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Case Report

A Case of Systemic Lupus Erythematosus

Basal Cell Carcinoma in a Case of Systemic Lupus Erythematosus – A Case Report

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Abstract: Systemic lupus erythematosus (SLE) is a chronic, systemic autoimmune with autoantibody production, complement activation, and immune complex deposition. SLE patients have higher mortality rates as compared to the general population Even though there have been improvements in finding and treating diseases, cancer is still a major reason for death in this group. The most common type of cancer is Basal cell carcinoma, and it is widespread. The risk of malignancy in SLE is of considerable interest because the immune and genetic pathways underlying the pathogenesis of SLE and the immunosuppressant drugs (ISDs) used in its management may mediate this altered risk. Here, we present a case of long-standing SLE in an elderly female who was on treatment for the same. She presented with a chronic ulcer over her scalp. She got checked, and a biopsy showed she had Basal Cell Carcinoma on her scalp. Doctors removed it with surgery and covered the open area with skin from another part of her body. At her first checkup, everything looked good, and the new skin was healing well. Even though skin cancers are known to happen in people with certain skin conditions, getting Basal Cell Carcinoma with lupus is very uncommon. In various cohort studies of adults with SLE and overall risk of malignancy, ranging from 1992 to 2017, it was found that though cutaneous malignancies did occur, not a single study reported Basal Cell Carcinoma.

Key Words: Systemic lupus erythematosus, Malignancy, Basal cell carcinoma, Immunosuppression, Wide local excision.

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1. INTRODUCTION

Systemic lupus erythematosus is reported to be one of the most common autoimmune rheumatic diseases. This disease affects various organs and systems. The renal, nervous system, cardiovascular system, skin, mucous membranes, and serous membranes are most commonly involved in the pathological process[1]. Skin is the second most commonly affected site in SLE. The lesions range from malar rash, maculopapular rash, photosensitive dermatitis, subacute cutaneous lupus erythematosus, discoid rash, and lupus profundus. The non-specific lesions of lupus, such as non-scarring alopecia, ulcers of the oral cavity, and vasculitic lesions, are also recorded in literature[2]. Therapy in SLE involves administering systemic glucocorticoids, both as monotherapy and/or along with secondary immunosuppressants. Despite therapy, patients with SLE have higher mortality rates as compared to the general population, as highlighted in various studies, with cancer not being included in the cause of mortality[3]. Apart from the involvement of various systems that contribute to mortality in SLE patients, the treatment itself with immunosuppressants can be a causative factor[4]. Over the last 10 years, numerous cohort studies and meta-analyses have attempted to establish a correlation between SLE and malignant neoplasms, but the results have varied [5-8]. Overall consensus shows that patients with SLE have a significantly increased risk of onco-hematological diseases (especially non-Hodgkin's lymphomas), as well as neoplasms involving the reproductive system in female patients. Numerous studies have demonstrated an association between malignancy and SLE, namely non-Hodgkin's lymphoma, lung, liver, vulvar/vaginal, and thyroid. As for cutaneous neoplasms, there is limited data on this subject. Association with skin cancers such as squamous cell carcinoma, basal cell carcinoma, and melanoma is rare. Basal cell carcinoma cases have not been reported thus far in the literature, making this case a rarity not only in our country. Here we present a case of an elderly female patient who is a known case of SLE on treatment. She presented to the OPD of our institution with complaints of a non-healing ulcer over the scalp for 10 years, which she sustained due to trivial trauma. Despite local, conservative management, the chronic and non-healing nature of the ulcer prompted her to seek medical attention. She was evaluated, her medical history was noted, and after baseline investigations were done, which were within normal limits, she underwent a biopsy from the lesion, which revealed a rarity - basal cell carcinoma of the scalp. She was surgically managed with wide local excision of the lesion with a split skin graft. This report aims to highlight the rarity of the case and enforce the need for prompt investigation, diagnosis, and management of these ulcers, which otherwise seem benign due to their indolent and long-standing nature. Malignant transformation due to chronic scarring, immunosuppressant therapy, inflammation, and ultraviolet radiation exposure all contribute to the pathogenesis of skin cancers in SLE patients. Though cutaneous neoplasms are rare in SLE patients, long-standing, non-healing ulcers, despite other modalities of treatment, must be looked upon with high suspicion.

2. CASE PRESENTATION

2.1. Chief complaints and history of presenting illness

A 73-year-old female patient presented with complaints of an ulcer over the right parietal region of the scalp for 10 years. Sudden in onset with a history of trauma, progressive in nature, associated with loss of hair. It was not associated with pain or any discharge.

2.2. Past medical and family history

She is a known case of SLE on treatment with no other comorbidities or relevant history. She had no relevant family history.

2.3. Examination findings

On examination, a single ulcer of size 7 x 5 cms was present over the right parietal region of the scalp. It was irregular and had raised and rounded margins without fixity to underlying structures.

2.4. Special investigation and diagnosis

A biopsy was done and reported basal cell carcinoma of the scalp.

2.5. Treatment plan and intervention

She was evaluated and baseline investigations were within normal limits. After fitness and consent, she underwent wide local excision of the lesion with a Split skin graft. The post-operative period was uneventful, and evaluation of the graft showed good uptake. The frozen section of the specimen confirmed the diagnosis and revealed margins free of malignancy.

2.6. Ethical Statement

The above case report has been approved by the ethical committee of sree balaji medical college and hospital, Chromepet, after scrutinizing the contents of the case report and the consent form as obtained from the patient ethical number Ref.no: 002/SBMCH/IHEC/2022/1762. Prior written consent for publishing this case report has been obtained from the patient after explaining the in a language understood by the patient.

2.7. Follow up

She is currently on follow up and is showing good progress with excellent graft uptake.

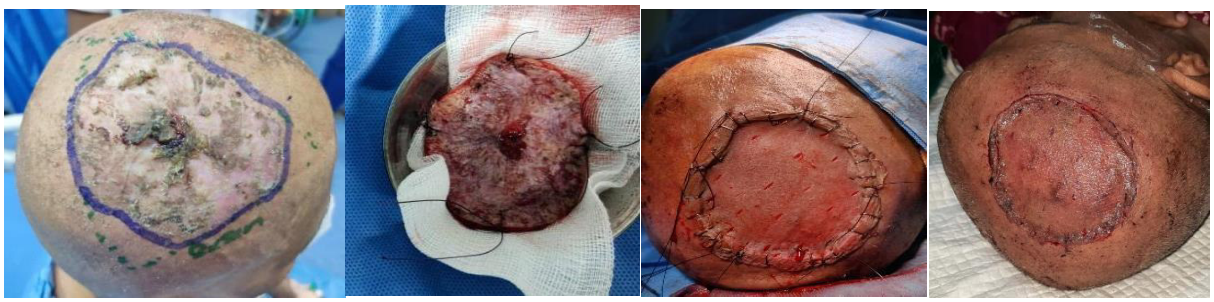

Fig 1: This shows the follow up of an ulcer of the scalp, which is in good progress.

The ulcer's edges are healing, and the tissue beneath them is starting to regrow. The patient is exhibiting indications of improvement, and the wound's degree of infection has decreased. The medication is working effectively on the individual receiving treatment. The patient is instructed to follow the prescribed treatment plan and to keep the wound dry and clean. For additional monitoring and evaluation, it is advisable to visit the doctor frequently. The patient is anticipated to recover fully.

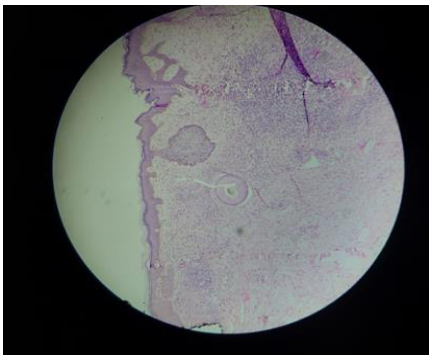

Fig 2: It shows the histopathological studies of the malignancy tissue. It depicts the histology of depigmented patches on the scalp.

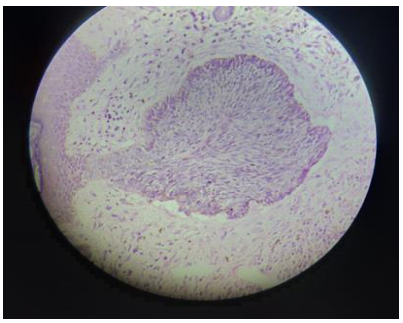

Fig 3: Histopathological slide over the scalp region.

The tissue sample underwent immunohistochemistry and H&E staining (Fig 2 and 3). The tissue tested positive for melanoma-related indicators, according to the findings. A qualified pathologist confirmed the diagnosis as melanoma. The collagen bundles in the cells were infiltrated by lar, pleomorphic, and abnormal melanocytes. Within the cells, there were also sizable melanin aggregates. The results called for more research since they suggested malignancy.

3. DISCUSSION

Systemic Lupus Erythematosus (SLE) is an autoimmune disorder affecting various systems. It can present with various symptoms and presentations, and the course and prognosis of the disease are also variable. Systemic lupus erythematosus increases cancer risk in Asian populations[7]. UV radiation is a risk factor for skin cancer[4]. It can occur as a single episode or multiple recurring and relapsing episodes. It is commonly seen in females in the reproductive age and manifests as generalized symptoms initially. Feldman CH et al. reported that SLE prevalence in females was 6 fold higher compared to males; 38.5% of individuals with SLE were African American, 13.9% Hispanic, 4.2% Asian, 1.5% Native American, and 36.2% were white[10], thus highlighting the uncommon occurrence in Asian, especially the Indian population. Discoid lupus erythematosus (DLE) is a benign skin disorder characterized by red scaly patches and vacuolar degeneration. Malignant transformation is rare[18]. The pathophysiology of DLE is thought to be influenced by genetic and somatic alterations, which lead to the growth of an illegal clone of lymphocytes that produce cellular autoantibodies[19]. Patients with lupus at increased risk of cancer should receive malignancy evaluations[6]. Generalized symptoms include fever, fatigue, generalized body aches, and weight loss despite a normal appetite. Due to the vague nature of symptoms, patients rarely approach a healthcare facility at the early onset of the disease. Systemic involvement produces specific symptoms that prompt the patient to seek medical care. Although uncommon, a subset of SLE called

late-onset SLE affects patients over 50 years, which was the case in our patient. Figure 1, explains the follow-up and good healing progress of the patient's scalp ulcers. Figure 1 and 2 shows the histopathological slides of the malignancy tissue over the patient's scalp. UVR exposure likely initiates skin cancer by inducing oncogenic mutations but does not appear to play a major role in the promotion of DLE-associated skin cancers[9]. A study of the tissue (Fig 2 and 3) revealed mutated cells suggesting malignant melanoma. Immunohistochemistry results confirmed the presence of active melanoma. Treatment and further observation were recommended. The patient was started on chemotherapy and radiation therapy. Follow-up visits revealed positive progress, with the patient responding well to treatment. After 6 months, the patient was declared cancer-free. The patient was monitored closely during the 6 months of treatment, and the scans and biopsies taken during follow-up visits showed that the cancer cells were responding well to treatment and were shrinking in size. This, combined with the patient's excellent response to the treatment, led to the successful outcome of being declared cancer free. The patient was elated at the result and grateful for the care and attention provided by the medical team. This successful outcome is a testament to the effectiveness of modern cancer treatments. Moving forward, the patient will continue to be monitored closely to ensure that the cancer does not return. The disease, where the underlying pathology is chronic inflammation, may enhance cellular dysplasia, leading to uncontrolled tumor cell proliferation 11,12. The mechanism for tumorigenesis in patients with SLE has yet to be delineated. However, an increased risk for Non-Hodgkin's and Hodgkin's lymphoma and laryngeal, lung, liver, vaginal, vulvar, thyroid, and non-melanoma skin cancers (NMSC) have all been reported [13-16]. SLE is related to various cutaneous pathologies, including Discoid lupus erythematosus (DLE). DLE is a benign cutaneous lesion of the head and neck region, and the development of the malignancy (SCC/BCC) over pre-existing DLE rarely occurs. SCC and BCC are feared complications associated with DLE, and BCC occurrence is rarely described in the literature[17-19].

4. CONCLUSION

Elderly patients are most often affected by the disease. Diagnosis of DLE is made mostly by clinical examination, which shows erythematous and scaly patches with healing features associated with the surrounding pigmentary changes. However, due to the indolent course of the disease with a non-painful skin lesion, the diagnosis is often delayed. Patients with high-risk factors, such as elderly patients (>50 years) with exposure to ultraviolet light, can undergo the malignant transformation over the parental DLE. Hence, the most common sites affected are the sun-exposed areas of the face, scalp, forearm, and lip. Chronicity and scarring are considered precipitating factors for malignancy. Thus, prompt and effective treatment is warranted. In refractory cases and suspicious lesions in DLE, whenever they are associated with a coexisting ulcerative or nodular lesion, a biopsy should be considered to rule out possible malignant transformation, even though the possibility of malignancy is rare. Early diagnosis and intervention with appropriate surgical intervention leads to better outcomes. The occurrence of BCC in a patient with SLE, i.e., without prior existing cutaneous lesions, is yet to be reported in the literature.

5. AUTHORS CONTRIBUTION STATEMENT

Dr. K. S. Ravishankar proposed the concept, interpreted the data, and critically edited the article. Dr. Suruchi Rubina, Dr. Manimaran, and Dr. Rathnaganapathy interpreted the data and reviewed the literature.

6. CONFLICT OF INTEREST

Conflict of interest declared none.

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