ORIGINAL ARTICLE

# Marjolin's Ulcer: A Preventable Cancer

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**Abstract**

Marjolin's ulcer is cutaneous malignant tumor arising from chronically inflamed or traumatized skin. It commonly occurs in burn scars but can also originate from venous ulcers, pressure sores and chronic osteomyelitis sinuses.

The major risk factors for development of neoplasms are healing by secondary intention, non healing wounds and fragile scars that ulcerate easily.

Our series reviews 17 cases of Marjolin's ulcer, who presented at Plastic Surgery Department Jinnah Hospital Lahore from June 2005 to June 2011. In 11 cases Marjolin's ulcer occurred after burns and six after traumatic wounds. In 16 cases wound had healed by secondary intention. After establishing diagnosis by incisional biopsy, 16 cases underwent wide excision and skin graft or flap for coverage. Below knee amputation was performed in one patient. Three patients had recurrence and above-knee amputation was performed in one patient, he is disease free one year after the amputation. Two patients died who had recurrence over scalp. Mean follow up period was 3 years. Three patients were lost to follow-up and 12 cases were tumor-free during the follow-up period.

We concluded that cure rate for early cases is high as compared with advanced cases. Marjolin's ulcer can be prevented by promoting wound healing by primary intention and early coverage of wound defects with graft or flap.

***Key Words:*** *Preventable cancer*

## Introduction

Marjolin's ulcer is cutaneous malignant tumor arising from chronically inflamed or traumatized skin. It commonly occurs in burn scars but can also originate from venous ulcers; pressure sores traumatic wounds, cystostomy sites, scarring from lupus, amputation stumps, chronic lymphedema, chronic pilonidal sinuses, hidradenitis suppurativa, chronic ulcers of leprosy and chronic osteomyelitis sinuses.

Malignant degeneration in post burn scar was first described by French Surgeon Jean-Nicolas Marjolin in 1828. In 1903, DaCosta suggested that carcinomatous change takes place in a chronic ulcer; indurations usually begin from around the margin and spread slowly. It is rare for the entire margin of a large ulcer to transform into a malignant disease.[1]

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Incidence of Marjolin's ulcer is 1.7% in chronic wounds.[2] All parts of the body can be affected but it most frequently occurs over the scalp and extremities.

Exact pathogenesis is unknown. Marjolin's ulcers are thought to be due to long-term, continuous mitotic activity as the epidermal cells attempt to resurface the open defect.[3] This cycle of damage, irritation, and repair especially in flexion creases, can lead to a malignant transformation.

Two variants have been recognized. In Acute variant malignant degeneration occurs within one year. In Chronic variety, it occurs after more than one year. Latent transformation period for chronic variant ranges from 25 to 40 years .[4]

The most common histological type is squamous cell carcinoma. Other types include basal cell carcinoma, Malignant fibrous histiocytoma, malignant melanoma, liposarcoma, fibrosarcoma, neuroendocrine (Merkel cell) carcinoma, and keratoacanthoma.[5-8] SCC developing in a Marjolin's Ulcers are reported to be more aggressive than other skin cancers of the same cell type arising de novo[9] Prognosis worsens with higher grades; thus

Lifeso[10] et al. described three grades (well differentiated, moderately differentiated and poorly differentiated. Marjolin's ulcers have 30-40% rate of metastasis[11].

## Materials and Methods

We retrospectively analyzed the case records of 17 patients of Marjolin's ulcer, who presented at Plastic Surgery Department Jinnah hospital Lahore from June 2005 to June 2011.Diagnosis was made on the basis of history, clinical examination and biopsy. Preoperative evaluation with CT scan or MRI was done for local extent of tumor and lymph nodes metastasis while distant metastasis was ruled out by CT scan, ultrasound abdomen, X ray chest and bone scan (if required). Tumor grading and histological type was determined from biopsy. In 16 cases Tumors were excised with 2cm wide margin of clinically involved area and after biopsy proven tumor negative margin on frozen section coverage was provided with graft or flap. Below knee amputation was done in one case due to lower tibia involvement. Post operative radiotherapy was given in patients who had tumor size more than 10cm ,regional lymph node involvement and high grade tumors(grade 3).All patients were followed up for recurrence.

## Results

Initial cause of injury was burns (70%), traumatic wounds (24%) and chronic osteomyelitis (6%) Seven patients were male and ten were female. Mean age of presentation was 48 years (22-68 years).

Latent period ranged from 8 months to 50 years with mean of 28 years. 76% of tumors were located over extremities, 18% over scalp and 6% over trunk. Histological types were Squamous cell carcinoma (94%) and basal cell carcinoma (6%). One patient (6%) developed acute Marjolin's ulcer. Rest had chronic Marjolin's ulcer (94%).Lymph node involvement and distant metastasis was not seen in any case. (Table 1)

**Table 1**

| Total Patients | 17 |
|---|---|
| Study location | Jinnah Hospital, Lahore |
| Time Period | 2005-2011 |
| Gender | 7 Male: 10 Female |
| Age | 18-68 years (mean, 48) |
| Latent Period | 1-50 years, (mean, 27) |
| Cause of initial injury | Burn 70% ,Trauma 24%, Ch Osteomyelitis 6% |
| Most common type | SCC -94%, BCC-6% |
| Most common location | Extremities 76%, Scalp 24% |
| Lymph node involvement | None |

Initial injury had healed by secondary intention in 94% cases. In 16 cases wide local excision with 2 cm margins was done. One case had involvement of lower tibia and below knee amputation was done. In seven (41%) cases defect was covered with skin graft, nine (59%) patients needed flap coverage .Post operative radiotherapy was given in seven cases (41%).

All patients were followed up for recurrence ranging from 1.5 to 3.5 years with mean period of follow-up of 3 years. Three patients (18%) had recurrence. Mean period of recurrence was 6 months. Two patients with scalp Marjolin's ulcer had recurrence and died with aggressive involvement of dura and brain. One patient had recurrence at knee joint and above knee amputation was done and he is disease free for past one and half years. Three patients (18%) were lost to follow up.12 cases (70%) are disease free now. (Table 2)

**Table 2**

| Initial wound healing | Secondary intention 94%<br>Skin graft 6% |
|---|---|
| Tumor excision | Wide local excision, 16 (94%)<br>Below knee amputation,1 (6%) |
| Wound coverage | 7 cases Skin graft (41%)<br>9 cases Flap (59%) |
| Post-op Radiotherapy | 7 cases (41%) |
| Follow up | 1.5-3.5 years (mean, 3 years) |
| Recurrence | 3 cases (18%),<br>mean period 6 months |
| Lost to follow up | 3 cases (18%) |
| Died | 2 cases (12%) |
| Disease free | 12 cases (70%) |

## CASE 1

40 years female presented with Marjolin's ulcer popliteal region for 03 years. She had history of flame burn twenty years back. There was no lymph node, bony or neurovascular involvement. Incisional biopsy revealed low grade Squamous cell carcinoma. Wide local excision of tumor with 2cm margin done and defect covered with skin graft.post operative radiotherapy was not given and patient is disease free for past 02 years. (Fig 1-4).

Pre-operative picture of a patient with Marjolin's Ulcer Left Popliteal area

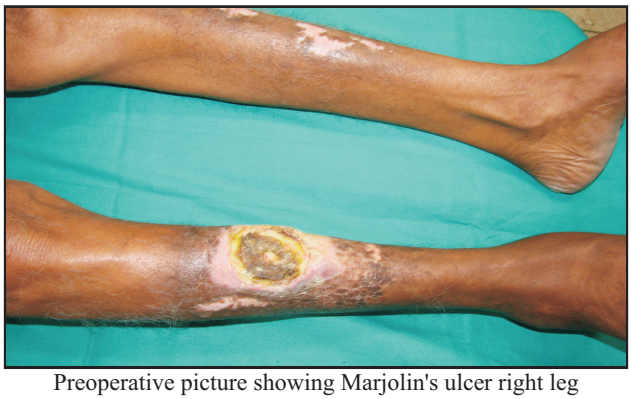
Per-operative picture showing the ulcer has raised irregular borders and surrounding area is hypo pigmented

Intra-operative picture showing 2cm wide excision margin

Intra-operative picture showing split thickness skin grafting over popliteal area

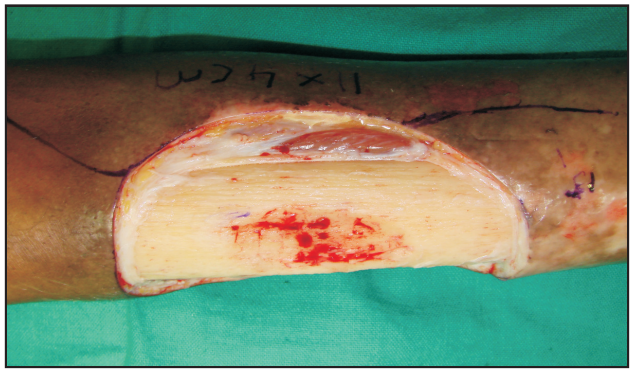
## Case 2

60 years male presented with non healing ulcer over right middle third of leg for past 03 years. He had history of flame burns 40 years back which healed by secondary intention. Preoperative evaluation didn't show lymph node or bony involvement. Histology revealed moderately differentiated SCC.Tumor was excised with 2cm margin. Tibia was exposed in middle third of leg. After negative resection margin confirmation by frozen section, defect was covered with proximally based medial hemisoleus flap. Post operatively patient received radiotherapy and he is disease free for past three years. (Fig 5-9).



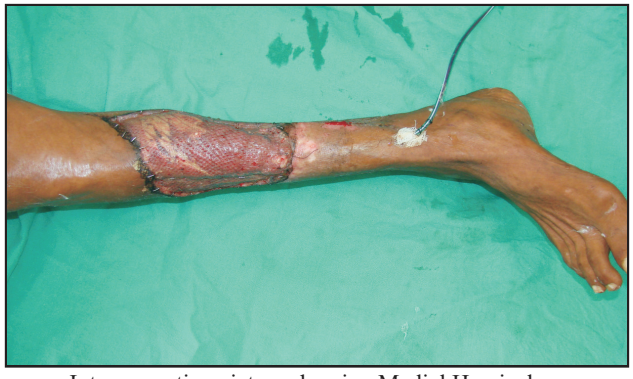
Preoperative picture showing Marjolin's ulcer right leg

Pre-operative picture showing tumor marking and 2 cm wide excision margin.



Intra-operative picture showing defect created after tumor excision.

Intra-operative picture showing defect covered with Medial Hemisoleus.



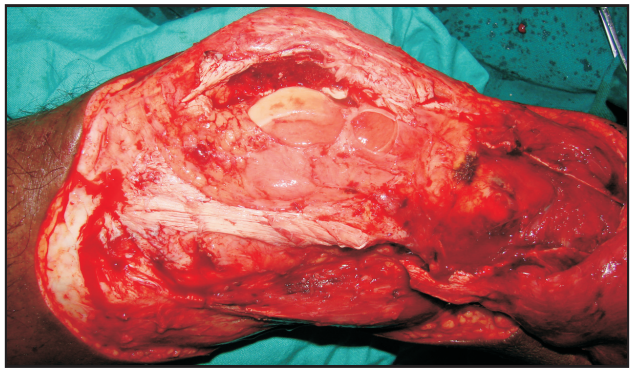
Intra-operative picture showing Medial Hemisoleus covered with SSG.

**Case 3**

48 years male presented with recurrent Marjolin's ulcer over knee for 03 years. He had history of flame burn 35 years ago which healed by secondary intention. He had already undergone tumor excision and postoperative radiotherapy 02 years back.MRI knee showed the involvement of knee joint capsule. Orthopedic opinion was to salvage knee joint by doing capsulotomy. Tumor was excised with 2cm margin, capsulotomy of knee joint was performed and defect was covered with vastus lateralis muscle flap and split thickness skin graft. Post operative radiotherapy was not given because patient had already received full dose of radiation, as advised by radiotherapist. Tumor recurred after 6 months and above knee amputation was done. (Fig10-14)

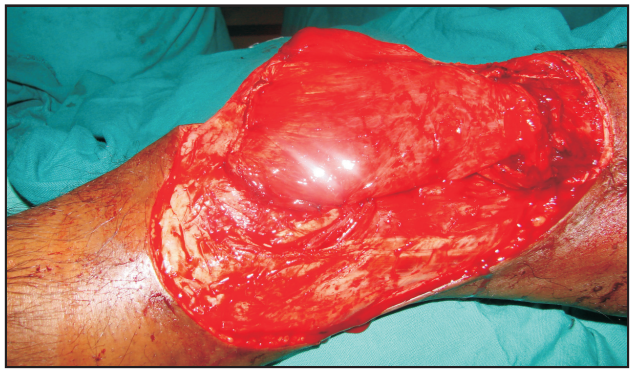
Preoperative picture showing Marjolin's ulcer left knee. Radiation was already given before referral

Pre-operative picture showing tumor marking and 2 cm wide excision margin



Intra-operative picture showing defect created after tumor excision over left knee

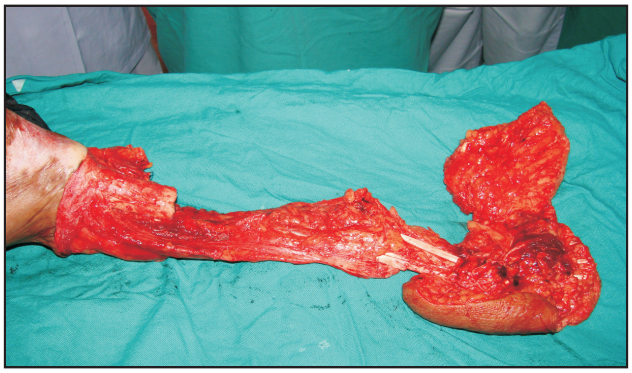
Preoperative picture showing Marjolin ulcer right leg after degloving injury twelve year back.



Intra-operative picture showing knee covered with vastas lateralis muscle flap

X ray right leg showing osteomellitus of tibia and fibula.

Intra-operative picture showing vastas lateralis covered with SSG



Intra-operative picture showing below knee amputation and preserving foot fillet flap.

## Case 4

17 years female presented with non healing wound leg for 12 years following degloving injury. Preoperative evaluation showed involvement of tibia. Below knee amputation with stump formation done using fillet flap of foot and split thickness skin graft. Post operative radiotherapy was given. Patient is disease free for past three years.(Fig 15-18)

Intra-operative picture showing stump of below knee covered with foot fillet flap and SSG.

**Case 5**

18 year female presented with non healing ulcer scalp for 06 months. She had toka machine avulsion injury of scalp 09 months back and wound was covered with split thickness graft. Histology revealed poorly differentiated SCC. Preoperative evaluation showed the involvement of outer and inner tables of skull. Excision was done by neurosurgery team with removal of outer and inner tables. Dura was involved by tumor, but dura was not excised because tumor was overlying superior sagittal sinus. Currettage of dura was performed and defect was covered with free Latissmus dorsi flap with skin paddle. Post operative radiotherapy was given. Patient died after 03 months due to invasion of brain by tumor. (Fig 19-22)

Preoperative picture showing Marjolin ulcer in SSG after degloving injury scalp nine month back.

Intra-operative picture showing defect created after tumor excision with 2 cm margin and excising all unstable scar.

Postoperative picture showing defect covered with free Latissimus Dorsi musculocutaneous flap and SSG. The cutaneous part of flap is over malignant area so that post operative radiation can be given

Lateral view showing well healed area after 6 weeks

**Discussion**

Marjolin's ulcer is aggressive malignancy with worse prognosis than any other skin malignancy. Best form of management is to prevent development of Marjolin's ulcer by promoting primary wound healing. In our study 94% of our patients of Marjolin's ulcer, initial wound healing occurred by secondary intention. Healing by secondary intention promotes unstable scarring and non healing ulcers.

There should be high index of suspicion for development of Marjolin's ulcer in patients in whom wounds healed by secondary intention. Ulcer features that are suggestive of malignant transformation include a chronic ulcer of greater than three months' duration, excessive granulation tissue beyond margins, everted wound edges, recurrent breakdown of ulcers after healing, static non healing ulcers after appropriate treatment, and ulcers that increase in size or pain despite appropriate therapy.[12] These non healing wounds should be biopsied especially when there is change in nature of ulcer. Lawrence[13] suggested that specimens be taken from both the center and the margins of suggestive lesions.

Preoperative evaluation should be done to rule out lymphatic involvement and distant metastasis. Prognosis worsens with lymphatic involvement. Wide local excision with 2cm margin should be taken. Amputation is indicated in case of bone or joint involvement, involvement of neurovascular tissue, extensive infection, hemorrhage (Erosion of vessel) or excision impairs function. Lymph node dissection is indicated when lymph nodes are suspected on palpation or radiological imaging. Role of sentinel lymph node biopsy is controversial.

Flap coverage is required when vital structures are exposed (Neurovascular bundle, bone or Joint, tendon) or patient needs post operative radiotherapy. Indications for radiotherapy are.

1. Inoperable regional lymph node metastasis.
2. Grade 3 lesions with positive lymph nodes after regional lymph node dissection.
3. Tumors with a diameter greater than 10 cm and with positive lymph nodes after regional lymph node dissection.
4. Grade 3 lesions with a tumor diameter greater than 10 cm and negative lymph nodes after regional lymph dissection.
5. Lesions of the head and neck with positive lymph nodes after regional lymph node Dissection.[14]

Long-term follow-up is recommended in all cases of Marjolin's ulcer. We had 18% recurrence in our cases. Most series indicate that the incidence of recurrence is in the range of 20% to 50%.[15]

## Conclusion

- Marjolin's ulcer can be prevented by promoting wound healing by primary intention and early coverage of wound defects with graft or flap.
- Index of suspicion should be high for patients in whom wounds healed by secondary intention.
- Chronic wounds should be biopsied to rule out Malignancy esp. when there is change in nature of ulcer.
- Marjolin's ulcer of head and neck should be treated more aggressively in term of excision and adjuvant radiotherapy.
- Limb salvage is possible with modern reconstructive options.

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