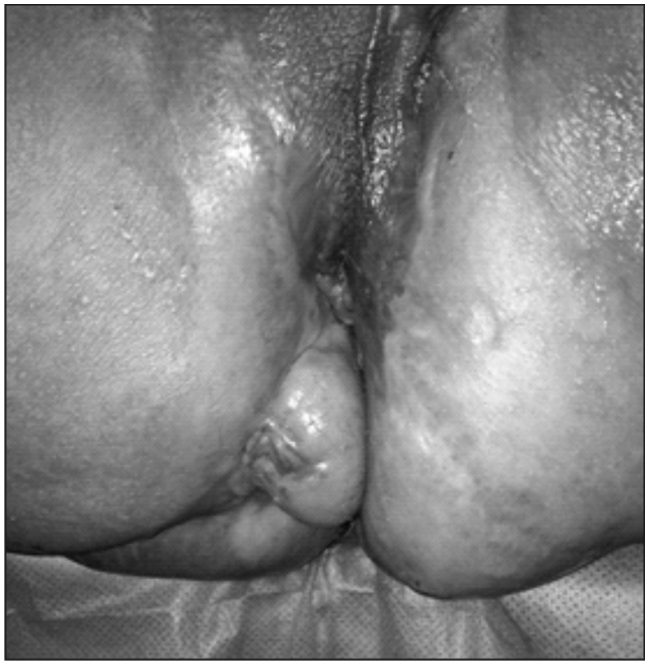

Fig. 1. Perineal Chronic Suppurative Hidradenitis, and scars from previous surgical resections. Patient in lithotomy position.

(excision of the fistula tracks and abscess drainage). He was diagnosed early with CSH and throughout his treatment he reported progressive worsening of perineal and perianal pain, alteration in stool frequency, and mucinous discharge from his fistulas. Physical examination showed multiple deformities of the perianal region, scars, the presence of perineal skin ulcers near the posterior anal canal, mucinous discharge from fistulas, and a bulge in the perineal region (Fig. 1). Pelvic MRI revealed a multiloculated cystic lesion in the left side wall of the distal rectum, measuring $5.2 \times 8.5 \times 6.2$ cm (Fig. 2).

The lesion occupied the intersphincteric space with caudal extension bulging the elevator muscle of the anus bilaterally. There were fistulas between the lesion and the gluteal skin surface as well as the intergluteus groove with paths going both posteriorly and anteriorly to the presacral space bordering the anal elevator muscle in the right. There was a marked hyper signal in T2 and skin scar retraction.

The patient underwent an examination under analgesia with lesion biopsies. This revealed mucinous adenocarcinoma with a moderately differentiated pattern in the perianal region and associated inflammatory reactions near the epithelium. The skin had pseudoepitheliomatous hyperplasia and dermal fibrosis with the presence of 70% of mucinous component in the sample. Computed Tomography (CT) scan did not reveal distant metastasis.

Given these findings, the patient elected to undergo an extended abdominoperineal resection (APR) with coccyx and partial sacrum resection, definitive colostomy, and pelvic lymphadenectomy. In the 12th post-operative day, a bilateral pedicled anterolateral thigh flap was performed by the Plastic Surgery team of the University of Campinas Clinical Hospital to cover the perineal defect. The surgical aspects are shown in Fig. 3. Unfortunately, he developed abdominal suture dehiscence with evisceration, making it necessary to perform a peritoneostomy. He also required cleaning of the perineal wound in the operating room and hyperbaric chamber therapy due to flap infection. He was discharged on post-operative day 60, in good condition and with good healing of the perineum. Histopathological sections confirmed a large mucinous adenocarcinoma with lymphatic and perineural invasion with clear margins and the absence of lymph node metastases (Fig. 4).

Five months after surgery, he presented with a hardened and painful lesions in the previous flap in the perineal area. A new biopsy was performed confirming tumor recurrence, although the margins of the surgical specimen were clear. He elected not to pursue any other therapies due to the large extension of the lesion and his poor clinical condition. He died 6 months after the operation.

3. Discussion

Perineal mucinous adenocarcinoma is a rare clinical condition, and only 5 cases (4 male) associated with CSH were described in the literature (Table 1). They were between 40 and 60 years-old, and they had developed clinical presentation of CSH between the ages of 3 and 15 years. Ariwa [9] performed histochemical analysis which suggested that the origin was from the anal glands. In the present case report, this analysis was not performed for technical limitations. However, there were no significant alterations in the histologic sections of rectal mucosa from the surgical specimen, suggesting that the origin of the tumor may be associated with CSH. Indeed, the mucinous adenocarcinoma was found in the perineal and perianal subcutaneous tissue, near the fistulae tracks of the CSH. In all cases of the literature, malignant degeneration was diagnosed through lesion biopsies. The surgical approach was

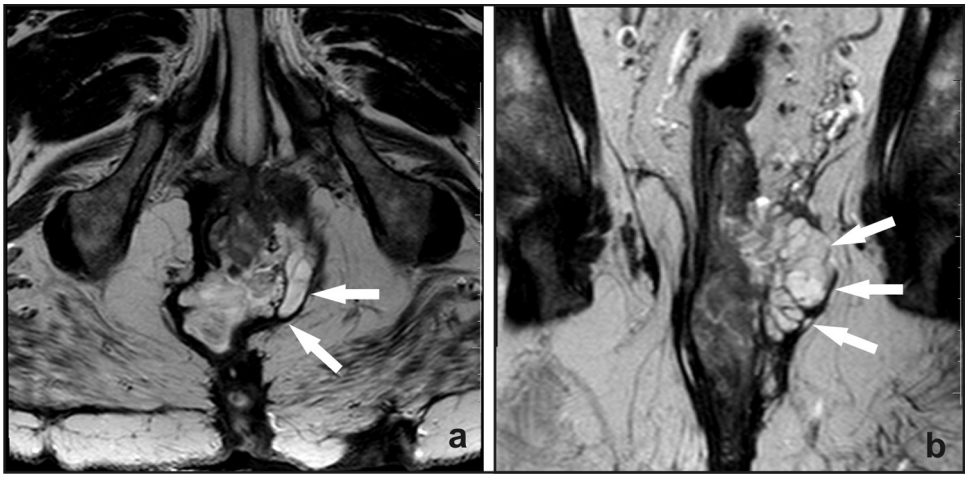

Fig. 2. Magnetic Resonance Imaging shows hyper signal in T2, suggesting mucinous adenocarcinoma. (a) Transverse section (b) Sagittal section.