Acute Marjolin’s Ulcer in a Venous Ulcer - Case Report

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Abstract: Marjolin’s ulcer is the eponym used to describe carcinomatous change occurring at the edge of any longstanding benign ulcer irrespective of cause\textsuperscript{1}. This should be suspected when part of an ulcer shows evidence of proliferation. This type of tumor is usually a squamous cell carcinoma\textsuperscript{2}. A chronic ulcer unresponsive to dressings and simple treatments should be biopsied to rule out neoplastic change\textsuperscript{3}. These cases typically appear decades after the original injury in wounds that healed primarily, but acute cases arising within a year of injury have been reported\textsuperscript{4}. Malignancy mandates wide excision, with potential amputation if lesion is on an extremity\textsuperscript{4}. A middle aged man presented with a non healing ulcer near the ankle for over one year. On examination there was an ulcer with pale granulation tissue, with varicose veins. Biopsy of the ulcer edge revealed well differentiated squamous cell carcinoma. X-ray of the affected part showed bone involvement by the tumor. A below knee amputation was done with the posterior flap well away from the tumor. This was a rare case where marjolin’s ulcer developed in a short span of one year.

Keywords: Non healing ulcer, varicose veins, squamous cell carcinoma, marjolin’s ulcer, amputation

1. Case Presentation

History of Presenting Complaint: A 65 year old man presented with a non healing ulcer on his left leg near the ankle for the past one year. He sustained a trivial trauma to the left ankle one year ago. The ulcer was slowly increasing in size. It was associated with serous discharge. He also complained of dull aching pain in the calf which was more towards the end of the day and was relieved on lying down and keeping his legs raised. No other symptoms. He used to go to the local hospital where wound dressings and medications were prescribed but in spite of regular dressings the wound was not healing hence referred to K. R. Hospital.

Past history/ Family history – Nothing contributory

Personal history – chronic smoker (30 pack years)

2. General Examination:

Elderly man, moderately built and nourished, alert. No pallor/icterus/cyanosis/generalized edema or lymphadenopathy. Vitals stable.

3. Local Examination

A solitary ulcer in the lower lateral aspect of the left leg extending on to the foot. Size 10x5 cm , irregular margins, raised edge [Fig 1:Ulcer showing raised edge (arrow) ]. Pale granulation tissue and slough on floor. Serous discharge from the ulcer. Base formed by the bone in one area and the muscle and overlying fascia in the rest. Edge and the surrounding area indurated. Limb proximal to the ulcer showed varicose veins involving the long and short saphenous system [Fig 2: Lipodermatosclerosis (white arrow) varicose vein (yellow arrow) ]. No evidence of Deep Vein Thrombosis. Peripheral arterial pulsations normal. No regional lymphadenopathy. Ankle movements restricted. Sensations preserved. Contralateral leg normal.

Figure 1: Ulcer showing raised edge (arrow)
4. Systemic Examination

CVS/CNS/ABDOMEN: Normal RS : occasional rhonchi

Clinical Diagnosis: Callous ulcer in the left leg probably due to varicose veins involving the long and short saphenous system.

Investigations:
Routine investigations – Normal

Edge Biopsy – Well differentiated squamous cell carcinoma [Fig 3: well differentiated SCC with Keratin pearls (arrows)].

Venous Duplex – SFJ incompetence, SPJ incompetence, Multiple perforator incompetence.

X-Ray – Osteolytic lesions in the distal fibula with loss of cortico-medullary differentiation [Fig 4: Involvement of fibula (blue arrow) and calcification in the base of ulcer (yellow arrow)].
Final Diagnosis: Marjolin’s ulcer in the distal part of the left leg secondary to varicose veins of the long and short saphenous system with infiltration of distal fibula.

Plan: In view of the involvement of the bone by the marjolin’s ulcer, it was decided to perform a below-knee amputation.

Perioperative period: Under spinal anaesthesia a below-knee amputation was done with a long posterior flap [Fig 5: Below knee amputation flap]. Care was taken to ensure the flap margins were well away from the ulcer [Fig 6: Well differentiated SCC with Keratin pearls (arrows)]. Prophylactic lymph node dissection was not done. A posterior slab was applied to prevent flexion contracture. Postoperative period was uneventful. After suture removal, patient underwent regular physiotherapy and a prosthetic leg was designed for him. Histopathology report confirmed well differentiated squamous cell carcinoma with margins being free [Fig 3: well differentiated SCC with Keratin pearls (arrows)]. Patient has been advised for regular follow up to detect recurrences.

Figure 4: Involvement of fibula (blue arrow) and calcification in the base of ulcer (yellow arrow)

Figure 5: Below knee amputation stump
5. Discussion

Jean Nicholas Marjolin (1780-1850) a French surgeon described the development of carcinomatous ulcers in scars in 1828. Squamous cell carcinomas (SCC) that arise in burn scar or a chronic, open wound overlying osteomyelitis is often referred to as a Marjolin’s ulcer. The various etiologic mechanisms for these tumors include release of local toxins following injury, induction of dormant neoplastic cells and induction of injury-induced preneoplastic cells by cocarcinogen. Marjolin’s ulcer is usually seen in low socio-economic societies with limited access to medical facilities. Most of the cases are from developing countries with late presentations. Malignant change seems to be prevented if early wound closure is undertaken. Anatomically there is a preponderance of lesions on the extremities. The Incidence of malignant tumors in chronic ulcer is 2%, and the incidence of squamous cell carcinoma (SCC) as the cause of the malignant tumor is 0.4% which is greater than the general population.

Malignant transformation occurs usually after a long period of latency of chronic infection; it takes approximately 35 years on average. But acute cases arising within a year of injury have been reported. Acute and Chronic types have been distinguished by the length of latency. Other than a discrepancy in lag time it appears unclear if there are any differences in the clinical, histological or prognosis between the acute and the chronic types. Malignancy can be diagnosed by a edge biopsy. Squamous cell carcinoma is most common, although Basal cell carcinoma can occur and rare tumors such as Malignant Fibrous Histiocytoma, Sarcoma and Melanoma have been reported. The tumors often are well differentiated and may arise in a background of pseudopitheliomatous hyperplasia, making diagnosis difficult. Well differentiated SCC exhibits polygonal squamous cells arranged in orderly lobules and exhibiting numerous large zones of keratinization ( Keratin Pearls). SCC arising in areas of chronic inflammation have a 10-30% rate of metastasis, whereas those arising not due to pre-existing inflammation or degeneration condition varies from 0.05-16%. A Marjolin's ulcer should be considered when a soft-tissue mass with a broad based skin ulcer shows a mass effect and invasion of the adjacent bone. Classically a 2cm margin is reserved for primary SCC Marjolin ulcer and 2.5 cm for recurrent cases. Malignancy mandates wide excision, with potential amputation if lesion is on an extremity. Prophylactic regional lymph node dissection has not improved survival, but Sentinel Lymph Node Biopsy is a promising modality to direct therapeutic node dissections and awaits validation in this population. On a selected basis, adjuvant radiation may be warranted. Recurrence rates are high despite current treatment advances and long term follow up are warranted. Recurrences are almost always local but distal metastasis to lung, brain, liver have been reported.

6. Conclusion

Marjolin’s ulcer is the eponym used to describe carcinomatous change occurring at the edge of any longstanding benign ulcer irrespective of cause. A chronic ulcer unresponsive to dressings and simple treatments should be biopsied to rule out neoplastic change. This type of tumor is usually a squamous cell carcinoma. These cases typically appear decades after the original injury in wounds but acute cases arising within a year of injury can occur. Malignancy mandates wide excision, with potential amputation if lesion is on an extremity. Prophylactic lymph node dissection is not necessary. In this case marjolin’s ulcer appeared within one year and in literature there are only very few cases reported, hence this is a rare case.

References


