ISSN: 2634 - 680X

Journal of Clinical Case Studies Reviews & Reports

SCIENTIFIC Research and Community

Case Report

Open Access

Hidradenitis Suppurativa, Can Be Fatal: Case Report

Badr I Abdulrauf* and Jamal A M Ilyas

Section of Plastic Surgery, Department of Surgery King Faisal Specialist Hospital and Research Center Jeddah, Saudi Arabia

ABSTRACT

Hidradenitis Suppurativa (HS) is a relatively common chronic debilitating skin condition characterized by recurrent abscessesand sinuses formation on apocrine sweat glands bearing areas. Although immune modulating therapies have beenpopular lately but definitive surgery with wide excision and reconstruction can provide permanent cure for the involvedareas. Here we are reporting an unfortunate case of a 47 years old male who has been suffering from HS in the buttocks andperineum since adolescence. Patient was first seen 8 years back and he was advised to consider wide surgical excision and skin grafting. Patient did not accept this solution and kept undergoing alternative treatments with prolonged courses ofantibiotics as well as steroids. Eventually he presented with fungating areas in the gluteal region which proved to be moderatelydifferentiated squamous cell carcinoma (SCC). Sacrum was already invaded with the disease and has metastasized toregional lymph nodes, liver and lungs. Patient was treated as palliative including conservative surgical debulking and skingrafts. He received further oncologic treatment in a different center overseas. Unfortunately developed brain metastasisand expired two years following diagnosis of Marjolin's ulcer (MU). Patients with chronic HS, recurrant infections andsinuses must be encouraged to seriously consider definitive surgical treatment to eliminate the risk of such long term fatalcomplication of their disease.

***Corresponding author**

Badr M I Abdulrauf, MD, FRCSC, Section of Plastic Surgery, Department of Surgery,King Faisal Specialist Hospital and Research Center PO Box 40047, Jeddah 21499, Saudi Arabia.

Received: March 26, 2020; **Accepted:** April 01, 2020; **Published:** April 06, 2020

Keywords: Hidradenitis Suppurativa, Squamous cell Carcinoma, Marjolin's ulcer, Fatal

Introduction

Hidradenitis suppurativa (HS), Acne inversa, is chronicrelapsing inflammatory suppurtaive and cicatricle skindisorder of the folliculo-pilosebaceous units, Describedoriginally by Velpeau in 1832. Clinically this debilitatingdisease is characterized by chronic inflammation, recurrentabscesses, nodules and fistulas over apocrine glands bearingareas, e. g. axillae, breast, groin, peri-anal, buttocks andperineal region [1].

Hurley Classification (Stage I-III) clinically states as deepseated follicular papules and pustules eventually turnsnodules, abscesses formation that discharge foul-smellingpus. The condition further progresses as tunnels, ultimatelyleads to inter-connected sinuses and eventually acute onchronic inflammatory changes and thickened scars.

Aberrant immune system may respond to the commensal peptides and the recruitment of inflammatory markers (TNF IL-1b, IL-17) and cytokines. This in turn gives rise to hyper metabolic plug of the follicular infundibulum and subsequent activation of inflammation by the keratin released in the dermis due to its rupture [2].

Heredity, hormonal imbalance, heat & humidity, sweating, rubbing of skin-to-skin, deodorants or other topical chemicals, are among the inciting factors. This is also associated with abnormal inflammatory responses to follicle rupture or to the normal bacterial flora in the skin, or unusual healing responses to follicular injury. Chronic inflammation produces an environment that favor oncogenesis through the dysregulation of tumor suppressor genes and through self-sufficient growth [3]. HS may be seen with Crohn's disease and rheumatoid arthritis. Several rare inflammatory conditions of the skin like Sweet's syndrome, Behçet's disease, or Pyoderma Gangrenosum may be seen with HS. The metabolic syndrome, with its tendencies toward diabetes; or hormone imbalances such as polycystic ovaries, are often found in patients with HS. The overall prevalence of HS is approximately 3.2 to 4.6 % in general population [4]. Though hidradenitis female to male ration is 4:1 however Marjolin's ulcer (MU) male to female ratio is 6.75:1. This is defined as the malignant degeneration of chronic inflammatory tissue or pre-existing cicatrix into the Squamous cell carcinoma [5]. Biological behavior of MU is much more aggressive than the SSC arising from the non-scar sources.MU metastasizes more frequently with relatively reduced long term survival. Around 50% of these patient died within 2 years after the diagnosis [6]. To the best of our knowledge, so far 94 cases are reported as MU with the history of HS.Most such cases developed in men who had long-standing HS on their genitals or around their anus. (Probably related to HPV? Androgen related). Studies have shown that long-lasting HS and the involvement of the buttocks and perineal areas increase the risk of malignant transformation to SCC [7]. The combination of SCC and HS is often associated with early metastasis and high mortality rates the overall 3-year survival rate is 65% to 75% and 10-year survival is 34%. However, for those with metastasis to the lymph nodes, the 3-year survival rate significantly drops to 35% to 50%.

Because of the extensive nature of this malignancy, surgery is often rejected by the patient or physicians [8]. The radiotherapy is usually ineffective because of the extent and volume of tissue which must be irradiated. Also, the moist nature of the perineum

and the extensive infection preclude an effective dose and this might produce severe morbidity. The relative chemo-resistance of SCC, severe infection and the advanced age of the patients all militate against the use of cytotoxic chemotherapy [9].Case presentationThis is a 47 years old smoker male, body mass index of 28 kg/m2, with almost 30 years history of HS. The main anatomical areas involved were gluteal, perineal, posterior thigh and groin. He had several incision and Drainage procedures over the years in addition to topical antimicrobials and antibiotics courses. Upon our first encounter with the patient he was advised for wide excision and reconstruction with skin grafting specifically to the buttocks and perineal areas. Patient refused this method of treatment and did not come for any follow up. In retrospect, as per review of medical record meanwhile the patient has been followed by other specialties including infectious disease, general surgery and Dermatology. Eventually he presented to emergency medical services (EMS). This was our second encounter with this patient almost 8 years after our clinic visit. Apart from the classic picture of extensive interconnecting gluteal abscesses, sinuses, fistulas and fibrotic scars the most suspicious lesion was an exophytic, fungating lesion of 4x3 cm above the natal cleft region.Multiple biopsies proved the histopathological diagnosis of moderately differentiated invasive SCC. Computed tomography (CT)) and Magnetic resonance imaging (MRI) of the pelvis and abdomen have shown, cystic tumor involving specifically the sacral region's skin and subcutaneous tissue with fistulas and tunneling along with the involvement of the sacral bone. Groin lymph nodes were also reported to be involved on the CT and MRI. There were also suspicious metastatic deposits found in the right lung and the liver.

Case was reviewed in the Multi-disciplinary tumor (MDT) board meeting and patient was considered to be a palliative on basis of distant metastases and chemotherapy would not be useful. Surgical excision and debulking was suggested with possible plan of postoperative radiation. The possible procedure of sacral bone resection (sacrectomy) as part of surgical treatment was thought to be of significant morbidity considering the poor expected overall prognosis.

Patient first undergone a right inguinal lymph node (LN) biopsy and a diversion colostomy. LN biopsy was also consistent with SCC. 10 days later patient undergone wide local excision of gluteal HS, MU, coverage with autogenous split thickness skin grafts (STSG) taken from right thigh and Vacuum assisted closure (VAC). Indeed, the objective was to palliate patient's offensive painful ulcers and to improve his quality of life. The patient and his family decided to take second opinion overseas, where he had radiotherapy as well as trial of chemotherapy (though this is not usually beneficial as per most of the literature). Following the completion of his therapy overseas patient was noted to have temporary improvement of overall condition and morale.

11 months later patient was re admitted this time unfortunately with brain metastasis. Neurosurgery gave an option of ventriculo-peritoneal shunt. However, the patient could not survive much longer at this admission. He deceased about 2 years following the diagnosis of MU.

Discussion

HS is a relatively common cutaneous condition usually beginning in early adolescence, and can be quite debilitating functionally and socially. Usually H.S. patients are seen by various specialists like general practitioners, family physicians, dermatologists, general surgeons, and Plastic surgeons.Unfortunately many patients would hesitate going under the knife for excision and reconstruction, instead would go for long term trials of immune modulating therapies, prolonged antibiotics, steroids, laser and even radiation. Surgical elimination of persistent sinus tracts is the only curative method that can eradicate chronic HS in a particular region.Medical treatments would benefit in early stages and not so when already sinuses and fistulae have developed. In fact these can mask an underlying cancer development and can give a false sense of healing. Our patient seemed to have all the risk factors of developing a MU as a complication to HS: chronicity of the disease for about 30 years, involvement of perineal areas, a male and smoker.

Although rare, SCC is considered the most severe complication arising from chronic HS, with high morbidity and mortality. The management of SCC in HS has not been standardized, but current reports and literature review shows, the most effective treatment for HS-derived MU is wide surgical excision with adjuvant radiotherapy based on tumor staging [10-12]. Aggressive treatment of established MU on top of HA may prevent recurrence and cancer-related deaths [13].

In conclusion, this is a preventable complication, we cannot overemphasize the importance of early wide surgical excision of chronically HS affected areas.

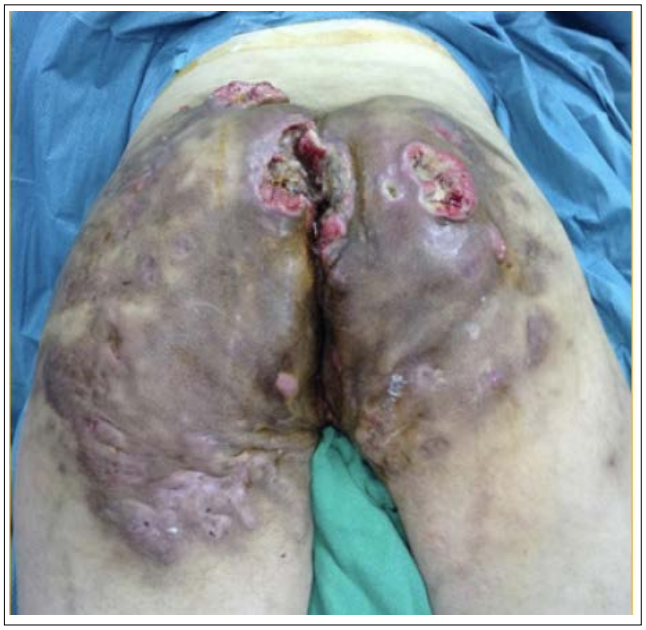

Figure 1: Recent Presentation in EMS.

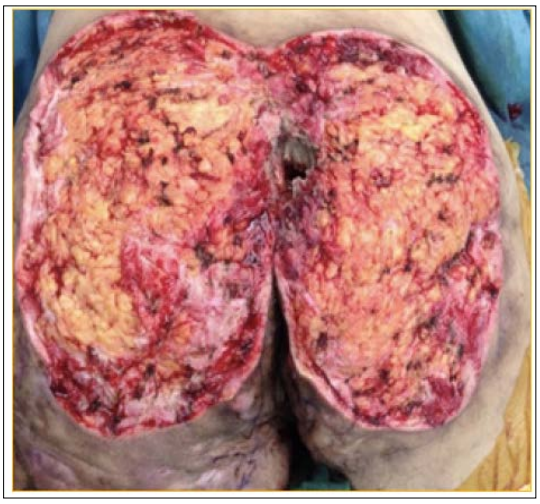

Figure 2: Post debulking.

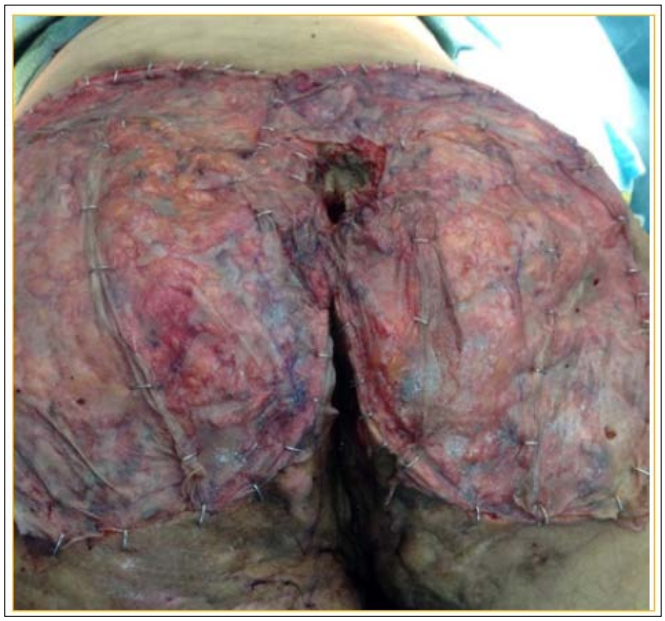

Figure 3: Post Skin grafts application.

References

1. Jansen T, Plewig G (1998) Acne Inversa. Int J Dermatol 37: 96-100.
2. Dufresne RGJr, Ratz JL, Bergfeld WF, Roenigk RK (1996) Squamous cell carcinoma arising from the follicular occlusion triad. J Am Acad Dermatol 35: 475-477.
3. Chapman S, Delgadillo D, Barber C, Khachemoune A (2018) Cutaneous squamous cell carcinoma complicating hidradenitis suppurativa: a review of the prevalence, pathogenesis and treatment of this dreaded complication. Acta Dermatovenerol Alp Pannonica Adriat 27: 25-28.
4. Lavogiez C, Delaporte E, Darras-Vercambre S, Lassalle EMD, Castillo C, et al. (2010) Clinicopathological study of 13 cases of squamous cell carcinoma complicating hidradenitis suppurativa. Dermatology 220: 147-153.
5. Choa R, Rayatt S, Mahtani K (2015) Marjolin's ulcer. BMJ 351: h3997.
6. Maclean GM, Coleman DJ (2007) Three fatal cases of squamous cell carcinoma arising in chronic perineal hidradenitis suppurativa. Ann R Coll Surg. Engl 89: 709-712.
7. Chandra Mohan K, Mathews A, Kurian A, Thomas S, et al. (2009) Squamous cell carcinoma arising from perineal lesion in a familial case of Hidradenitis suppurativa. Int Wound J 6: 141-144.
8. Vijay K Shukla, L E Hughes (1995) a case of squamous cell carcinoma complicating hidradenitis suppurativa. European Journal of Surgical Oncology 21: 106-109.
9. Maugham TS (1994) Basic principle of radiotherapy for surgical oncologist, Effect of Radiotherapy on superficial tissues. Implications of treating skin canceer Eur J Surg oncol 20: 74-79.
10. Jennings L, Schmults CD (2010) Management of high-risk cutaneous squamous cell carcinoma. J Clin Aesthet Dermatol 3: 39-48.
11. Jambusaria-Pahlajani A. Miller CJ, Quon H, Smith N, Klein RQ, Schmults CD (2009) Surgical monotherapy versus surgery plus adjuvant radiotherapy in high-risk cutaneous squamous cell carcinoma: a systemic review of out-comes. Dermatol Surg 35: 574-585.
12. Veness MJ (2008) The important role of radiotherapy in patients with non-melanoma skin cancer and other cutaneous entities. J Med Imaging Radiat Oncol 52: 278-286.
13. Williams ST, Busby RC, Demuth RJ, Nelson H (1991) Perineal hidradenitis suppurativa: presentation of two unusual complications and a review. Ann Plast Surg 26: 456-461.