CASE REPORT
Vulval Squamous Cell Carcinoma Arising in Chronic Hidradenitis Suppurativa

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Hidradenitis suppurativa is a chronic inflammatory disease of the sweat glands and hair follicles which is rarely associated with squamous cell carcinoma (SCC). A case of vulval SCC complicating hidradenitis suppurativa is presented. In addition to being the first case to report the association of vulval SCC and hidradenitis suppurativa in the English language literature, it also illustrates the difficulty inherent in distinguishing between invasive SCC and pseudoepitheliomatous hyperplasia on histological examination. The success of wide local excision of the vulval lesion and primary closure without recourse to skin grafts, rotational flaps, or healing by secondary intention is demonstrated. © 1999 Academic Press

INTRODUCTION
Hidradenitis suppurativa is a chronic suppurrative disease of the skin that is caused by follicular hyperkeratosis leading to retention of follicular products with secondary infection and inflammation of the apocrine and eccrine sweat glands. The disease is often misdiagnosed and inadequately treated. Mild cases are often treated medically; however, there is a high recurrence rate and severe cases almost always require surgical excision of the affected area [1]. Squamous cell carcinoma (SCC) is an uncommon complication of hidradenitis suppurativa, with only five cases being previously reported in women [2–6]. We report the first case in the English language literature of vulval cancer complicating hidradenitis suppurativa.

CASE REPORT
The patient was a 52-year-old Maltese woman, gravida 2, para 2, with type II (adult onset) diabetes who presented with a vulval mass. She had a 30-year history of hidradenitis suppurativa, previously managed in other institutions. Her father and two of her seven siblings had also been diagnosed with hidradenitis suppurativa. She had undergone surgical excision of affected areas in the neck, groins, axillae, and chest wall in 1970, 1980, and 1985. The right-sided vulval mass had been present for about 8 years, initially increasing in size until it was excised 2 years ago. The histology at that time was reported as hidradenitis suppurativa with no evidence of malignancy. She had been under regular review for many years by a dermatologist who had been managing her condition with antibiotics and Acitretin.

The vulval lesion recurred soon after excision and gradually increased in size with pain and discharge over a period of 2 years until she presented for the first time to Westmead Hospital and was referred to the Gynecologic Oncology Unit. On examination she had widespread scarring, induration, and occasional pustule formation on her back, thighs, axillae, and groins consistent with the diagnosis of hidradenitis suppurativa. In addition there was a 5-cm-diameter fungating mass arising from the right vulva which was highly suspicious of malignancy (Fig. 1). There were no clinically enlarged lymph nodes. The vulval lesion was biopsied under local anesthesia. However, a definitive diagnosis could not be made as histological examination of the biopsy specimen could not clearly distinguish between pseudoepitheliomatous hyperplasia and well-differentiated SCC (Fig. 2).

Due to the large size of the lesion, its persistence despite previous conservative therapy, and the possibility of malignancy, it was considered that the lesion would be best managed by wide local excision/hemivulvectomy with recourse to regional lymphadenectomy only if the final histology of the excised specimen confirmed malignancy. Radical local excision of the lesion with primary closure was subsequently performed under general anesthetic. Postoperatively, she was given a course of flucloxacillin for 1 week with early commencement of salt baths. The patient was highly motivated toward personal hygiene and meticulous in keeping the wound clean and dry. The wound subsequently healed well with no infection or breakdown.

The histology of the vulval specimen showed a well-differentiated SCC (Fig. 3), with a maximum depth of 20 mm from the surface of the overlying epidermis. The tumor cells showed keratinization (Fig. 2) and there was focal marked cellular atypia (Fig. 4). The excisional margins were clear of malig-
FIG. 1. Fungating mass arising from the right vulva. Note scarring and induration characteristic of hidradenitis suppurativa affecting the adjacent thigh.

FIG. 2. Initial biopsy of the vulval lesion showing a well-differentiated squamoproliferative lesion with associated chronic inflammation. It was not possible to distinguish pseudoepitheliomatous hyperplasia from a well-differentiated squamous cell carcinoma in this material (hematoxylin and eosin ×100).
FIG. 3. Well-differentiated squamous cell carcinoma showing keratin pearl formation (hematoxylin and eosin ×100).

FIG. 4. Tumor cells showing nuclear atypia and a mitotic figure (arrowhead) (hematoxylin and eosin ×400).
nancy. Four weeks later, a right inguinal-femoral lymphadenectomy was performed and all nodes were negative for malignancy. She made an uncomplicated recovery and the groin wounds healed well by primary intention.

DISCUSSION

Hidradenitis suppurativa results from a cycle of localized infection and inflammation, as the apocrine sweat glands become plugged with keratinaceous material with progressive dilation, stasis, and bacterial infection with skin flora to form localized abscesses. These subsequently rupture onto the skin surface, spreading the infection to surrounding tissues leading to a repeated cycle of abscess formation and chronic draining sinuses with subsequent scarring and fibrosis [7]. The disease often starts as a localized area of inflammation but commonly progresses to involve wider areas of skin with the axillae, groins, external genitalia, perineum, and perianal regions being the most commonly affected areas [7].

This case highlights the difficulties often encountered in the diagnosis of SCC on a background of hidradenitis suppurativa. Even though multiple superficial biopsies were taken and the histopathology was reviewed by two pathologists experienced in the field of gynecologic pathology, the presence or absence of malignancy was disputed prior to the definitive surgery. Difficulty may arise in distinguishing between a well-differentiated SCC and florid pseudoepitheliomatous hyperplasia. A number of features allow for accurate identification of the two entities. Pseudoepitheliomatous hyperplasia is usually associated with chronic irritation, whereas SCC is far less commonly so. SCCs have associated tissue destruction, necrosis, and often keratin pearls. Vascular and lymphatic invasion may be present. Mitotic activity is seen in both conditions, although abnormal mitoses are only seen in SCCs.

Hidradenitis suppurativa is rare before puberty and occurs most commonly in the third and fourth decades. Its sex prevalence is controversial but it is probable that anogenital disease is more common in males and disease at other sites is more common in females [4]. Apocrine sweat glands are stimulated by androgen and suppressed by estrogen which partly explains why hidradenitis suppurativa is rarely seen prior to puberty and never develops after menopause [8]. The exact etiology is unknown; however, poor personal hygiene, shaving, the use of depilatories and antiperspirants, obesity, excessive sweating, and diabetes are all thought to be associated factors [1].

Conservative management of hidradenitis with salt baths and antibiotics is sometimes successful in early and mild cases. A variety of therapies have been reported in the management of this condition including local injection with steroids, systemic steroids, isotretinoin, antiandrogens, and even radiation, all with limited success and high recurrence rates. Surgery remains the treatment of choice for chronic or severe cases [7]. Wide excision of the lesions is usually indicated as simple incision and drainage have resulted in recurrence rates of over 80% [9]. Some authors recommend the use of delayed primary closure or split skin grafts in cases associated with extensive disease [7, 9]. Goldberg et al. [10] describe a very advanced case of hidradenitis suppurativa presenting as bilateral vulval masses managed with an extensive resection of the affected area requiring delayed wound closure with local flaps and skin grafts, allowing antibiotic therapy and debridement during the interval between the primary surgery and delayed closure. The case reported here demonstrates that primary closure of the vulval wound can be successful in a highly motivated patient.

Complications of hidradenitis suppurativa may include severe psychosocial and economic disabilities in addition to anemia, fistulas, and osteomyelitis [1]. Squamous cell carcinoma is an uncommon complication, with only 5 of the 33 cases reported in the English language literature occurring in women. All 5 previously reported cases of SCC complicating hidradenitis suppurativa in women have involved the buttocks or perianal region [2–6].

The case reported here illustrates the pitfalls in long-term conservative management of advanced vulval hidradenitis suppurativa without gynecological review. Malignancy should always be considered in the presence of a vulval mass and adequate biopsies taken. Even when not complicated by malignancy, such cases of advanced vulvar disease may require extensive surgical resection which is most appropriately performed by a surgeon skilled in the techniques of radical vulvar surgery. In many institutions this radical surgery is most often performed by a gynecological oncologist.

REFERENCES