Adenocarcinoma secondary to hidradenitis suppurativa Case report and literature review

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INTRODUCTION

Hidradenitis suppurativa (HS) is a chronic, recurrent, and potentially disabling systemic inflammatory autoimmune disease that originates in the hair follicle in areas with a high density of apocrine glands.[1]

According to the European series, worldwide incidence ranges from 1 to 4%.[2] The 2018 national registry documented 253 cases in Argentina.[3] The female-to-male ratio is 3-5:1. The prevalence of the condition is significantly higher in women between the ages of 20 and 40 years, as well as in men older than 45 years. In contrast, the incidence in childhood is reported to be only 2-3%.

Genetic load, obesity, and smoking are described as predisposing factors.[1] The diagnosis is made by clinical examination and ultrasound. In this entity, clinimetry or severity assessment is of fundamental relevance. Several systems have been described for staging the disease: The Hurley staging system, the Sartorius score, the Physician Global Assessment (PGA), the Clinical Response in HS (HiSCR-Hidradenitis Clinical Response), and the Hidradenitis Suppurativa Severity Score System (IHS4-International HS 4). The most commonly used in our country are the Hurley staging system and the Sartorius score (Fig. 1). In Argentina, the Hurley staging system and the Sartorius score are the most commonly utilized staging systems (Fig. 1).

An infrequent complication of HS is the development of neoplasms. Squamous cell carcinoma is the most prevalent type, while mucinous adenocarcinoma (MCA) is extremely infrequent, with no more than eight cases documented in the international literature.[5]

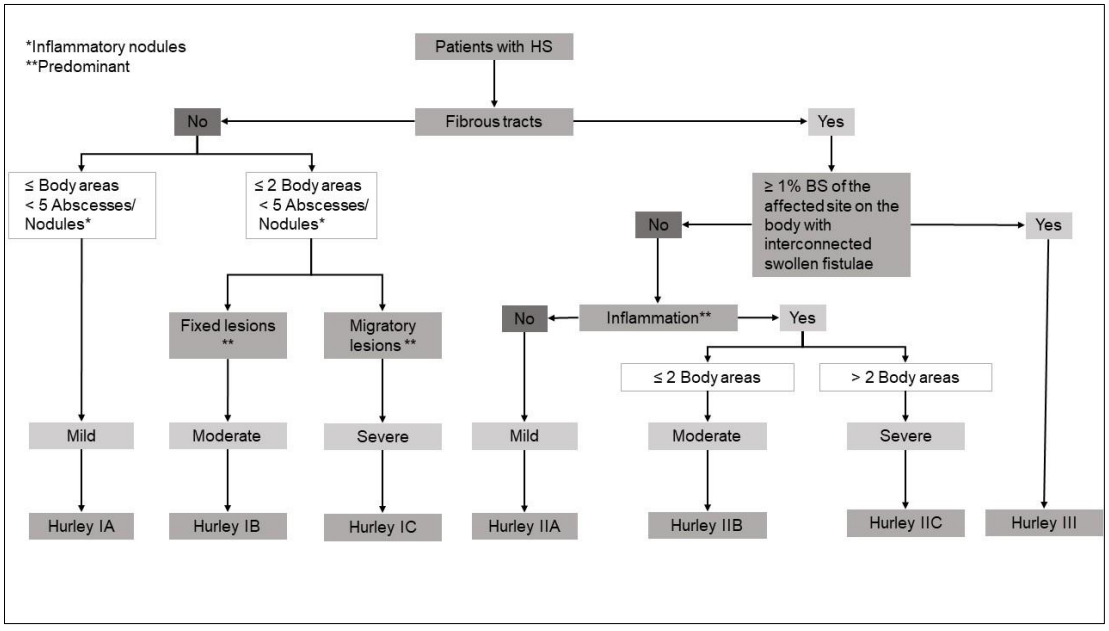

Figure 1. *Hurley's classification of hidradenitis suppurativa (HS). BS: Body surface area.*

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CASE

We present the case of a 46-year-old female patient who has been diagnosed with HS for 10 years, with a smoking history amounting to approximately 20 pack-years. Despite undergoing various treatments, both local and systemic, there was no satisfactory response until Adalimumab 40 mg was administered subcutaneously every 15 days from 2013 to 2014. The treatment resulted in a positive response, as evidenced by the absence of secretion and no progression. However, the sequelae of this pathology had escalated to a Hurley III and Sartorius 100 grade (Fig. 2A). Subsequently, due to difficulties in obtaining the immunomodulator, the patient interrupted treatment and follow-up for two years. In 2016, she returned to the consultation with a gluteal tumor with substantial mucinous secretion (Fig. 2 B).
The case was approached via a multidisciplinary team, and a biopsy was conducted. The biopsy revealed mucus-secreting adenocarcinoma. Colorectal pathology was ruled out with a complete colonoscopy.

The lesion was resected, ensuring oncologic margins, resulting in the removal of the right buttock, a portion of the left buttock, the anococcygeal raphe, and a segment of the homolateral levator ani (Fig. 2C). A perforation of the posterior rectal wall was identified, which was repaired and protected with a sigmoid loop colostomy.
The histopathology revealed mucus-secreting adenocarcinoma with mucin lakes and atypical glandular cells.
After ruling out the presence of disease, the colostomy was closed 10 months later. However, due to factors inherent to the health system and a lack of adherence, follow-up was deferred for one year. After this period, a voluminous local recurrence was found (Fig. 2D), which was associated with tomographic images compatible with metastases. However, a PET scan did not show uptake at that level. Due to a deterioration in quality of life and subocclusive symptoms, another sigmoid loop colostomy was performed. The case was discussed in a multidisciplinary oncology meeting, and adjuvant treatment was recommended. However, the patient did not accept the proposal.

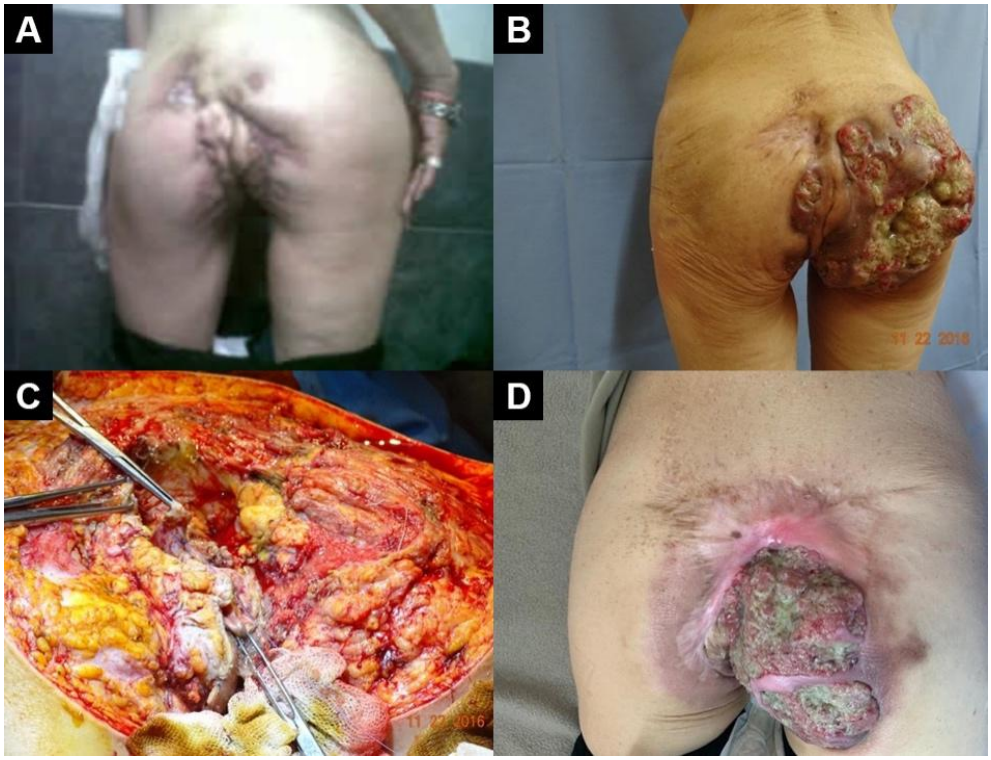

Figure 2. *Hidradenitis suppurativa.* ***A.*** *Disease classified as Hurley III after 10 years of progression and a poor response to multiple treatments.* ***B.*** *A large gluteal tumor with abundant mucinous discharge developed after 2 years without follow-up. A biopsy revealed a mucus-secreting adenocarcinoma.* ***C.*** *Large local resection.* ***D.*** *Large local recurrence can be observed one year after surgery.*

Given the progression of the disease without treatment and the progressive deterioration in the quality of life, life, a new surgical procedure was undertaken. This procedure involved a posterior pelvic exenteration, and the reconstruction of the defect using a a vertical rectus abdominis myocutaneous flap 41 months after the initial resection (Fig. 3, A and B). This resection revealed the same tumor lineage, and given the positive margin in the right ischial bone, oncologic treatment was deemed necessary and partially implemented with capecitabine.
Four months later, a biopsy revealed disease progression, which was treated locally with curettage for one year (Fig. 3C). Concurrently, tomographic lesions consistent with pulmonary metastasis were identified.

After one year, the systemic stability of the oncologic disease was documented. However, alterations in the quality of life due to local growth (Fig. 3D), associated with difficulties in hygiene and posture, resulted in a palliative partial resection. One month later, adrenal metastasis was observed, and six months later, in the context of an impregnation syndrome of one month of evolution, the patient died.
Notably, during the six-and-a-half-year period in which the oncological disease was present, even with a poor quality of life, the performance status remained at 0 until the final month of life.

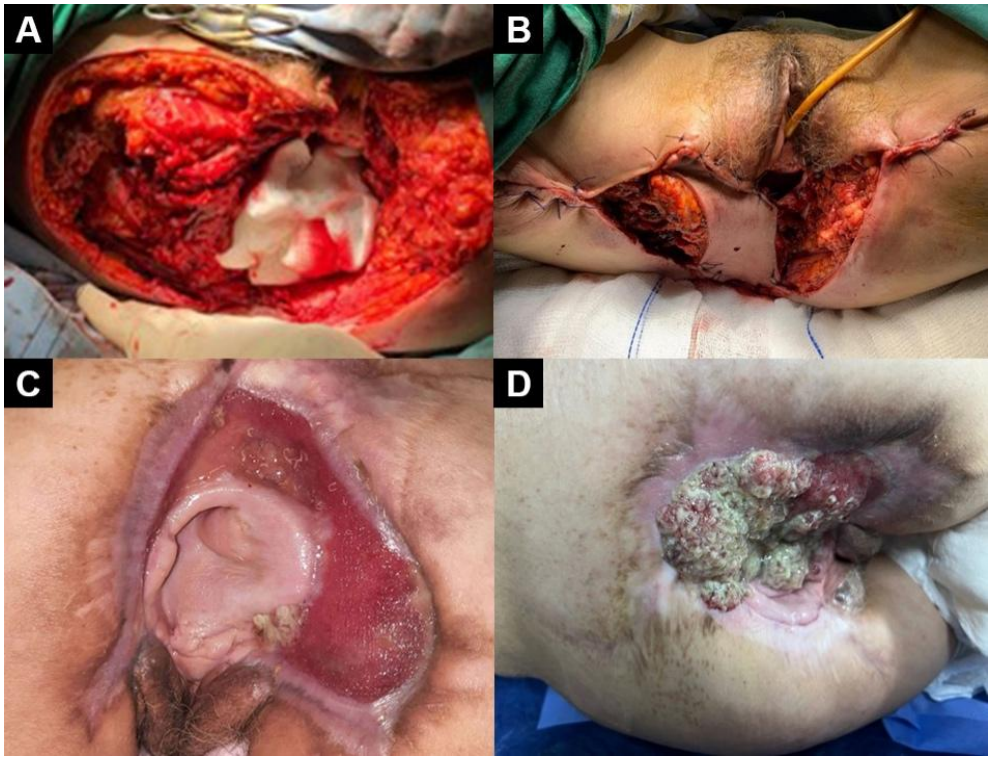

Figure 3. *Local recurrence of mucus-secreting adenocarcinoma.* ***A.*** *Posterior pelvic exenteration of the recurrent mucus-secreting adenocarcinoma.* ***B.*** *Partial defect reconstruction using a vertical rectus abdominis myocutaneous flap (VRAM).* ***C.*** *Disease progression 4 months after exenteration, treated with curettage for 1 year.* ***D.*** *Recurrent tumor.*

DISCUSSION

MAC of the perineum/genital area was first described by Rosser[6] in 1934, who identified it in cases of long-standing fistulas. In the few cases published secondary to HS, patients have reported a history that has persisted for 10 or more years.[4] The most prevalent tumor identified in these cases is squamous cell carcinoma. However, cases of MAC have been documented.

In 2022, Tekbas et al.[7] described 20 cases of neoplasms of the perianal region, including only 4 of MAC, without distinguishing their origin. In 2016, Mukai et al.,[4] in a systematic review, found only 5 cases of MAC secondary to HS. Two case reports have also been published. The cases mentioned, and this report, confirm that there are currently 8 documented cases published in indexed journals.[4,5,8,9]

According to Kim et al,[8] the initial step in determining the origin of an adenocarcinoma in an anal fistula is the exclusion of a rectal neoplasm as the primary source. Moreover, the presence of a fistula must precede the development of carcinoma, and the primary orifice must be located within the anal canal and not within the tumor itself. Following these principles, it can be concluded that the present case is attributable to the malignization of fistulous tracts secondary to HS.

Concerning the progression of this chronic complication, it is noteworthy to mention the concept outlined in the consensus on HS of the Argentine Society of Dermatology,[1] which introduces the term "window of opportunity." This term refers to when interventions to regulate inflammatory activity may be most effective.

This window is often lost due to diagnostic delays in HS caused by issues inherent to the pathology, the healthcare system, and the patients themselves.[10]

The treatment for adenocarcinomas in the perineal and perianal regions depends on their location, extent, and origin. Treatment options range from wide local resections to abdominoperineal resections. In the present case, a progressive approach was chosen after a discussion among the multidisciplinary team, respecting the patient's autonomy.

Lastly, it is important to highlight the role of mental healthcare for patients with this disease, as the sequelae are usually related to depression, shame, stigmatization, and irritability. These sequelae can produce isolation and aversion to intimate relationships, and often exceed the patient's will to comply with the best treatment.[11,12]

CONCLUSION

MAC originating in HS is a very infrequent complication. Surveillance of patients with long-standing disease is essential, and the development of a neoplasm should be suspected in the presence of disease progression despite treatment, or torpid evolution after a previous favorable response.

Multidisciplinary teams are recommended for the comprehensive management of patients with HS, including mental health care and treatment of possible addictions.

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