



ISSN: 0975-833X

REVIEW ARTICLE

A RARE CASE OF MARJOLIN'S ULCER WITH A SHORT LATENT PERIOD

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ARTICLE INFO

Article History:

Received 15th September, 2015
Received in revised form
27th October, 2015
Accepted 27th November, 2015
Published online 21st December, 2015

Key words:

SCC, Marjolin's Ulcer,
Biopsy, Non Healing Ulcer,
Burn.

ABSTRACT

Marjolin's ulcers (MU) are rare malignancies arising from chronically inflamed, or scarred skin especially in burn wounds. Persistence of burn ulcer, induration or elevation of margin of such ulcer, ulceration or nodule formation over a burn scar may indicate malignant transformation. The commonest malignancy arising in MU is squamous cell carcinoma (SCC). Common clinical presentation is flat indurated ulcer and less frequent presentation is exophytic papillary growth. Diagnosis is made by tissue biopsy from multiple sites including the margins of the lesion. These tumors are aggressive and have poor prognosis. Here we report a case of MU in a 45 year female, who presented with non healing ulcer in a neglected burn wound after short latent period i.e. within six months of burn injury and biopsy taken from multiple sites from the ulcer showed a well differentiated SCC. Wide surgical excision was advised, but the patient lost to follow up.

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Citation: Dr. Kishori Moni Panda, 2015. "A rare case of marjolin's ulcer with a short latent period", *International Journal of Current Research*, 7, (12), 23664-23666.

INTRODUCTION

Currently, MU is used for carcinomas arising from chronically inflamed, or scarred skin. It is a rare type of cancer. The incidence varies due to sociocultural and economic influences. Earlier reported incidence of 1.2-2% mostly constitutes carcinomas arising from burn scars. About 6.8% incidence have been reported amongst the Kashmiri Indians, where "Kangri ulcers" occur due to repeated burn injuries. Similar ulcers called "Kairo burn cancer" are also reported in some parts of Japan. In North-western China, the term "Kang Ulcers" is used for MU arising from burn injuries. "Erythema ab Igne," a chronic thermal dermatitis also occasionally undergoes malignant change. A higher incidence of 63% have been reported in some series of MU, due to chronically irritated or scarred skin (Chase Tobin *et al.*, 2014; Ali Akber Mohammadi *et al.*, 2013). MU occur predominantly in middle age, although they have been reported in every age group. Sisrat and Shrikhande have reported average age of onset 44 years in India. Asuquo *et al* have reported 44.1 years as the average age of onset in south eastern Nigeria. Nthumba recently reported an average age of onset of between 36 and 42 years in Sub Saharan Africa. These averages may not be truly representative, as many variables such as aetiologic agent, and age at time of onset of the predisposing condition, influence the age of onset of MU.

The latent period, that is the time from onset of the predisposing condition, to the development of cancer, has been shown to be shorter for the elderly and longer for the young patients (Ali Akber Mohammadi *et al.*, 2013; Kingsley O Opara and Otene, 2011).

The diagnostic criteria for MU was established by Ewing & also adapted by Treves and Pack include the following

- The cancer must arise within the boundaries of the scar or wound.
- There should be evidence of trauma or pre-existing ulcer as evidenced by the wound or scar.
- The absence of any precursory or similar neoplasm on the site before development of MU.
- The histologic variety of MU must be compatible with the native tissues.

The interval time between the trauma/ulcer and the onset of the cancer must be appropriate. A period of one month has been proposed as the minimum acceptable time between the trauma/ulcer and the onset of the cancer (Kingsley O Opara and Otene, 2011; Treves and Pack, 1930). Here a case of acute MU arising in a neglected burn wound in a 45 year female with a short latent period of less than six months is reported.

CASE REPORT

A 45yr old female presented to surgery OPD with a non-healing ulcer since 6 months following burn injuries in her

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right forearm and complained of bleeding on contact. On local examination ulcer was (2.5x2) cm, irregular, unhealthy, foul smelling with indurated base and focally covered with slough and blood clots. There was no regional lymphadenopathy. A provisional diagnosis of marjolins ulcer was made and biopsy was taken from multiple sites. Grossly 5 gray white irregular tissue bits measuring (1.5x1) cm received for histopathological examination. Microscopically majority of tissue bits showed proliferating nests of atypical dyskeratotic squamous cells infiltrating superficial dermis with squamous pearl formation, consistent with well differentiated SCC (Fig. 1 & 2). one tissue bit showed pseudoepithelial hyperplasia (Fig. 3). Peritumoral lymphocytic infiltration was also seen.

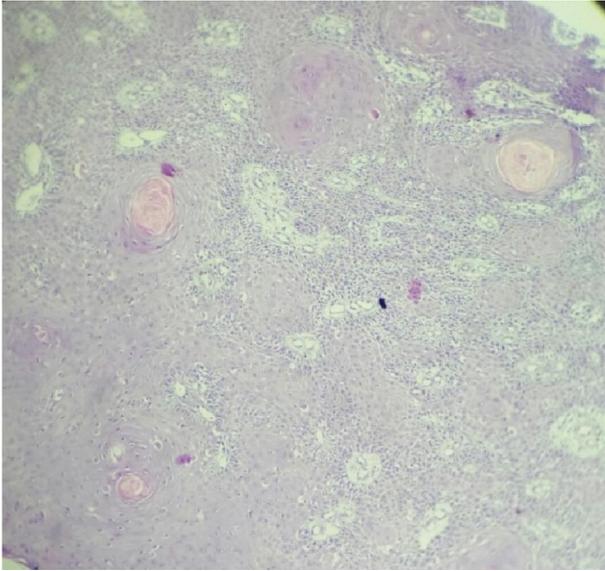


Fig. 1. Well differentiated SCC Infiltrating superficial dermis with pearl formation (HEX10)

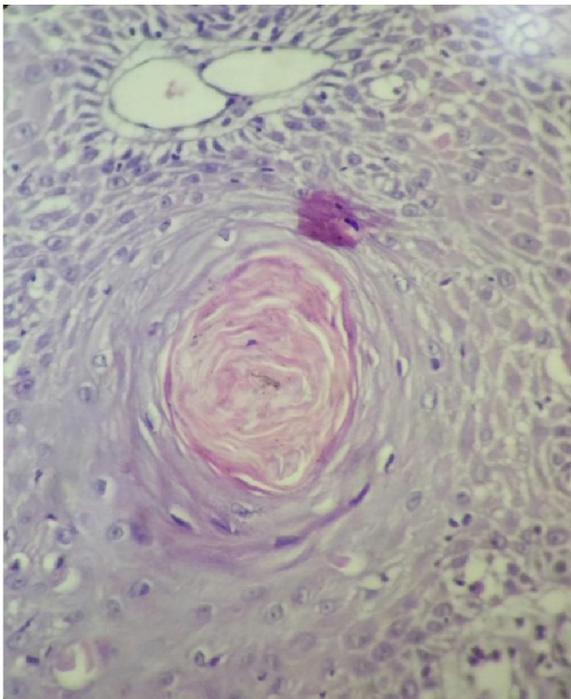


Fig. 2. Malignant squamous cells with pearl formation (HEX40)

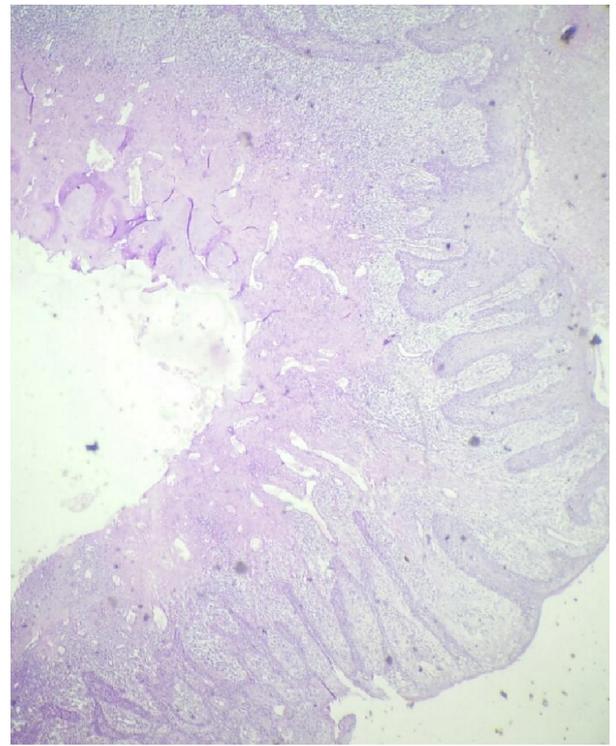


Fig. 3. Pseudoepithelial hyperplasia in burn ulcer (HEX10)

DISCUSSION

It was Celcius who first observed malignant transformation in burn scars in 100 AD. Jame Nicholas Marjolin in 1828 published the classical description of MU. The term 'MU' was first used by Da Costa in 1903. MU can be clinically described as acute with a latent period of less than one year, and chronic longer than a year. The commonest region of body involved are the lower limb (40%), head & neck (30%) and (20%) for the upper extremities. Treves and pack describe two clinical types of MU. Our case belonged to the commonest form, which is flat indurated, ulcerative MU with poor prognosis. The less frequent exophytic papillary form has good prognosis (Kingsley O Opara and Otene, 2011; Treves and Pack, 1930; Muhammad Saaiq *et al.*, 2014).

MU typically take years to develop, but occasionally, acute onset i.e. latent period of less than 12 months has been documented. Latency has been described as the time between initial injury and the confirmation of a pathologic diagnosis of MU. The report by Baskara *et al* which describes the development of an acute MU in a 72-year-old within 9 months of a plantar pressure ulcer. In our case acute MU developed within 6 months of burn injury (Baskara *et al.*, 2010).

Histologically, seventy-one percent of Marjolin's ulcers are squamous cell carcinomas, although basal cell carcinoma, melanoma, fibrosarcoma, angiosarcoma, liposarcoma, leiomyosarcoma, osteosarcoma, dermatofibrosarcoma protuberans, malignant fibrous histiocytoma, malignant schwannoma, and other malignant mesenchymal tumor have also been described. When MU occurs in its squamous cell form it is a very aggressive malignancy but constitutes only 2%

of all SCC. Histologic diagnosis in this case was well differentiated SCC, which is the commonest type (Chase Tobin *et al.*, 2014; Marjolin's, 2014).

MU can be suspected, when non-healing ulcer develops in an area of abnormal or scarred skin. However, it should be confirmed histologically by taking tissue biopsy from multiple locations including the margins to minimize a false-negative finding. Most common cause of false negative diagnosis is pseudoepitheliomatous hyperplasia, which is a common finding in chronic ulcers and often coexists with MU as seen in our case. Also, MRI is helpful in determining the extent of local invasion of MU on the extremities (Chase Tobin *et al.*, 2014; Kingsley O Opara and Otene, 2011; Marjolin's, 2014).

The reported causative agents include burn and non-burn traumas, osteomyelitis sinuses, pressure sores, urinary fistulas, pilonidal sinuses, gumata, radiation scars, and scars from flogging, etc. The pathogenesis of this cancer is not completely understood, but appears to be multifactorial, affected by both environmental and genetic factors. Key pathogenetic factors are slow healing process and chronic instability of scar tissue. MU occurs in locations where there is constant trauma or a compromised blood supply. It has been suggested that in the chronic wound, decreased vascularity combined with weakened epithelium creates a susceptibility to carcinogens. It has also been proposed that scar tissue is an immunologically privileged site because of relative avascularity and inadequate cell-mediated immunity (CMI). Direct mutagenic effect of released toxins by lysis of scar tissue is also carcinogenic.

Mutations in the p53 gene and Fas gene may cause dysregulated apoptosis and cell homeostasis, respectively, and have been documented in patients with MU. Chronic inflammation and repair provide a continuous stimulus for cellular proliferation and may increase the rate of spontaneous mutations. These findings support the idea that chronic irritation or inflammation is an inciting factor (Chase Tobin *et al.*, 2014; Kingsley O Opara and Otene, 2011). Tumor type, location and rate of metastasis affect prognosis. The overall survival for patients with MU is 65%-75% at 3 years post diagnosis, but falls to 35%-50% if there is metastasis. Poor prognosis is associated with palpable regional lymphadenopathy, which predicts death within 2 years.

The poor prognostic indicators on histology are poorly differentiated carcinoma, scarce or absent peritumoural T cell infiltration, invasion of reticular dermis or deeper structures, and ≥ 4 mm vertical thickness of the neoplastic lesion. Prevention is by early excision and grafting of deep burns adequately in acute stage (Chase Tobin *et al.*, 2014; Kingsley O Opara and Otene. Marjolin's, 2011; Novick *et al.*, 1977). Wide local excision and wound coverage with skin grafting or flaps is the treatment of choice.

Currently there is no universal consensus or treatment protocol regarding excision margins, lymph node dissection, or the use of neoadjuvant radiotherapy or chemotherapy. A combination of these procedures is often necessary. Amputation may be required in extremity lesions where there is invasion of major neurovascular structures, when adequate surgical margins are not possible, or if there is aggressive recurrence. In patients with inoperable metastatic disease, palliative adjuvant radiotherapy and chemotherapy can be useful as well. Lymphadenectomy is required if there is persistent adenopathy (Chase Tobin *et al.*, 2014; Kingsley O Opara and Otene, 2011; Novick *et al.*, 1977).

Conclusion

MU can present as non-healing ulcer in neglected burn wounds. Regular monitoring and prompt biopsy in suspected cases helps in early diagnosis and improved outcome. Awareness about early excision and grafting of deep burns adequately in acute stage could prevent this deadly complication.

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