Case Report

Squamous Cell Carcinoma Arising in Hidradenitis Suppurativa, a Rare but Dreadful Complication

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Abstract |

Squamous cell carcinoma (SCC) arising in the setting of long standing hidradenitis suppurativa (HS) is a rare consequence with a high morbidity and mortality. It can often remain unrecognized and delayed diagnosis can lead to advanced stage at presentation and thus a poor outcome. We present a case of HS involving bilateral axillae, groin and natal cleft, for the past five years. The diseased tissue in the natal cleft transformed to SCC. The patient had a stormy course with multiple complications. It was a moderately differentiated tumor with such an aggressive disease course that within a period of five months the patient lost the battle. We suggest a high index of suspicion in long standing HS cases. Early diagnosis, detailed metastatic workup, wide excision, sentinel lymph node biopsy and adjuvant radiation therapy can decrease the morbidity and mortality.

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Introduction

Squamous Cell Carcinoma (SCC) is the most dreadful complication of hidradenitis suppurativa (HS) of perineal and gluteal areas.[1] Chronic relapsing and remitting nature of the disease superimposed with lymphedema plays a pivotal role in metastatic transformation into SCC in long standing HS.[2] We hereby present a case of HS complicated by metastatic transformation into SCC.

Case Presentation

A thirty-eight year old Pakistani male, driver by profession for the past 12 years, presented with a history of hidradenitis suppurativa of both axillae, both groin and natal cleft for past 5 years. He was managed conservatively for most of the areas, except for right axilla for which he underwent surgical excision and coverage with split thickness skin graft in 2018. He presented with Hurley's grade III HS of the natal cleft (Figure 1a & b). Physical examination showed an area of about 16×18 cm, with irregular margins and multiple discharging sinuses and cysts, spanning cranially from the level of sacrum into the natal cleft till the level of coccyx bone caudally, with loss of soft tissue in central area of 8×7cm. Incisional biopsy was performed along with contrast enhanced MRI Scan of the abdomen and pelvis. MRI revealed disease extending from S1 level to the tip of coccyx bone. Multiple linear sinus tracts extending from skin to the natal cleft also involving the gluteus maximus muscles. No coccygeal or sacral osteomyelitis was seen. The biopsy revealed non keratinizing moderately differentiated squamous cell carcinoma. After performing the baseline and metastatic workup, case was discussed in multi-disciplinary team (MDT) meeting. On 22nd July, 2020,resection was performed by the Oncological surgeon with 2cm margin of clearance from the palpable margins of tumor under frozen section control. The deep margin from above the coccyx came out to be positive and additional clearance with negative margin was performed. A defect of about 25×25cm

was created and was reconstructed by plastic surgery team using left inferior gluteal artery rotation advancement flap (Figure 2 a & b).

On consensus of MDT, patient was put on adjuvant radiotherapy; he received 08 fractions of radiotherapy which were completed on 30th September, 2020. His radiation therapy was complicated by wound breakdown and multiple sinuses which were debrided and managed with VAC therapy. On the 5th of October 2020, he presented to the emergency department with swollen and tender right lower quadrant of the abdomen and foul smelling dirty colored discharge from right groin. On investigations he was found to have an extremely low Hb level of 4.9g/dl. The findings on CECT abdomen and pelvis were consistent with the features of necrotizing fasciitis and also showed a mass in the right sacral region of size 48×37mm inseparable from the sacral bone and causing bone erosion. The scan also showed multiple enlarged and necrotic bilateral inguinal and pelvic lymph nodes. Patient underwent aggressive debridement and multiple blood transfusions to build up the Hemoglobin level. He was started on palliative chemotherapy, 1st cycle was given on 27th Oct, 2020. Unfortunately the patient experienced a fatal episode of hemorrhage from femoral artery blow-out and expired on 1st Nov, 2020.

Discussion:

Hidradenitis suppurativa is a chronic inflammatory disorder of the pilosebacious units of the skin with a prevalence of 1% in European population and 0.05% to 0.20% in American population.[3] There is lack of literature about the prevalence of this disease in our part of the world. With a penetrance of 100% this autosomal dominant disease has a variable phenotypic expression.[4,5] The complications associated with this disease range from hyperpigmentation, scarring, fibrosis, lymphatic obstruction, paraneoplastic syndrome and the most dreadful being SCC.[6] The relapsing disease with its associated complications not only affects the patient physically, psychologically and socially but may also cost the patient his life.[1,3] Over 80 cases of HS have been reported to develop SCC, with the average age at presentation being 52.4 years and the mean duration of HS prior to development of SCC being 25years.[7] This disease predominantly affects women, but, there is a male preponderance in SCC transformation.[1,8] Various factors have been postulated to contribute to the process of malignant transformation, with chronic inflammation and epidermal hyperplasia being on top of the list.[1] The chronic lymph stasis in HS disturbs the circulation of immunocompetent cells to the area thus making that tissue immunocompromised and thus prone to malignancy.[2] The predominance of SCC in perineal, perianal and gluteal areas is possibly linked to the presence of bacteria and viruses in these regions. The presence of HPV is one of the established risk factor in ano-genital squamous cell carcinomas.[8] The use of anti-TNF alpha therapy in the management of HS has been reported to contribute to rapid transformation to SCC.[1,8] Smoking also increases the susceptibility of these patients to SCC.[8] Any unexplained neuropathy on background of HS, should alert the clinicians to look for SCC.[7]

Conclusion

In the context of high associated mortality with reported death rate of above 40%[9] with more than 57% of patients dying within the first two years,[1] there should be a high index of suspicion for SCC in underlying HS. Prompt histological diagnosis, metastatic workup using MRI/ PET scan, surgical excision with a minimum of 2cm margins, sentinel lymph node biopsy to rule out occult lymph node metastasis and adjuvant radiation therapy, should all be incorporated as practice guidelines to decrease the associated morbidity and mortality.

Figure I *A; Extent of theLesion, b; Resected Specimen*

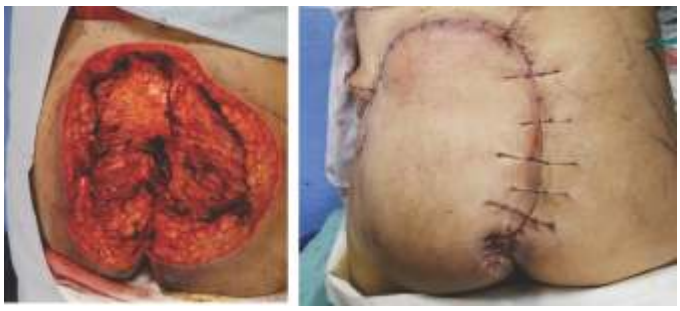

Figure 2: *A; Defect Size, B; Soft Tissue Coverage with Left Inferior Gluteal Artery Rotation Advancement Flap*

Conflict of Interest *None*

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