

## Extensive Marjolin's Ulcer of Scalp in a Patient with Cardiac Dysfunction - A Reconstructive Challenge

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### 1. Abstract

Marjolin's ulcer originate in long standing non healing ulcers of various origin. It used to be the ulcerative variety of squamous cell carcinoma, encountered on rare occasions. These lesions are usually treated by wide local excision followed by reconstruction preferably using vascularised tissue of similar nature. We came across a young male patient with a large Marjolin's ulcer over temporo-parieto-occipital scalp extending intracranially with multiple cardiac valve dysfunction. The ulcer was a malignant transformation of a recurrent ulcerating scar resulting from thermal burn sustained during early childhood. We experienced a turbulent episode during excision of the pathology. The reconstruction was delayed for 48 hours and achieved using free Latissimus dorsi muscle flap covered with split thickness skin graft. This report emphasises the importance of early diagnosis of this clinical entity, reconstructive challenges for such extensive defects after its excision, anticipating various possible fatal complications and preparedness of the treating team to overcome them.

### 2. Introduction

Marjolin's ulcer is a rare and aggressive cutaneous malignancy that arises on previously traumatized and chronically inflamed skin, especially after burns. This clinical condition was first described by Marjolin in 1828 [1]. The term "Marjolin's ulcer" generally refers to squamous cell carcinoma arising from long standing scars or chronic non healing ulcers mostly arising from burn. However, wounds or scars other than burn, like scar of vaccination, snake bites, osteomyelitis, pilonidal abscesses, pressure sores, and vel-

nous stasis may also induce this tumour [2, 3, 4]. The treatment of these lesions usually include wide local excision followed by reconstruction using different autologous tissues depending upon site, extent and patient related factors.

### 3. Case Report

A 20-year-old male presented to the department of Burns and Plastic Surgery with recurrent ulceration of scalp of 3-years duration associated with recurrent haemorrhage and infestation with maggots. Patient sustained thermal burn during early childhood (at the age of 1 year) affecting major portion of his scalp and both the ears. The wound at that time was allowed to heal by secondary intention. A part of the wound did not heal completely and recurred intermittently till 6 years of age when it was debrided and covered with partial thickness skin graft. After an asymptomatic period of about 10 years, his scalp developed recurrent ulceration with occasional haemorrhagic events. Over the last 3 years he had an ulcer over his temporo-parietal scalp that did not respond to conventional non-surgical modalities of treatment and kept on increasing in size.

Examination revealed an ulcer measuring 15 X 10 cm over right temporo-parieto-occipital region of scalp with alopecic hypopigmented scar over the surrounding area. It was friable, had a bony base and irregular everted margins. There was no evidence of focal neurological deficit and cervical lymph nodes were not enlarged clinically. Systemic examination revealed signs of multiple cardiac valve dysfunction. Echocardiography demonstrated severe aortic regurgitation, moderate mitral regurgitation, mild Tricuspid

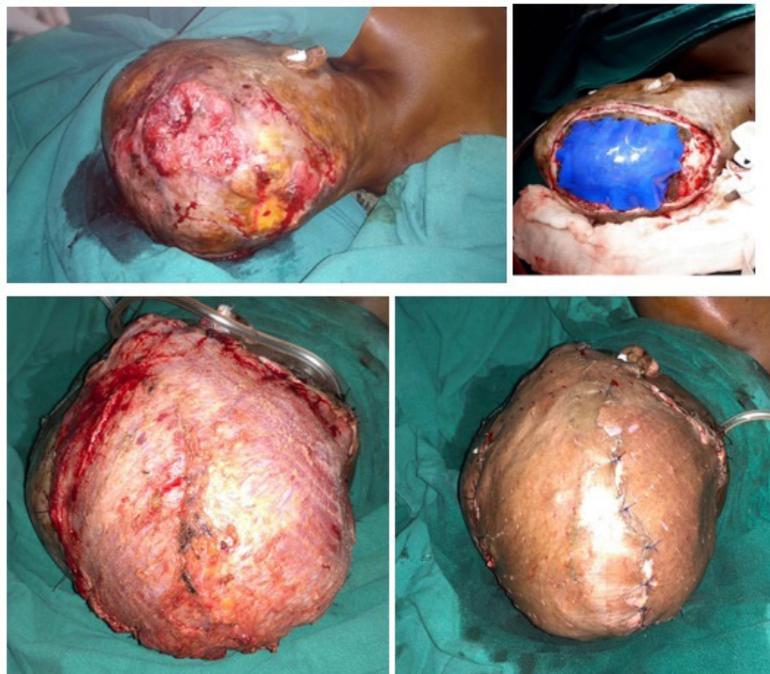
regurgitation and moderate pulmonary arterial hypertension. Incisional biopsy of the ulcer confirmed the lesion to be a Squamous cell carcinoma. MRI of the head showed huge irregular ulcerative lesion in the temporo-parieto-occipital region of scalp with intracranial extension, involving the dura mater over the posterior end of superior sagittal sinus and focal involvement of brain parenchyma of right parietal lobe.

The case was discussed in the multidisciplinary clinic for treatment of malignant tumours of our institution and decided to proceed for wide local excision of the tumour with an attempt to remove the part of the dura and brain parenchyma infiltrated by tumour. Post-excisional defect was calculated to be 18x13 cm with a bony defect of similar dimension. The dural defect was expected to be of 10x6 cm. The resultant defect was planned to be reconstructed using facia lata for dura, titanium mesh for bone and Latissimus dorsi muscle with split thickness skin graft for the scalp. The anaesthesia team was advised to prepare for management of anticipated intra-operative blood loss and any eventuality anticipated in view of severe cardiac abnormality.

While excising the lesion under general anaesthesia, the superior sagittal sinus was exposed accidentally due to adhesion of lesion to the dura overlying it. This led to torrential haemorrhage and probably air embolism. Eventually he developed hemodynamic instability. The patient could be resuscitated by aggressive interventions including necessary blood replacement and administration of inotropic medications. In the wake of such a turbulent intra-operative episode during excision, the reconstruction was deferred to a later stage since it was expected to impart another major surgical stress considering the need for a microvascular free tissue transfer. The

exposed brain was covered by Poly Tetra Fluoro Ethylene (PTFE) dural patch, reinforced by moist collagen sheet dressing. After being the patient stable haemodynamically over next 48 hours, he was taken back to operation room for definitive reconstruction of the scalp defect. In order to prevent compression of oedematous brain in that situation, it was decided by the treating team not to reconstruct the skeletal element of the defect and left for future sitting. Soft tissue reconstruction was performed by using free Latissimus dorsi muscle flap harvested from right side with sheets of Split Thickness Skin Graft (STSG) covering it. The thoracodorsal artery and vein were anastomosed to the right superficial temporal artery and vein at the level of zygomatic arch. Lumbar sub arachnoid space drain was placed to maintain the Cerebro-Spinal Fluid (CSF) pressure at a lower level in an attempt to prevent CSF leak from the flap margins and aid in uninterrupted healing of the wound. The patient recovered uneventfully and the surgical site healed completely by the end of 4th week. The histopathology report revealed the tumour to be a well differentiated squamous cell carcinoma, keratinizing type with all the margins free of tumour except the deeper one. He received adjuvant radio-therapy (60 Gray spanned over 6 weeks) after complete healing of the scalp without any obvious adverse effect either local or systemic.

After 2.5 years of completion of treatment, the patient did not show any evidence of local recurrence, awaiting his Calvarial (skeletal) reconstruction if considered necessary. Of course, any further reconstructive surgery for him will be planned after his corrective cardiac surgery. He has been advised for a regular follow up in view of residual tumour tissue in the brain parenchyma which could not be resected completely (Figure 1 and 2).



**Figure 1:** 1. A – The scalp lesion, 1. B – PTFE dural patch covering brain after excision  
1. C – Latissimus dorsi muscle flap over dural patch, 1. D – STSG covering the muscle flap



**Figure 2:** 2. A, 2. B, and 2. C – Follow up pictures after 2.5 years

#### 4. Discussion

Marjolin's ulcer developing in a post burn scar over the scalp although rare, not very uncommon. Owing to its local aggressiveness it is possible to erode the underlying skull and invade into the adjacent meninges or brain parenchyma. This cutaneous malignancy most commonly arises from a long standing ulcer or scar due to burns. Whereas vaccination, snake bites, osteomyelitis, pilonidal abscesses, pressure sores, and venous stasis may also induce this tumour [2-6]. Metastasis in this disease has also been documented and once it does, prognosis worsens [7, 9, 11]. The most common age of affection of this malignancy is 53 to 59 years and the lag period ranges from 4 weeks to 70 years with a mean of 36 years [7,8,9]. In case of our patient the lag period was 16 years and post ulceration period of 36 months. In this case the lag period is short and age of presentation is much less despite the post ulceration period being comparable to that of Xiao H et al 9. The disease may manifest in individuals even in their teens as reported by Daya & Balakrishnan [10]. Ignorance about the nature of such tumour delays the treatment, thereby affecting the prognosis adversely.

In this particular case intracranial extension of tumour and Multiple cardiac valve dysfunction made the clinical decision making and management more complex. The tumour eroded through the underlying skull involving dura over a part of superior sagittal sinus and surrounding brain parenchyma. The aortic, mitral and the

tricuspid valve functions were compromised significantly with the aortic valve being the worst. In such a situation, the task of multi-disciplinary team was horrendous to decide whether to address the malignancy first or the cardiac disease. Since the boy was capable of doing all his normal activities other than strenuous exercises, the team decided in favour of managing malignancy first.

Reconstruction of such a big defect after excision of the lesion with replacement of all the tissue components was a challenge. Although titanium mesh was decided to be used to replace the skeletal component and free Latissimus dorsi muscle with split thickness skin graft for soft tissue before contemplating the surgery, per operative situation did not permit the same. During the tumour excision, rupture of the wall of superior sagittal sinus led the patient to sink into severe haemodynamic instability. After an erstwhile struggle to resuscitate the patient, further extensive reconstruction with microvascular tissue transfer was deferred till 48 hours with the temporary covers for exposed brain tissue. This clinical judgement carries points both in favour and against a favourable outcome of treatment as a whole. Proceeding immediately for an extensive and time consuming reconstructive surgery could have been detrimental in terms of surgical stress, in a situation when massive haemorrhage and air embolism has just been taken care of. On the other hand, delay in reconstruction was posing the exposed brain parenchyma to threat of infection and cerebral oedema. At the end

our decision was found to be appropriate, as we could complete the reconstruction as per our pre-treatment plan without any eventuality. Skeletal reconstruction was not done considering the cerebral oedema by then and risk of prosthesis infection in such a scenario. Here we emphasise that free tissue transfer is the most appropriate mode of soft tissue reconstruction in such large defects which are far away from all available locoregional pedicle flaps. Of course the defects of lesser dimensions can be resurfaced with various local or locoregional pedicle flaps as reported by Mishra et al 12. In present context, non-microvascular techniques of distant tissue transfer should not be adopted, unless the microvascular surgery is not feasible in terms of availability of expertise or infrastructure.

Complete excision of tumour in such situations may not be feasible on every occasion and adjuvant radiotherapy may come handy as a saviour. This aspect occupied most of the part of discussion when the treatment was planned by multidisciplinary team. The residual tumour in brain parenchyma was subjected to adjuvant radiotherapy as soon as the surgical site healed. At the end of 2.5 years, the patient did not develop any clinical manifestation of intra cerebral space occupying lesion nor of local recurrence while the correction of cardiac dysfunction is still awaited.

## 5. Conclusion

The challenges involved in managing extensive Marjolin's ulcer of scalp invading into intracranial tissues are multitude. Multidisciplinary treatment planning with members from surgical oncology, neurosurgery, plastic surgery, anaesthesiology, medical oncology and radiotherapy is essential to achieve the treatment goal. The members of the team should be amenable to mould themselves and take judicial decisions appropriate to the situation. After all it has mostly been seen that "fortune favours the brave".

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