

CASE REPORT

Penetrating Marjolin's ulcer of scalp with intracranial extension: A multidisciplinary experience

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ABSTRACT

Marjolin's ulcer invading the skull, dura mater, and the brain is quite exceptional. We report such a rare case of massive Marjolin's ulcer arising in the chronic scar tissue of the scalp of an elderly male. The ulcer had invaded the skull, dura mater, and underlying brain tissue. The patient was managed successfully by wide excision and scalp reconstruction. The importance of appropriate and timely wound management of scar ulcers with high malignant potential is highlighted. The role of multidisciplinary approach in the scalp reconstruction is discussed.

Key words: Brain tissue, dura mater, Marjolin's ulcer, scalp reconstruction, skull

Introduction

Marjolin's ulcer is rare, often aggressive and well-recognized long-term malignant complication developing in non-healing scar tissue.^[1] Marjolin's ulcer of the scalp, usually epidermoid carcinoma in nature has been variously reported in literature.^[2-4] In this part of the globe, due to poverty and ignorance, many a time this entity has been neglected by the patients leading to delayed diagnosis. Owing to the behavior of the lesion and late presentation, the treatment results are often disappointing. Though it is known as a plastic surgical problem, when it invades the dura and brain, it becomes neurosurgical interest.

Case Report

A 58-year-old male developed an ulcer over his chronic scalp scar, which he developed due to bear mauling about 50 years back. The lesion developed following a minor trauma to the scar tissue and went through a cycle of apparent healing and deterioration. The ulcer continued to increase in size, and a cauliflower-like granulomatous mass appeared in the center

with repeated episodes of foul smelling serosanguinous discharge and contact bleeding.

On examination, he was conscious, with stable vital signs. A fungating growth of size 12×10×5 cm with irregular and indurated margin was found over the scar on his occipital area. The surface was covered with pale granulation tissue with intervening hemorrhage and necrosis [Figure 1]. Scrap cytology revealed squamous cell carcinoma. Fine-needle aspiration cytology of the enlarged right occipital and posterior triangle lymph nodes showed features of inflammation and reactive hyperplasia. Computed tomography scan showed a right occipital extra-calvarial isodense lesion with underlying bone erosion and meningeal thickening [Figure 2].

Definitive surgery was planned based on prior discussion with the plastic surgeons. The patency of scalp vasculature was confirmed and marked over skin by Doppler ultrasound to plan rotational flap. With the patient in prone position, wide excision of the lesion was carried out with a 2 cm margin of normal skin. The underlying bone was found to be eroded with the dura infiltrated and adherent to the brain tissue below it. The unhealthy bone margins were nibbled away. The involved dura and an area of abnormal cortical brain tissue were excised and the dural defect was repaired using a G-dura patch [Figure 3]. The scalp defect was closed with local rotation flap based on left occipital artery in the same sitting. The secondary defect was grafted with split thickness skin graft. Histopathology of the mass revealed squamous cell carcinoma. One hundred percentages flap survival was achieved post-operatively with uneventful wound healing and acceptable cosmetic outcome [Figure 4]. On the 8th day, he was discharged with advice to attend radiotherapy department for possible radiotherapy.

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Figure 1: Pre-operative (a) and per-operative. (b) Photograph showing the ulcerative growth on the occiput of the scalp

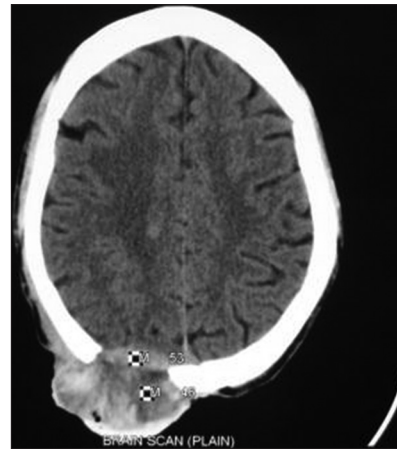


Figure 2: Computed tomography scan showing a right occipital extra calvarial isodense lesion with underlying bone erosion and meningeal thickening

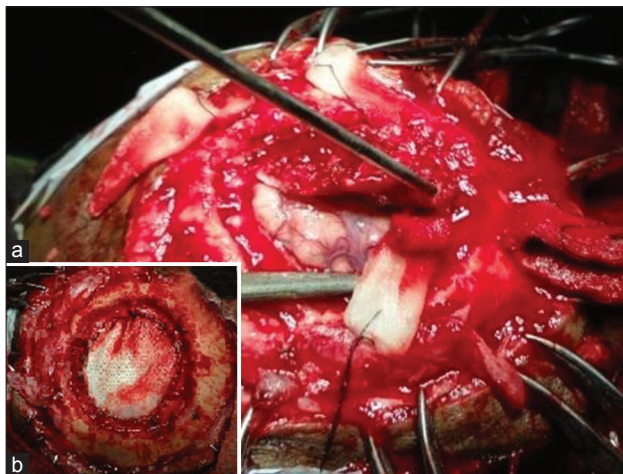


Figure 3: Intra-operative photograph showing (a) the eroded skull bone with exposed underlying brain and (b) the repaired dural defect using G-dura patch



Figure 4: Post-operative photograph at the time of discharge with 100% graft survival and acceptable cosmetic outcome

Discussion

Although it was described originally in a burn scar, Marjolin's ulcer can arise in any chronic wound of diverse origin.^[5] In our case, it is due to bear mauling, an unusual mode of injury. Rich vascularity of scalp is responsible for its lower incidence in scalp.^[3,4] Invasion of the skull, dura mater, and the brain is quite exceptional and has only been reported in a limited number of cases.^[3,4,6-8]

The reason for malignant transformation is largely unknown. Virchow suggested that any cutaneous scar subjected to continuous irritation has increased potential for malignant transformation.^[5,9] Some factors such as decreased vascularity, compromised lymphatic system, and decreased regenerative capacity of scars have also been proposed.^[9] The cicatricial tissue which prevents the action of the immune system favors new tumor formation. We suppose that trauma associated with repeated ulceration and healing in the scar tissue is the

most important factor in the pathogenesis of the Marjolin's ulcer as in our case.

In the scalp tissue, the transition time is quite variable. The average latency in chronic form of Marjolin's ulcer as has been described in literature is 36 years,^[3,7] and in our case, it was longer. Another striking feature of our case was the rapid tumor progression not only in the scalp but also in the underlying skull bone and brain parenchyma.

The classical presentation is a fungating growth with everted margin, often with foul smelling discharge and contact bleeding. When the tumor is confined to scar tissue, the growth is slow but once it spreads out of the scar, it metastasizes rapidly. The diagnosis is confirmed by biopsy. Marjolin's ulcer can be very aggressive tumor that necessitates a well-thought-out treatment plan to optimize care and assure patient survival.^[10] Early diagnosis and prompt surgical intervention are mandatory in Marjolin's

ulcers of the scalp as they may invade vital structures. The main stay of treatment is wide local excision with 2 cm tumor free margin.^[5,9] It is imperative that the surrounding scars to be excised. Reconstructive techniques are useful.^[2,7,10] Small defects (<3 cm) closed primarily, split skin graft is sufficient for larger defects with intact pericranium. For full thickness scalp defects, local rotation flaps and in case of very large defects, free tissue transfer is used.^[4] Primary radiotherapy has not been effective because of the poor vascular supply of the tumor and its role is therefore palliative.^[5,9] Adjuvant radiotherapy should be given for incomplete removal and positive nodal status. Regional lymph node dissection is controversial.^[5,9] The optimum treatment for advanced Marjolin's ulcer remains controversial.

The outcome for scalp Marjolin's ulcer is often dismal with early recurrence and death.^[3,6] Prognosis is generally poor in patients with Marjolin's ulcer of the scalp invading the brain. Early recognition, when it is confined to the scar, offers best chance for cure and wide local excision alone at this stage may be curative.^[11]

Conclusion

A delay in diagnosis as a consequence of poverty and ignorance remains a major stumbling block in improving the outcome of aggressive Marjolin's ulcer of scalp. Health education is needed to discourage its late presentation. Appropriate and initial wound management of scar ulcers in time will facilitate wound healing and minimize the risks of malignant transformation. A team approach including plastic surgeon is needed for the optimum reconstruction of scalp following wide excision of the ulcer.

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