Rare leiomyosarcoma in a burn scar

**ABSTRACT**

Leiomyosarcomas presenting as a cutaneous malignancy in a burn scar are rare due to scarce mesenchymal elements within the cutaneous tissues. We here present a case of a 73-year-old woman with a past history of burns, came with a recurrent ulceroproliferative lesion in the right lumbar region over previous burn scar. She had a history of a similar lesion in the same region 1 year ago, for which wide local excision and grafting were done, and histopathology revealed as sarcomatoid carcinoma. Examination revealed multiple right axillary lymph nodes. Evaluation was done with contrast-enhanced computed tomography to rule out systemic metastasis. She was managed surgically with wide local excision, right axillary dissection, and split skin grafting. She had aggressive local recurrence along with lung metastasis within 6 months and was lost to follow-up. This report presents the second case of leiomyosarcoma arising in a burn scar on the right flank.

**Keywords:** Burn scar, latent period, leiomyosarcoma

**INTRODUCTION**

Cutaneous malignancy in chronic burn scar has been reported in the literature, with squamous cell carcinoma (71%) being the most common followed by basal cell carcinoma (12%), melanoma (6%), and rarely sarcomas (5%). Sarcomas are rare due to the deeper location of mesenchymal cells in the dermis or subcutaneous tissue making it less vulnerable to trauma and lesser requirement of tissue regeneration when compared to the epidermis. Leiomyosarcoma is rarer with one case reported so far. We report the second case of leiomyosarcoma in burn scar over the right flank. The first case had been reported in 1998 by Can et al.

**CASE REPORT**

A 73-year-old female with a history of burns at the age of 10 years presented with recurrent painless ulceroproliferative lesion in the right flank over the burn scar [Figure 1] for 3 months. She had similar lesion in the same region 1 year ago, for which she underwent wide local excision with split skin grafting. Histopathology was suggestive of sarcomatoid carcinoma. On examination, 8 cm × 9 cm ulceroproliferative, nontender growth over the burn scar with everted margins and surrounding induration was found. Multiple, large, mobile, firm-to-hard right axillary nodes were palpable with normal overlying skin. The patient was evaluated with contrast-enhanced computed tomography of the thorax and abdomen which showed few well-defined lobulated partially exophytic, heterogeneously enhancing soft-tissue lesions involving the skin and subcutaneous tissue of the right hypochondrium and flank regions, showing ulcerations with infiltration into the external oblique muscle suggesting malignant lesions [Figure 2]. Multiple enlarged heterogeneously enhancing necrotic lymph nodes in the right axillary region were likely metastatic [Figure 3]. She was scheduled for elective surgery and wide local excision of the lesion with 2-cm margin, right axillary dissection [Figure 4], and split skin grafting [Figure 5] was done. Routine histopathological examination showed ill-circumscribed tumor composed of sheets, interlacing fascicles of spindle cells with hyperchromatic nuclei and increased mitosis.

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cells with elongated to plump oval nuclei, coarse chromatin, prominent eosinophilic nucleoli, moderate eosinophilic cytoplasm, and mitotic figures (3–5/10 HPF), with atypical mitosis surrounded by fibrocollagenous stroma with smooth muscle fibers suggestive of inflammatory leiomyosarcoma. On immunocytochemical staining, tumor cells are focally positive for desmin [Figure 6]; diffusely positive for vimentin [Figure 7]; and negative for cytokeratin, smooth muscle actin, and anaplastic lymphoma kinase confirming diagnosis of leiomyosarcoma. The patient had aggressive recurrence with lung metastasis within 6 months and was lost to follow-up.

**DISCUSSION**

Marjolin was the first person to describe regarding malignant transformation in scars, hence called Marjolin ulcer. The pathogenesis of malignant transformation in burn scar is, however, uncertain. Dermal and subdermal tissues have embryonic mesodermal cells which consist of primitive mesenchymal cells, fibroblasts, and histiocytes and usually protected by the epidermis from the external agents such as infections, physical or chemical damage, and radiation. When tissue damage occurs due to ulceration, protective barrier is lost and damage of the underlying mesodermal cells occurs. In the process of regeneration, there is acceleration of turnover of constituent cells and there is increased possibility of mistranscription of DNA, and thus, malignant transformation begins. This could be one of the underlying pathogeneses in malignant transformation of a burn scar.

Fleming and Rezek suggested that deeper tissues are usually subjected to lesser trauma due to the overlying protective barrier and thus lesser tissue regeneration when compared to the superficial epidermis; thus, carcinomas are more common. The appearance of sarcoma is very rare as such when cases are found, they are worthy of note on this basis itself. Fourteen cases of sarcoma in burn scar have been reported in the literature so far.
Among sarcomas, the most common is fibrosarcoma with four cases reported in the literature so far followed by malignant fibrous histiocytoma, with three cases being reported. The reason behind this is that both of them arise from undifferentiated mesenchymal cells and fibroblasts which are abundant in skin scar tissue. Our patient had lesion 1 year ago which was sarcomatoid carcinoma which could have occurred from the process of epithelial to mesenchymal transition.\(^\text{[4]}\) It is a complex process where epithelial cells lose their polarity and cytoskeletal remodeling occurs with the acquisition of mesenchymal components and manifestation of migratory phenotype.\(^\text{[3]}\) These cells have the ability to breakthrough basement membrane\(^\text{[6]}\) and invade blood vessels and lymphatics, thereby leading to distant metastasis. Pathogenesis of development of other types of sarcomas however is not described in the literature.

Lawrence reported that the age of the patient at the time of burn appeared to be inversely proportional to the induction period of malignancy.\(^\text{[7]}\) A latent period of 40–50 years is required for malignant transformation in a patient with burns before the age of 5 years. In burn scar sarcomas, the mean time between the occurrence of burns and development of malignancy was 35.7 years with age range 3–71 years, whereas in burn scar carcinoma, the latent period is less with early onset of malignancy. The earliest onset of postburn sarcoma was 3 years which was mesenchymal malignancy.\(^\text{[8]}\) Prognosis of postburn sarcomas is very poor considering all cases of sarcoma that have been reported, as majority of the patients had distant metastasis within 1–2 years followed by death despite adequate excision of lesion. Disease-free period noted in majority of the cases is only 6 months to 1 year.

In 1981, Fields and Helwig reported 65 primary cutaneous and 15 subcutaneous leiomyosarcomas. Although exact etiology is not known, Fields and Helwig recorded a history of trauma in 15 of 80 patients.\(^\text{[9]}\) Therapeutic irradiation has also been reported as one of the causes. A case of leiomyosarcoma arising in a smallpox vaccination scar was also reported.\(^\text{[10]}\) However, only one case of leiomyosarcoma following a burn scar has been reported till date.

Treatment of burn scar sarcoma is a wide local excision with negative lateral margins and depth including subcutaneous tissue and fascia. Despite this, tumors have a high local recurrence rate with eventual distant metastasis suggesting the poor outcome of the disease. For cases with high risk of recurrence which include tumors larger than 5 cm, high mitotic rate, positive surgical margins, and subcutaneous extension of dermal tumor, radiation therapy is considered. Both radiotherapy and chemotherapy should be reserved for cases of recurrence or metastasis. Classical regimens include doxorubicin, ifosfamide, gemcitabine, docetaxel, and dacarbazine. A combination of gemcitabine and docetaxel has shown the best cure rate against metastatic leiomyosarcoma. The behavior of sarcomas is not well established due to low incidence and very little literature demonstrating the importance of reporting such findings in an effort to better understand the disease and provide the most appropriate management.
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**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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