Case Report
Marjolin’s Ulcer In Young Age: A Rare Case Report
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ABSTRACT
BACKGROUND: A Marjolin’s ulcer describes malignant degeneration in any chronic wound. This condition is most commonly seen post burn scar formation as originally described by Jean Nicholas Marjolin in 19th century. The majority of cases arise in burn scars and are often latent for decades. However, it can originate from other chronic wounds of long duration.

Materials And Methods: In the present case report 13 years old girl with Marjolin’s ulcer. A review of literature was also undertaken. Results: In the reported case, the ulcer was found on the lower extremity. Squamous cell carcinoma is the most prevalent malignancy identified in Marjolin’s ulcer and was confirmed in many studies which is also confirmed in present study.

Conclusion: Based on observations and a review of the literature, early detection and aggressive surgical management either with wide local excision and prompt coverage or in case with clear evidence of bone involvement or with harbouring osteomyelitis, amputation yield optimal results when treating with Marjolin’s ulcer.

INTRODUCTION
The earliest observation of malignant changes within burn scar epithelium dates back to first century by Aurelius Cornelius Celsus. The French academic Surgeon Jean Niclas Marjolin is honoured with eponym. Marjolin’s ulcer classically refers to Squamous cell carcinoma arising in thermal burn scars, although the term has more recently been used to describe malignant degeneration in any chronic wound. Marjolin himself simply documented chronic ulcers arising fro scar tissue in the first edition of Dictionnaire de Medicine, published in 1828. The association of malignancy and thermal burn scars was first reported in literature in 1833 by the English surgeon Marjolin’s ulcer was first used by Professor Robert William Smith in 1850 and later by Dr. John Chalmer Da Costa and Dr. John Addison Fordyce, which helped establish the eponym in medical vocabulary.

Upwards of 75% of reported Marjolin’s ulcer occurs in burn scar sites. The overall rate of malignant transformation in burn scars is no more than 2%. Malignant degeneration has also been reported in chronic ulcers from trauma, forst bite, Discoid lupus erythematous, skin graft sites, pilonidal abscess, pressure ulcer, snakebites, venous stasis ulcers, leprosy ulcers, hidradenitis suppurativa and osteomyelitis.

Marjolin’s ulcer has been seen in all age groups, though the average age of patient tends to be near 50 at the time of diagnosis. There is male to female ratio of 2:1. Marjolin’s ulcer have the ability to develop in almost any anatomical location, although their incidence is highest in the lower extremities though rare, Malignant degeneration in chronic wounds continue to occur sporadically.

CASE REPORT
A 13 years old female girl came to hospital with chronic ulcer on right leg since 3 months. Patient reported having suffered a burn at age of 5 months followed by healing with contracture of Ankled joint. Since then patient was asymptomatic then she developed a pea sized ulcer over right ankle joint over past burn contracture which was insidious in onset and progressed to present size of 7x4x2 cms with irregular margins, slough over floor with everted edges and serosanguineous discharge of foul smell. Exceeding fixity of Ankle other joint movements are normal. Except anterior tibial, posterior tibial and dorsalispedis other lower limb pulses are felt. No inguinal lymphadenopathy is present. The remainder of physical examination was unremarkable. Histopathology shows pseudo epithelmatous hyperplasia with hyperkeratosis of one corner of section, epithelial cells show loss of pricking. Basement membrane is destroyed by dense MRI of right ankle shows-Malignant mass lesion in soft tissue of foot with underlying bony erosion. Doppler study shows-visualised vessels show normal arterial and venous study except for luminal irregularity of arteries. Remaining other investigations are normal. Patient posted for below knee amputation in view of biny erosion. Histopathology showed invasive squamous cell carcinoma invading periostium of underlying bone, patient displays no evidence of local, regional or distant metastasis.

DISCUSSION
Marjolin’s ulcers are malignant transformation of chronic wounds. Malignant transformation, as demonstrated in present case, has been recognised since the 19th century. In 1828 Jean Nicolas Marjolin first described an indolent ulcer arising in a burn scar. Urgently the term Marjolins Ulcer most accurately describes malignant degeneration of a chronic inflammatory skin lesion, regardless of origin of lesion or the type of cancer that develops in the lesion seventy one percent of marjolins ulcers develop as a squamous cell carcinoma, Melanoma, fibrosarcoma, angiosarcoma, liposarcoma, leiomysarcoma, osteosarcoma, dermatofibrosarcoma protuberance, malignant fibrous histiocytoma, malignant schwannoma and mesenchymal tumour have also been identified. According to Bozkurt et al the wound may also appear as pseudoepitheliomatous hyperplasia. When Marjolin’s ulcer occurs in its squamous cell form it is a very aggressive malignancy but constitutes only 2% of all squamous cell carcinomas. Marjolin ulcer typically take years to develop though, on occasion, acute onset has been documented. Latency has been described as the time between initial injury and confirmation of a pathological diagnosis of Marjolin’s ulcer. There is variation in latency period, but many studies report at least 2-3 decades between injury and malignant transformation. Marjolin ulcer can be classified as acute or chronic, with malignant transformation that takes place within 12 months of a burn considered acute. Although over whelming majority are squamous cell carcinomas, basal cell carcinomas is relatively more common in acute transformation and in scars resulting for a superficial burn injury. It is thought that there is an inverse relationship between latency period and patient age at the time of burn injury, with older patients having a shorter latency period.

The report by Baskara et al which describes the development of a
Marjlon’s ulcer in a 72-year old within 9 months of a plantar pressure ulcer provides some validation for this premise. The exact mechanism for development of Marjolin’s ulcer in burns or other wound types has not yet been identified, but is likely multifactorial, affected by both environmental and genetic factors. Key factors in development of Marjolin’s ulcer seem to be slow healing process and chronic instability of scar tissue. Marjolin’s ulcer tend to favour locations where there is a constant trauma or a compromised blood supply. It has been suggested that in the chronic wound, decreased vascularise combined with weekend epithelium creates a susceptibility to carcinogens. It has also been proposed that relative avascularity of scar tissue leads to a locally depressed immunological state or immunologically privileged site leaving the body without an adequate cell mediated response. Release of toxins by lysis of scar tissue may have a direct mutagenic effect on cells. Mutation in p53 gene and Fas gene may disrupt regulated apoptosis and cellhomeostasis, respectively, and have been identified in patients with Marjolin’s ulcer. Chronic irritation and repeated attempts at healing provides a prolonged stimulus for cellular proliferation and may increase the spontaneous mutations. The evidence supporting this theory is represented in many cases of Marjolin’s ulcer occurring at skin zones that have been exposed to long term irritation, including areas where clothing might cause trauma. These findings support the idea that chronic irritation is an inciting factor. Marjolin’s ulcer can be suspected based on a nonhealing ulcer in an area of abnormal or scarred skin. However, the suspicion must be confirmed with pathological interpretation of tissue biopsy specimen taken from multiple locations of ulcer and its margins to minimize the false negative findings. A standardised biopsy procedure involving excision biopsy that is limited by the size of neoplasm has been proposed. This biopsy procedure may increase the percentage of correctly diagnosing the tumour but a more focused investigation may be needed to determine the minimum number of biopsies required. Also, the use of MRI has been found to be helpful in determining the degree of soft tissue invasion of Marjolin’s ulcer on the extremities and extent of bone destruction and periosteal reaction. Tumour type, location, rate of metastasis affect prognosis. Location of the tumour is a key prognostic factor influencing metastasis, and the rates of metastasis in descending order of ulcer locations most likely to metastasize are lower extremities, trunk, scalp, face, neck and upper extremities. Marjolin’s ulcer was approximately 27.5%. The rate of metastasis in Marjolin’s ulcer arising in pressure ulcer has been listed to be as high as 61%, a rate higher than that resulting from Burn scars (38%) and osteomyelitis (14%). The survival of patient with Marjolin’s is 65%-75% at 3 years post diagnosis, but falls to 35%-50% if metastatic disease is detected on presentation. Palpable regional lymphadenopathy predicts death within 2 years. 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, lymphnodes dissection, or the use of Neoadjuvant radiotherapy or chemotherapy. A combination of these procedures is often necessary. Based on their review, Bozukart et al found a very low recurrence rate when 3cm-5cm margins were obtained. 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, it is documented that adjuvant radiotherapy and chemotherapy can be useful as well. Lymphadenectomy is warranted if there if persistent adenopathy.

Ilaih et al.,

**Fig: 1**

**Fig: 2**

**Fig: 3**
REFERENCES