

Two Different Cases of Marjolin's Ulcer and Recommendations for Practice

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Abstract: Marjolin's ulcer is a malignant tumor developing in the chronic skin lesion (e.g. burn scar, vaccination scar, non-healing wound). We report two histological different cases: ulcerated squamous cell carcinoma growing in the burn scar and basal cell carcinoma arising in the vaccination scar. We present clinically-oriented literature review and management recommendations. In general all non-healing chronic wounds should be surgically closed and large open wounds must not be left for secondary intention healing to prevent delayed malignant transformation. All burn scars should be monitored by appropriately informed patient and family physician. All suspected masses within chronic wounds/burn scars should be promptly biopsied. If Marjolin's ulcer diagnosis is established, wide local excision comprising fascia should be performed as well as careful clinical assessment of regional lymph nodes.

Keywords: Marjolin's ulcer, malignant, skin lesion, carcinoma, lymph nodes.

INTRODUCTION

Marjolin's ulcer is a malignant tumor developing in the chronic skin lesion (burn scar, vaccination scar, non-healing wound etc.). We present two histological different cases of this disease: squamous cell carcinoma arising in the burn scar and basal cell carcinoma developing in the vaccination scar. We shortly discuss current data on Marjolin's ulcer epidemiology and its clinical characteristics. As there is no consensus on the management of this disease, we present subjective management recommendations based on clinically-oriented literature review [1-10].

CASE #1

63-years old Caucasian woman presented with a large ulceration (6 x 10 cm) on the lateral surface of her left thigh (Fig. 1). Her both legs and buttocks were burned when she was 7-years old. Ulcerated area was bleeding on contact and had elevated margins. Incisional biopsy was performed, and the diagnosis of squamous cell carcinoma (SCC) with extensive suppurative changes was made. The patient was referred to regional cancer center for further treatment. On CT 3 cm thick superficially spreading lesion with no signs of large vessels/major nerves/joints/bones invasion was seen. No enlarged regional (iliac or inguinal) lymph nodes were noted neither physically nor on CT. No distant metastases were present on chest X-rays and abdominal imaging. The ulceration was excised with 2 cm clear margin (the specimen included femoral fascia). The wound was closed with rotated

skin-subcutaneous flap. The postoperative course was uneventful and the patient is followed-up for 14 months with no signs of recurrence.



Fig. (1). Ulcerated squamous cell carcinoma arising within burn scar on left thigh.

CASE #2

67-years old Caucasian man was referred to the regional cancer center with firm nodule on his left arm arising in a childhood smallpox vaccination scar. The nodule appeared 4 months ago and grew quickly to the diameter of 1 cm; the surface of the tumor was minimally ulcerated (Fig. 2). Incisional biopsy was performed, and microscopically basal cell carcinoma was diagnosed. Apart from the lesion physical examination and imaging studies were normal. Subsequently the lesion was excised with 1 cm clear deep and lateral margins. No further treatment was necessary and

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patient remains in follow-up for 8 months with no symptoms of relapse.



Fig. (2). Basal cell carcinoma arising in vaccination scar on left arm.

DISCUSSION

The association of burn injury and delayed development of malignancy was documented 2000 years ago by Celsus [1]. The first modern description of the condition discussed here was presented by French surgeon Jean Nicholas Marjolin, who in 1828 observed and classified cellular changes in burned skin (“*ulcère cancroïde*”) [1, 2]. Today Marjolin name serves as an eponym (Marjolin’s ulcer) for skin malignancies (mainly squamous and basal cell carcinoma, but other histology is possible) arising in old burn scars, chronic wounds and other chronic skin conditions, including vaccination scars and frostbites [1-4].

In 2009 160 000 cases of squamous cell carcinoma were expected in the United States. Because about 2% of all squamous cell carcinoma arise in the burn scars [1], at least 3200 new Marjolin’s ulcers per year appear in the USA [5]. Squamous cell carcinoma is the most frequent (70%; see case #1, Fig. 1), but not the only malignancy developing in the chronically wounded (e.g., burned) skin. Other malignancies known to occur in this setting are: basal cell carcinoma (12% of all Marjolin’s ulcers; see case #2; Fig. 2), melanoma (5%) and sarcoma (4%) [1].

According to the recent literature the transformation from non-healing wound/scar to the malignant disease is slow, ranging from 20 to 40 years (“chronic Marjolin’s ulcer”) [1, 2, 6]. Seldom rapidly progressive cases were noted (“acute Marjolin’s ulcer”), with malignant transformation occurring within few weeks or few years [1, 2, 6]. The latency period is inversely proportional to patient’s age at the time of skin injury [1]. The pathophysiology of Marjolin’s ulcer development is distinct from regular squamous cell carcinoma. Local ischaemia, decreased immunity of the affected skin/scar tissue, fragile epithelium exposed to repeated injuries are among main triggers of neoplastic transformation [6]. The skin lesion underlying development of Marjolin’s ulcer is predominantly burn scar (75%), traumatic non-healing wound (8%), venous stasis ulcer (6%), pressure ulcer (3%) and other (e.g., frostbite, vaccination scar) [3, 4]. Hot oil

burn scars are associated with higher risk of developing more aggressive lesions [7].

The Marjolin’s ulcers usually locates on the extremities: majority of 443 cases analyzed by Kerr-Valentic *et al.* were located on the lower extremities, 20% on upper extremities, and the rest on the torso and head [2].

Marjolin’s ulcer with squamous cell histology is usually more aggressive compared to typical non-melanoma skin cancers. The risk of regional lymph node metastases exceeds 22-25%, and the risk of distal dissemination – 6-14% [1, 2]. Five and ten years survival rate for Marjolin’s ulcer is lower compared to typical squamous cell carcinoma, respectively 52% vs 85% and 34% vs 60% [6]. Inferior results can be also attributed to the lack of vigilance, frequent overlooking of the lesion growing in the chronically wounded skin and inadequate treatment (e.g. non-radical excision) [8].

As there is no consensus on the management of Marjolin’s ulcer with squamous cell histology, we present subjective recommendations for practice based on clinically-oriented literature review (Table 1) [1-10]. In brief it is authors’ subjective opinion that: all chronic wounds should be closed surgically (preferably with skin graft or skin-muscle flap) and large wounds must not be left for secondary intention healing. All burn scars from childhood should be monitored by appropriately informed patient and primary care physician. All suspected masses visible and/or palpable

Table 1. Recommendations for Prevention, Early Diagnosis and Treatment of Aggressive Squamous Cell Carcinoma Arising in Burned or Chronically Wounded Skin (Marjolin’s Ulcer) Based on Literature Review [1-10] and Authors’ Opinion

1. Excise and (if possible) primary close all chronic wounds (preferably with skin graft or skin-muscle flap)
2. Closely and regularly monitor all childhood burn scars and chronic wounds which cannot be surgically closed (and inform your patient about risk of Marjolin’s ulcer to increase her/his vigilance)
3. Prevent and treat all infections in chronic wounds
4. Do not delay incisional biopsy of any suspected [crusting, ulcerated, painful, growing, bleeding] lesion visible/palpable within the chronic wounds/vaccination scars/burn scars (biopsy both central and the marginal zone)
5. Any venous ulcer unresponsive to therapy for >3 months should undergo incisional biopsy to rule out malignancy
6. Pay attention to the regional lymph nodes, as the risk of lymph node involvement is higher compared to regular skin cancer (the risk of distant metastasis is also higher)
7. Excise confirmed squamous cell type Marjolin’s ulcer with wide margins of at least 2 cm and include fascia with the specimen
8. Perform axillary/inguinal lymphadenectomy only if lymph nodes are suspected on palpation or by imaging modalities (sentinel node biopsy [§] procedure is not fully validated, but could be performed if regional nodes are clinically negative)
9. Amputation is rarely an option, but cannot be excluded in very large primary/recurrent lesions with erosion of the large vessels and/or major nerves, loss of extremity function, invasion of joint cavities/bones
10. Radiotherapy and/or chemotherapy could be instituted on individual basis (mainly in advanced cases)

within chronic wounds/vaccination scars/burn scars should be promptly referred to surgical consultation and biopsied. Every venous ulcer should be biopsied if non-responsive to therapy after 3 months [10]. If diagnosis of Marjolin's ulcer with squamous cell carcinoma histology is established, excision with 2 cm lateral margins, comprising fascia, should be performed as well as careful clinical (physical and imaging) assessment of regional lymph nodes [6]. Regional (usually axillary/inguinal) lymphadenectomy should be performed only if lymph nodes are suspected on palpation or by imaging modalities. (Some authors advocate sentinel node biopsy [8] if regional nodes are clinically negative). Patient should remain in a postoperative follow-up program, as there is a constant risk of recurrence, particularly in case of squamous cell histology.

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