Squamous cell carcinoma arising from perineal lesion in a familial case of Hidradenitis suppurativa

K Chandramohan, Anitha Mathews, Anu Kurian, Shaji Thomas, K Ramachandran


ABSTRACT
Hidradenitis suppurativa (HS) is a chronic dermatologic disorder characterised by multiple vesiculopustular cutaneous lesions in skin. It sometimes expresses autosomal dominant mode of inheritance. In some patients, it can give rise to squamous cell carcinoma of skin. Unlike the usual marjolin’s ulcer which develops in chronic scars, this disease carries a grave prognosis with high recurrence rate and mortality.

Here we are reporting the case of a 40-year-old gentleman with hidradenitis suppurativa, who presented with squamous cell carcinoma in the perianal region, and had multiple family members affected with HS. A short history and review of literature are discussed. Squamous cell carcinoma arising from HS is a major therapeutic challenge. This malignancy carries very high rates of recurrence and mortality. Careful surveillance of the lesions of HS, especially occurring in perineal region is very important to diagnose secondary malignancies. Similarly, meticulous surgical excision with careful resection of all sinus tracks and frozen section examination of margins are very important to eradicate the disease.

Key words: Hidradenitis Suppurativa • Squamous cell carcinoma

INTRODUCTION
Hidradenitis suppurativa (HS) is a chronic relapsing inflammatory skin disorder with recurrent acneiform eruptions. The term ‘hidradenitis’ is derived from ‘hidros’ which means sweat and ‘adenos’ which means gland. It usually affects axilla, perineal region, and inframammary area and perianal region, areas where apocrine sweat glands are present. But it can affect any region of the body. It is a chronic disease which is often refractory to treatment. It was Verneuil who first described the etiology of the disease (1). This condition is also noticed to have autosomal dominant inheritance (2). Usually along with HS, acne (particularly conglobate type), pilonidal sinus, and chronic scalp folliculitis frequently coexist, as parts of ‘follicular occlusion tetrad’.

Major initiating event in pathogenesis is occlusion of follicular infundibulum followed by abscess formation and rupture. Three key elements in the diagnosis of HS are typical lesions, characteristic distribution and recurring nature. As a result of inflammation, follicles rupture and spill contents that include

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This causes further chemotactic response and abscess formation. A very common sequela is the formation of sinus tracks. Later, this leads to chronic scars and fibrosis. Squamous cell carcinomas develop from the lesions (3,4). Here we are reporting a case of HS giving rise to squamous cell carcinoma in the perianal region.

CASE REPORT

A 40-year-old man, who was suffering from HS, from the age of 18, presented to the surgical oncology outpatient department with recurrent squamous cell carcinoma of perianal region, which was first excised 2 years before. On examination, the patient had widespread cutaneous abscesses all over body especially axilla, groin and perineal region. A computerised tomography was done, which showed mass lesion at the natal cleft reaching up to the anus. The lesion was confined to subcutaneous fat and there was no infiltration to gluteal muscles. Incision biopsy from perineal lesion confirmed the squamous cell carcinoma. Wide excision of the lesion with bilateral gluteal rotation flaps and split-AQ3 thickness skin grafting was done. A protective colostomy was also done.

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On examination, the patient had widespread cutaneous abscesses all over body especially axilla, groin and perineal region (Figure 2). He also had an ulcer with everted margins in the left perianal region with satellite nodules extending to upper medial thigh and gluteal region. A computerised tomography was done, which showed mass lesion at the natal cleft reaching up to the anus. The lesion was confined to subcutaneous fat and there was no infiltration to gluteal muscles. CT scan of the abdomen and chest doesn’t show any metastatic disease. Incision biopsy from perineal lesion confirmed the squamous cell carcinoma. Wide excision of the lesion with bilateral gluteal rotation flaps and split-thickness skin grafting was done. A protective colostomy was also done as the lesion was very...
close to the anal canal, and this was closed after 6 months. Histopathological examination showed well differentiated squamous cell carcinoma. All margins were free of disease.

Postoperatively there was minimal wound infection and partial skin graft loss. This was managed conservatively. Patient was disease free one year after surgery (Figure 5).

DISCUSSION
HS is a chronic inflammatory disease of the skin and subcutaneous tissue in areas where apocrine glands are plenty. Apocrine glands are widely scattered in all parts of the body, but more abundant in axillae, perianal region, labia, scrotum, chest, and abdomen and around breasts. Rarely the disease presents as autosomal dominant mode of inheritance (5,6). The available evidence points towards the single gene transmission. Recently, the genetic locus responsible for HS has been identified in a Chinese family at chromosome 1p21.1–1q25.3.24 (7).

In the reported case, the mode of transmission is autosomal dominant, because more than 50% of siblings are affected with the disease. Squamous cell carcinoma can develop in HS lesions. Review of literature showed 33 cases of squamous cell carcinoma arising in the setting of HS. Among these, 15 originated in the buttocks/thigh, ten in the perianal region, five in the perineum, two in the vulva and one arose in the groin. It occurs as Marjolin’s ulcer, similar to those arising from chronic scars. Perianal lesions are more common in males and have more propensities to become malignant, compared to lesions in other sites of the body. Jackman et al. reported 3.2% incidence of squamous cell carcinoma in perianal hidradenitis (4). HS lesions usually take 19–32 years to become malignant. But there are no reports to suggest more risk of malignancy in familial cases. Review of literature by MacLean et al. showed that majority of patients presented with aggressive disease and nearly 50% of the patients who developed squamous cell carcinoma died within 2 years (8). This may be due to two reasons. Firstly, delay in diagnosis because of the failure in detecting the development of cancer in the chronic wound. Even multiple biopsies may be negative. Secondly, the surgical excision carries a chance of leaving behind microscopic foci and hence chance of recurrence. This happens because cancer spreads through multiple fistulous tracks into the surrounding tissue. Hence, meticulous surgical excision with careful resection of all sinus tracks and frozen section examination of margins are very important. Re-excision should be done if margin is positive for malignancy by frozen section or by permanent paraffin sections. Various choices available for reconstruction are myo-cutaneous flaps, split-thickness skin grafting, or healing by granulation. In the management of perianal wound, protective colostomy is often a valuable help.

In conclusion, squamous cell carcinoma arising from HS is a major therapeutic challenge. This malignancy carries a very high recurrence rate and very high mortality. Aggressive radical excision is the mainstay of treatment. There is no role of adjuvant treatment like chemotherapy or radiotherapy.

CONSENT
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

REFERENCES
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