Squamous Cell Carcinoma Arising in Verneuil's Disease: A Case Report with Literature Review

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Abstract:
Verneuil's disease, or hidradenitis suppurativa (HS), is a chronic inflammatory skin condition that affects apocrine glands in the axillary and anogenital regions. It is a suppurative and fistulizing disease and its degeneration into squamous cell carcinoma (SCC) is exceptional. We report a rare case of a 61-year-old man with 44 years history of Verneuil's disease confined to the perineal region. The patient was admitted to the emergency department for worsening symptoms over the last two months. Histological analysis of a skin biopsy revealed a squamous cell carcinoma treated with surgical excision. After one year of follow-up, outcome has remained favorable. He didn't present any sign of recurrence or metastatic spread. The transformation of hidradenitis suppurativa to squamous cell carcinoma is rare and often diagnosed too late and thus at advanced stage. It is considered the most severe complication of hidradenitis suppurativa. Close monitoring of patients with HS helps finding modification or worsening of skin lesions and allows early treatment.

Keywords: Verneuil’s Disease, Hidradenitis Suppurativa, Squamous Cell Carcinoma.

I. INTRODUCTION

Verneuil’s disease is a chronic suppurative disease of the apocrine glands of the axillae, groin, vulva and anal region. Its malignant transformation is a rare dreaded complication. We report a case of cutaneous squamous cell carcinoma complicating hidradenitis suppurativa.

II. CASE REPORT

A 61-year-old man with a 45 year pack smoking history and Verneuil’s disease was admitted to the emergency department for clinical deterioration of his skin lesions. His disease started 44 years ago with recurrent painful nodules, abscesses, and fistulas of the perianal region treated initially with courses of systemic antibiotics (tetracycline) but with no cure. The patient was lost to follow up for several years.

He presented, two months prior to his admission, a tender painful nodule turning into diffuse tracts and abscesses with non healing sinuses and extending to all the perineal area. Palpable lymph nodes were found and ultra sound was used to identify and measure them “Fig. 1”.

The patient was in a good health. He was hemodynamically stable. Skin biopsies proved the histopathological diagnosis of invasive SCC. Lymph nodes’ biopsies didn’t show any signs of cancer.

Staging investigations including computed tomography (CT) did not find any evidence of systemic disease elsewhere.

The patient received surgical treatment and underwent wide excisions of the involved areas of the perineum “Fig. 2”. Reconstruction was performed with musculocutaneous flap. Neither recurrence nor signs of metastatic dissemination were noted during the last two years of follow up.

Fig. 1. The loss of substance involving the perineum of our patient
Fig. 2. Flat lay with wide excision of the perineal lesions presented by our patient

III. DISCUSSION

Verneuil’s disease is a chronic suppurative disease of the apocrine sweat glands. It was first described by Velpeau in 1839, then by Verneuil in 1854. It is a recurrent, painful skin condition with nodules that develop into suppurative and fistulizing abscesses of apocrine gland-bearing skin of the axillae, groin, perineal and perianal regions and inframammary folds [1]. The axillary region is most commonly affected in women, the buttocks and perianal skin in men [2,3,4].

The exact aetiology of Verneuil’s disease still remains unclear. Smoking and obesity are more common in patients with HS. They are two environmental factors that contribute to the incidence and severity of the disease. In fact our patient is a heavy smoker.

Chronic complications may occur during the HS course. They include bacterial infection, fistulas, the involvement of important structures (urethra, bladder, rectum, and peritoneum) and malignant transformation into squamous cell carcinoma. The malignant degeneration is an exceptional and very dreaded complication with less than 100 cases reported in medical literature [2–15].

The first cases of SCC associated with HS were reported by Anderson in 1985 and Jackman in 1959 with a prevalence of 1.7 and 3.2 % respectively [5-6]. It’s more common in men. Only two cases of women that were reported in literature [7,8]. In more than 2/3 of the cases, malignant degeneration occurs mainly in the anogenital region.

The average age at onset is 51 years (range 27-68 years). The mean history of HS prior to SCC diagnosis is 20 years (range 3-50 years).

Neoplastic transformation occurs over an extended period of time. Once the degeneration is made, the clinical behavior becomes very aggressive over short periods of time. For our patient, it only took two months for the SCC to rapidly progress to extend to all the perineum. The development of SCC is multifactorial and superimposed on chronic HS. It is often associated with early metastasis (40 % of cases) and high mortality rates with most deaths occurring in the first few months [9,10].

Chronic states of inflammation and irritation produce an environment that favors malignant transformation. The immune response generates free radicals that increase the risk for DNA mutations [11;12]. The ability of HS to form squamous cancers reinforces the argument for early and complete resection [13]. Close follow up is necessary to detect early signs of malignant transformation in sufferers whose quality of life is highly affected by the recalcitrant painful lesions, the inefficient treatments, depression and social isolation.

The radical excision of the whole affected tissue is considered to be the only curative treatment. The surgical defect is either left to heal by secondary intention or covered with flap [15,16].

IV. CONCLUSION

SCC is considered the most dreaded complication arising from HS, with high morbidity and mortality. Close monitoring of worsening skin lesions and early skin biopsy of chronically HS affected areas help prevent malignant degeneration.

REFERENCES


