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Case report

Squamous cell carcinoma of the vulva arising in the setting of chronic hidradenitis suppurativa: A case report

Rekawek Patricia [a], Mehta Shailja [a], Andikyan Vaagn [b], Harmaty Marco [c], Zakashansky Konstantin [b]

[a] Department of Obstetrics, Gynecology and Reproductive Science, Icahn School of Medicine at Mount Sinai, New York, NY, United States
[b] Department of Gynecologic Oncology, Icahn School of Medicine at Mount Sinai, New York, NY, United States
[c] Department of Plastic Surgery, Icahn School of Medicine at Mount Sinai, New York, NY, United States

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1. Introduction

Hidradenitis suppurativa (HS) is a chronic and recurrent inflammatory follicular occlusive disease involving the follicular portion of folliculopilosebaceous units (FPSUs) of the skin (Jemec and Hansen, 1996). The pathophysiology of the disease involves follicular occlusion, follicular rupture, with an associated immune response that leads to the formation of abscesses, sinus tracts, scarring, and ultimately severe diffuse involvement (Pena et al., 2015). Associated factors include obesity, smoking, genetic susceptibility, and mechanical stresses on the skin. Most commonly affected areas of the body are those bearing apocrine-glands, such as the axilla, inguinal area, and anogenital regions. In females, it usually targets the gluteal and pudendal areas and is characterized by painful nodules, abscesses, fistulas, sinus tracts, comedones and scarring, which may lead to severe functional and psychological impairment (Alikhan et al., 2009).

It has been theorized that these chronic insults to skin can lead to proliferative epidermal changes as well as malignancy (Donsky and Mendelson, 1964; Anstey et al., 1990). Squamous cell carcinoma arising in hidradenitis suppurativa/acne inversa (HS/AI) is rare and more commonly found in men (Maclean and Coleman, 2007; Mendonça et al., 1991). A review of all published cases of SCC showed that 48% of these patients died within 2 years of SCC recognition (Maclean and Coleman, 2007). To our best knowledge, it has been reported in at least 64 cases (Alikhan et al., 2009).

We report a case of a woman who developed squamous cell carcinoma of the vulva in the setting of chronic, long-standing hidradenitis suppurativa.

2. Case presentation

Patient is a 61 year old P3 postmenopausal female who presented to the ED with a left labial mass and vulvar pain, worsening over the past 2 months. Patient's past gynecologic history is significant for chronic vulvar hidradenitis suppurativa (Hurley Stage III) and obesity (BMI 31). Other comorbidities included well controlled hypertension.

On Image 1, the initial external pelvic exam is shown. The patient's external genitalia showed areas affected by severe, chronic hidradenitis suppurativa with marked hyperkeratosis with darkened areas of fibrosis. Most significantly, her left labia majora contained a large, friable 10 × 7 cm mass with white scaling and verrucous appearance. Patient underwent a vulvar biopsy which revealed a focus of a well-differentiated, keratinizing squamous cell carcinoma. MRI of the pelvis revealed diffuse, nodular skin thickening along the pelvic folds, up to 2.3 cm in depth with subcentimeter, rim-enhancing nodules that may represent abscesses with no fistula or intrapelvic extension. Additionally, the patient had a 3.1 cm diffusely thickened endometrial stripe and a fibroid uterus. The findings were noted to be compatible with chronic hidradenitis. Patient had a subsequent PET scan which was negative for distant metastasis.

Patient underwent a radical hemivulvectomy, bilateral groin nodal dissection and endometrial curettings which left a large defect in the perineum measuring 12 × 6 cm (Image 2). Reconstruction of perineum was performed using perforator flap followed by two vascularized fasciocutaneous flap closures of thigh donor site. At the completion of the procedure, 4 Jackson-Pratt drains were placed (2 in bilateral inguinal node dissection and 2 in reconstructive flap).

In this obese patient undergoing vulvar reconstruction with superimposed hidradenitis suppurativa, the postoperative course was further complicated by surgical site infection requiring multiple debridement procedures and prolonged courses of IV antibiotics (Vancomycin, Piperacillin, Tazobactam, Ceftrizone, Flagyl) with twice daily dressing changes with Dakins solution (Image 3). After 2 weeks of therapy the surgical site was minimally healed with persistent swelling, deep fissures and purulent drainage noted. The patient was started on high dose steroid therapy (IV solumedrol 100 mg IV daily as well as one time injection of triamcinolone 60 mg), Vitamin C 1 g PO daily and Zinc Sulfate 220 mg PO. This regimen resulted in significant improvement in healing of the surgical wound. In addition, infliximab therapy was considered however the patient declined due to potential associated side effects.

E-mail address: Shailja.Mehta@mountsinai.org (S. Mehta).

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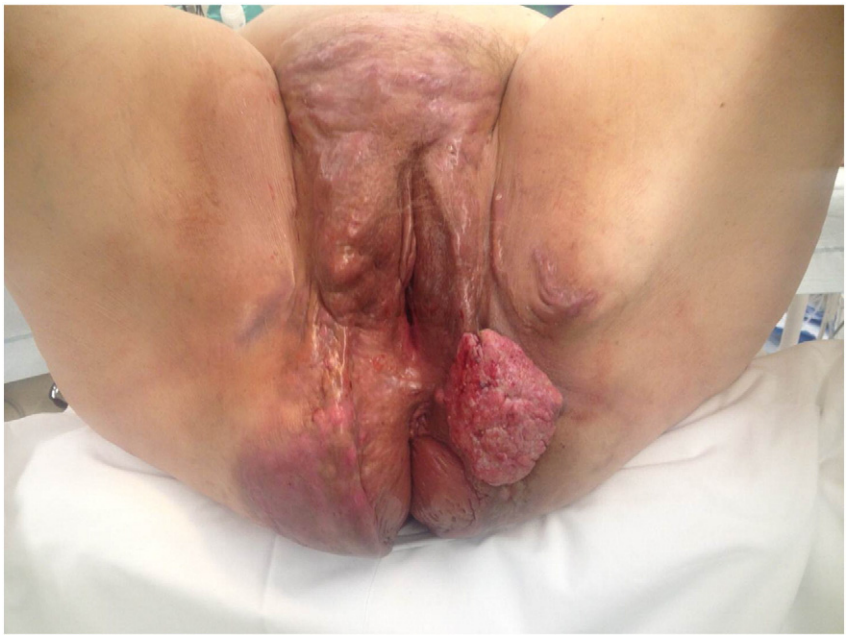

Image 1. Initial external pelvic examination.

The patient continues to follow up with plastic surgery and the gynecologic oncology team. The most recent images taken 8 months after surgery demonstrate a well healed vulva (Image 4). The final pathology revealed a mass with a well to differentiated, keratinizing squamous cell carcinoma; the tumor penetrated the specimen to 60% of its thickness (15 mm/25 mm). No definite lymphovascular space invasion was identified. All margins were free of tumor with the closest approaching the medial margin at 4 mm. The patient did not require postoperative adjuvant therapy and remains without evidence of disease.

3. Discussion

Our case highlights a rare consequence of long-standing poorly controlled hidradenitis suppurativa: squamous cell carcinoma. Although

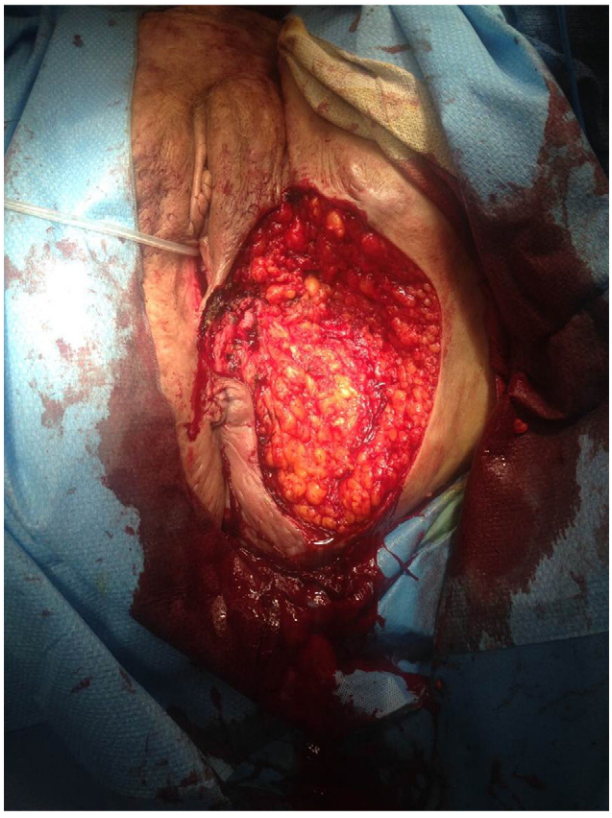

Image 2. Status post radical hemivulvectomy creating a large defect that required intervention by plastic surgery.

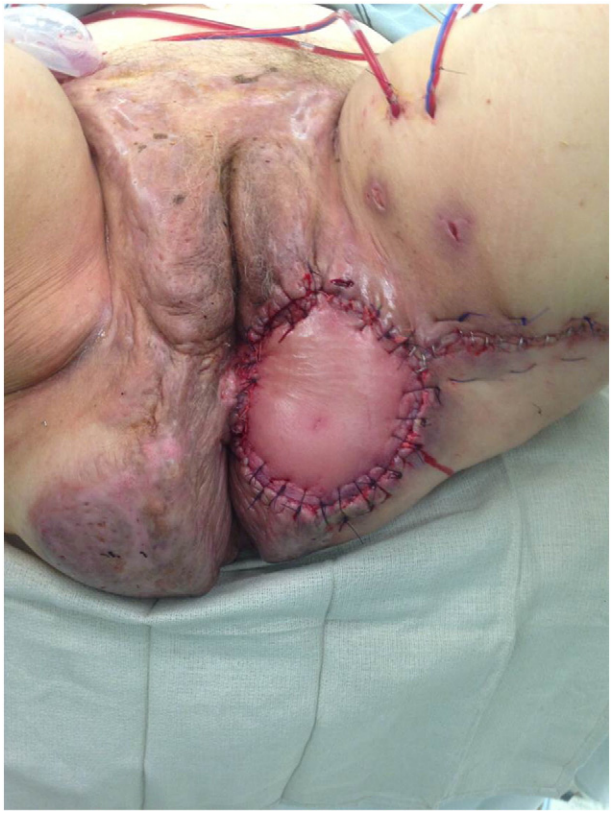

Image 3. Status post wound revision secondary to wound separation and infection.

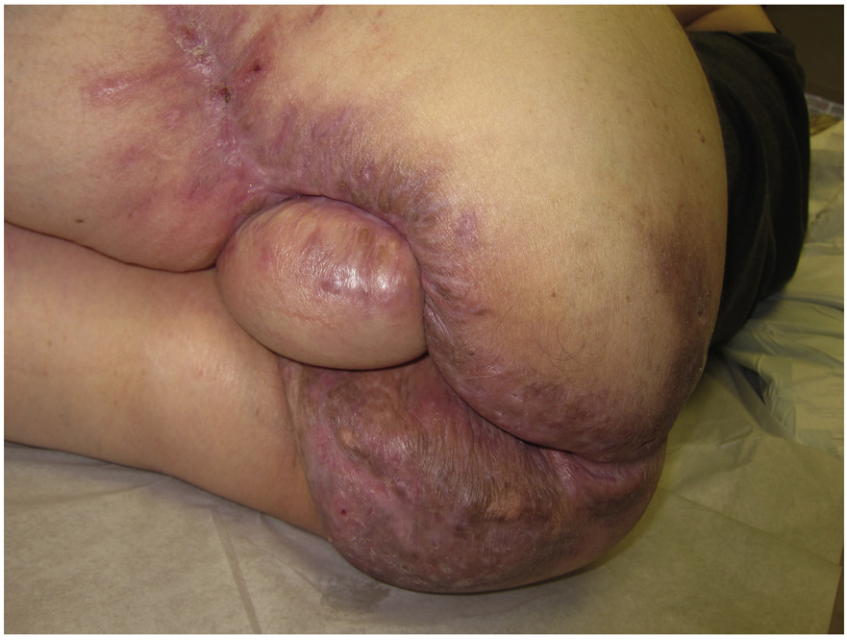

Image 4. Surgical site 8 months after surgery.

hidradenitis suppurativa is relatively common, the development of squamous cell carcinoma in the setting of HS/AI is thought to be a rare occurrence. Lavogiez et al. (2010) quoted an incidence of 4.6% in their cohort of 217 patients with HS with a stronger preference for males (4:1 ratio) (Maclean and Coleman, 2007; Mendonça et al., 1991). The prognosis is poor due to the advanced stage of SCC at time of diagnosis and difficulty in obtaining a biopsy. Frequently there is a delay in diagnosis of SCC and definitive surgical management. In a recent review of published cases of squamous cell carcinoma originating from HS/AI (Losanoff et al., 2011), the majority of events were described in the context of long-standing, chronic HS/AI (20–30 years) with 50% mortality noted from metastatic disease. It is thought that infection with the human papillomavirus is a risk factor for the development of SCC in patients with HS/AI.

HS should be treated early to avoid chronic progression of the disease that could ultimately lead to a rare but fatal consequence of HS/AI: the development of SCC (Lavogiez et al., 2010). Treatment should be tailored to the level of disease severity of HS/AI. In early stages, approaches to treatment include non-pharmacologic methods such as avoidance of skin trauma, hygiene, smoking cessation and weight management. Pharmacologic methods include topical clindamycin as first-line therapy, topical resorcinol (a chemical peeling agent), intralesional corticosteroids or systemic antibiotics. The cases of chronic and severe HS/AI require a more aggressive approach to avoid the chronic progression of the disease; in these cases, early surgical treatment in the form of complete excision is often warranted when medical therapies fail (Losanoff et al., 2011). Furthermore, a high index of suspicion and early tissue diagnosis should be performed in those with suspicion of malignancy. Previous authors have advocated that HS/AI arising in extra-axillary sites be regarded as a pre-malignant condition and not treated conservatively. Close follow-up and repeated skin biopsies should be performed in those with suspected malignancy.

In our patient, further complications after surgery included poor post-operative wound healing due to underlying HS/AI. Though previously thought to be related to infected sweat glands, current hypotheses is that hidradenitis suppurativa/acne inversa results from the "follicular triad" which includes follicular occlusion, follicular rupture, and an associated immune response (Danby and Margesson, 2010). Despite this new hypothesis, the role of the immune system as a contributor to the initiation of HS/AI has not been previously well studied. In our patient, high dose high potency steroid therapy resulted in a significantly decreased inflammatory reaction associated with HS. This ultimately led to better wound healing despite concomitant superimposed infection. The difficulty in treating severe and refractory cases of HS/AI has prompted researchers to study the effects of biologic agents routinely used for psoriasis and rheumatoid arthritis (Lee and Eisen, 2015). In particular, adalimumab and infliximab have been shown to be efficacious in treating HS/AI, although further investigation is warranted; such novel use of biologic therapies to decrease the inflammatory reaction associated with HS/AI is important for the gynecologic oncologist to consider when treating these complex surgical cases.

In summary, there should be a high index of suspicion for malignancy in patients with chronic, long-standing HS/AI; these patients should not undergo a delay in diagnosis or delay in definitive treatment in the form of surgical resection. In addition, prior to surgical intervention, every effort should be made to optimize surgical outcomes and wound healing, including a multi-disciplinary approach including plastic surgery and dermatology consultation.

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