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Three fatal cases of squamous cell carcinoma arising in chronic perineal hidradenitis suppurativa

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ABSTRACT

INTRODUCTION Hidradenitis suppurativa (HS) is a chronic, inflammatory and suppurative disorder of skin bearing apocrine glands. The most severe complication is squamous cell carcinoma (SCC) and we here present three cases, all of which proved fatal, and review the past 40 years of published cases.

PATIENTS AND METHODS Three advanced cases of SCC arising in chronic HS have been referred for reconstructive surgery over the past 8 years. Another 28 cases published over the past 40 years were identified using a Medline search (search items in combination: hidradenitis, squamous, carcinoma).

RESULTS The male:female ratio was 4:1, most (61%) were perineal or buttock. We found no reports of SCC arising in axillary disease. The symptomatic history of HS prior to SCC diagnosis ranged from 3–50 years with a mean of 25 years. Age at diagnosis of SCC ranged from 27–71 years, and 15 patients (48%) died within 2 years of SCC diagnosis.

CONCLUSIONS We advocate that hidradenitis suppurativa arising in extra-axillary sites is a pre-malignant condition, and should not be treated conservatively; curative resection is the mainstay of management.

KEYWORDS

Squamous cell carcinoma – Hidradenitis suppurativa – Management

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We report three cases of squamous cell carcinoma (SCC) arising in hidradenitis suppurativa where there was delayed diagnosis of malignant change. All presented late, and all patients died from their disease. These cases highlight the need for a greater clinical awareness that long-standing buttock and perineal hidradenitis suppurativa is a pre-malignant condition and should not be managed conservatively. We also present a review of all cases published in the English language in the last 40 years of SCC arising in hidradenitis suppurativa.

Case 1

A 50-year-old man was first diagnosed with hidradenitis suppurativa when aged 18 years after the development of an abscess on his buttock. At that time, he was told that this was an incurable condition, and he endured many years of recurrent abscesses. He was referred to the gastroenterologists with peri-anal suppuration who diagnosed Crohn's disease although he had normal small bowel imaging and normal rectal biopsies. Steroids gave no symptomatic relief, and the disease continued to progress. He developed an ulcerated lesion on his right buttock, biopsy of which showed SCC (Fig. 1). Surgery was not considered at this stage. A 6-week course of radiotherapy and chemotherapy followed, which gave moderate symptomatic relief. Eight months later, he developed an SCC in his left buttock, and was referred to our service. The extent of his disease required a radical excision from both buttocks and perineum along with an abdominoperineal resection of the rectum and a groin dissection. The defect was reconstructed with a pedicled rectus abdominis musculocutaneous flap and split skin grafts. Two months later, he developed a recurrence in the other groin and proceeded to a second groin dissection. All excised nodes were positive for SCC metastasis. Nine months later, he developed a recurrence in his right thigh. Investigations showed the tumour involved much of his posterolateral thigh musculature and femur. Local control was achieved by a right hip disarticulation and anterior thigh flap cover. He

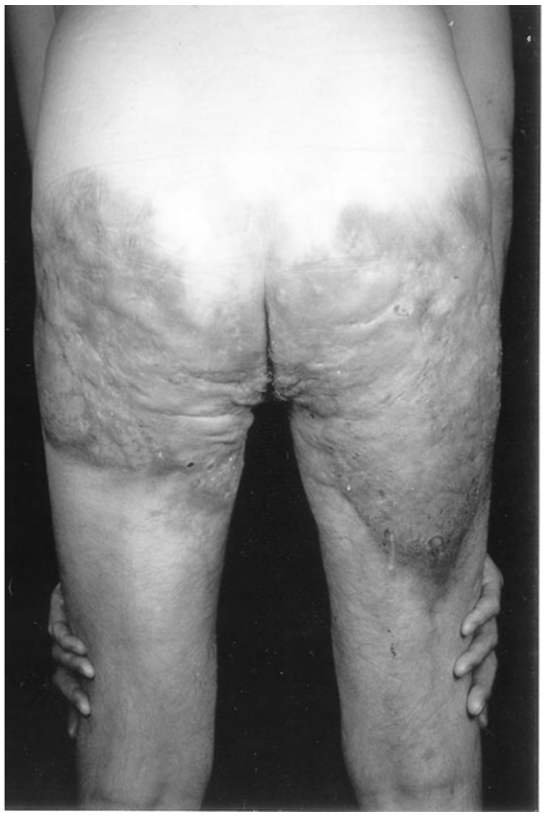

Figure 1 Case 1 showing very extensive buttock hidradenitis suppurativa. There are ulcerated areas on the medial aspect of each buttock that proved positive for SCC.

suffered a further buttock recurrence 4 months later, and died following a rapid deterioration 26 months after diagnosis.

Case 2

A 61-year-old woman was referred to the general surgeons with a right groin abscess. The patient gave a 40-year history of hidradenitis, initially confined to her axillae, with her first episode of perineal hidradenitis 10 years previously. In the previous 18 months, she had been troubled with discharge from her perineum. The right groin abscess was incised and 300 ml of pus drained. Further sepsis involving her right vulva and perineum was noted, but no biopsies were taken. The patient was referred for a dermatology opinion. She developed a right iliofemoral vein thrombosis, and was commenced on heparin and warfarin. She was then referred to our service. An examination under anaesthetic was carried out. She had a large fixed mass in the right iliac fossa arising from her pelvis with a very large, deeply fixed sacral ulcer extending anteriorly (Fig. 2), a large ulcer with gross induration of her right vulva extending to the groin, and extensive left groin lymphadenopathy. Multiple biopsies

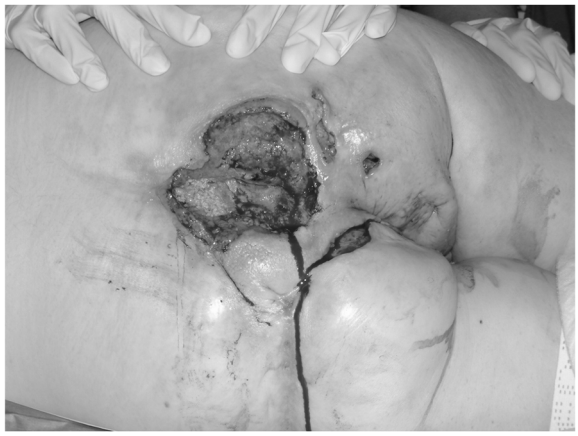

Figure 2 Case 2 showing a large ulcerated SCC on the sacrum. The tumour extended anteriorly to the perianal region, vulva and groin.

were taken of all affected areas, and a fine needle aspiration of the left groin node. Colonoscopy was normal. All specimens contained SCC. In view of the extent of the disease, surgery would not have been curative, and the patient was offered palliative radiotherapy. She received half of her planned treatment before she died from an episode of sepsis and acute haemorrhage 2 months after diagnosis.

Case 3

A 47-year-old man with a 9-year history of hidradenitis suppurativa affecting both buttocks, groin and peri-anal regions was referred to our service for wide excision and split skin grafting. During the previous 4 years, he had multiple treatments for his abscesses and sinuses. MRI imaging 2 years previously had shown changes that were suspicious, but not diagnostic, of osteomyelitis in his ischial tuberosity. He had developed a large peri-anal ulcer. A biopsy showed SCC. He was also under the care of the orthopaedic surgeons for severe hip pain. An MRI at this stage showed that the tumour had spread extensively and affected his ischial tuberosity, pelvic floor, obturator internus and medial border of gluteus maximus. Given the extensive spread, the tumour was deemed inoperable, and the patient was given palliative chemotherapy and radiotherapy. He died 9 months after SCC diagnosis.

Discussion

Using a search on Medline (search items in combination: hidradenitis, squamous, carcinoma), we reviewed the identified 28 cases published in the past 40 years (Table 1). Out of 31 individual cases (including the three reported

Table 1 Reported cases of SCC arising in hidradenitis suppurativa

Author	Age	Sex	History of HS (years)	Nodes	Surgery performed	Outcome
Case 1	50	M	32	+	Abdominoperineal resection	Died at 24 months
Case 2	61	F	40	+	None	Died at 2 months
Case 3	47	M	9	+	None	Died at 9 months
Rosenzweig *et al.*[1]	50	M	20	?	Excision and skin graft	Well at 1 year
Bocchini *et al.*[2]	36	M	?	?	Multiple surgical interventions	Died at 18 months
Altunay *et al.*[3]	54	M	30	+	None	Died at 3 months
Manolitsas *et al.*[4]	52	F	30	?	Radical local resection	Full recovery
Lin *et al.*[5]	55	M	30	+	Wide excision	Died at 2 months
Ritz *et al.*[6]	61	M	45	+	Radical resection	Died at 4 months
Li *et al.*[7]	68	M	50	?	Resected	No comment
Malaguarnera[8]	65	M	20	+	Radical excision	Died at 7 months
Dufresne *et al.*[9]	52	F	36	+	Mohs micrographic excision	Died at 7 months
Shukla *et al.*[10]	71	F	50	?	Massive resection	Symptom-free at 4 years
Perez-Diaz *et al.*[11]	60	M	25	+	Completely excised	Disease-free at 1 year
Welsh *et al.*[12]	50	M	20	+	Incomplete resection	Died at 2 months
Mendonca *et al.*[13]	57	M	35	–	Wide excision and skin graft	Well at 1 year
Williams *et al.*[14]	27	M	11	–	Wide local excision	Well at 1 year
Anstey *et al.*[15]	67	M	40	?	Massive resection	Died at 2 years
Weber *et al.*[16]	48	F	37	–	*En bloc* dissection	No comment
Chicarilli[17]	59	M	30	?	Massive resection	Disease-free at 2 years
Zachary *et al.*[18]	55	M	3	?	Radical excision	Disease-free at 1 year
Sparks *et al.*[19]	35	M	10	+	None	Died
Johnston *et al.*[20]	54	M	4	?	Wide resection	Died at 6 months
Mora *et al.*[21]	47	M	5	?	Abdominoperineal resection	Disease-free at 2 years
Mora *et al.*[21]	54	M	Many	?	None	Died at 5 months
Black *et al.*[22]	44	M	21	?	Radical excision	Died at 2 years
Gordon[23]	28	F	17	+	Wide local excision	No comment
Thornton *et al.*[24]	?	?	20	?	No comment	No comment
Alexander[25]	40	M	20	–	Abdominoperineal resection	Died at 18 months
Humphrey *et al.*[26]	48	M	8	+	Radical excision	Alive – unknown follow-up time
Donsky *et al.*[27]	44	M	23	?	Radical excision	No comment

HS, hidradenitis suppurativa.

here), 15 originated in the buttocks/thigh, nine in the perianal region, four in the perineum, two in the vulva and one arose in the groin.

Although the cases are limited in their reported follow-up, 15 patients (48%) died within 2 years of SCC diagnosis. The age at diagnosis of SCC ranged from 27–71 years with a mean of 51 years. The symptomatic history of hidradenitis suppurativa prior to SCC diagnosis ranged from 3–50 years with a mean of 25 years.

The male:female ratio was 24:6 with one case unknown. This is in keeping with the finding that although hidradenitis suppurativa overall is more prevalent in women, the incidence of extra-axillary disease is much more common in men.[10] We found no reports of SCC arising in axillary disease. Of the 13 cases (42%) with positive nodes at SCC diagnosis in which outcome at 2 years is known, only one patient was still alive.[11]

A common theme in the reported cases is multiple minor surgical interventions, under different specialists, over months or years prior to a diagnosis of SCC. The three cases we present are notable for the delayed histological diagnosis, and thus advanced stage at referral to our service. As radical

surgery can be curative, they serve as a pertinent reminder of the importance of early histological diagnosis, either by incision or excision biopsies, in all non-healing or otherwise suspicious lesions in chronic hidradenitis suppurativa.

Conclusions

We advocate that hidradenitis suppurativa arising in extra-axillary sites is a pre-malignant condition, and should not be treated conservatively; curative resection is the mainstay of management.

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