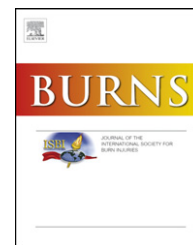


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## Case report

# Penetrating scalp Marjolin's ulcer involving bone and dura mater in a Nigerian hospital: Case report and literature review

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### ARTICLE INFO

Article history:

Accepted 9 April 2009

## 1. Introduction

Marjolin's ulcers are epidermoid carcinomas developing in non-healing scar tissue [1]. Marjolin's ulcer of the scalp is an occasional form of cutaneous cancer seen in our environment. Owing to the behaviour of the lesion and late presentation the treatment results are often disappointing. They are commonly found in the lower limbs, occasionally around the head and upper limbs, and rarely on the trunk. Marjolin's ulcers of the scalp have been variously reported [2–4], but not in the paediatric age group in this environment [5,6]. This report hopes to contribute to that.

## 2. Report

A 12-year-old female was presented to us in 2004 with a fungating scalp lesion having sustained full-thickness flame burns to the scalp when she was three months old from a kerosene lantern, which ignited the bed she lay on. The wound never completely healed, but went through cycles of apparent healing and deterioration, as she was taken through several health facilities—traditional and orthodox. Fifteen months from presentation the ulcer began widening rapidly, associated with foul smelling discharge, contact bleeding and nocturnal pains. There was no history of personality changes

or seizures, or clinical evidence of cranial nerve deficit or a space-occupying lesion.

Examination revealed a well-nourished girl with a roughly triangular depigmented left parietal scalp scar (13 cm × 10 cm) with an ulcer at the lateral margin overlying the temporal region and measuring 10 cm × 8 cm × 2 cm (Fig. 1). The edges were everted, and unhealthy granulation tissue exuding foul smelling yellow–brown discharge was on the floor. The base was fixed to bone. There was no regional lymphadenitis. There was no demonstrable clinical evidence of intra cranial lesions, or distant spread. She had had normal milestones, and secondary breast development had begun. X-ray findings were that of a soft tissue mass with underlying sclerosis of the bone, and patchy lytic areas, indicative of cranial involvement (Fig. 2). Computerised tomography studies were requested but not done for financial reasons. Wide excision and ostectomy were planned.

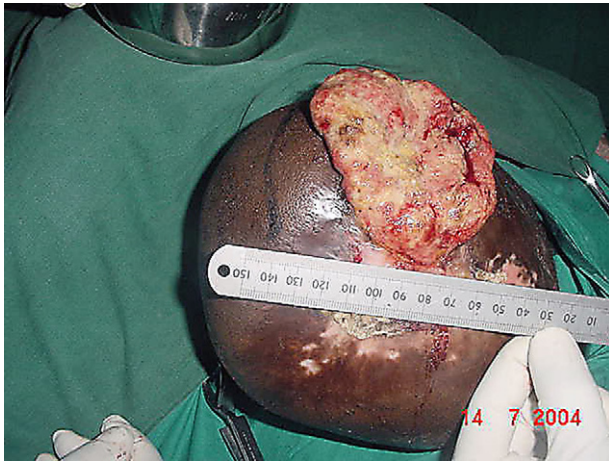
With the patient in the prone position wide excision was carried out involving the dura mater exposing the brain. The scalp excision involved the entire unstable scar and a contiguous ulcer excision with a 3 cm margin. Frozen section was not done: it was unavailable in the sub region. The cranial defect was 11 cm × 12 cm. A modified orticochea flap reconstruction using the rest of the scalp was carried out. A large occipito-parietal flap (19 cm × 14 cm) based on the left occipital and retro-auricular vessels was mobilised down to

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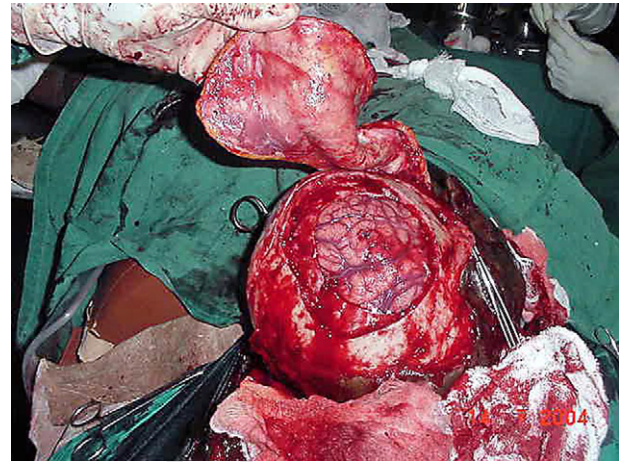
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0305-4179/\$36.00 © 2009 Published by Elsevier Ltd and ISBI.

doi:10.1016/j.burns.2009.04.010



**Fig. 1 – Mass on scalp.**  
Note the fleshy growth arising in depigmented skin, and the bleeding from the lesion. Note also the unstable scalp scars.



**Fig. 3 – Excised lesion with raised flap.**  
Note the exposed brain and the posteriorly based flap raised to cover the defect.

the nape of the neck and transposed to completely cover the calvarial defect (Fig. 3). A frontal flap measuring 9 cm × 7 cm based on the right superficial temporal vessels covered the rest of the primary defect. This was assisted by wide undermining of the temporal and forehead skin. The flap included a small part of the forehead. The secondary defect in the occipital region was grafted primarily with fenestrated partial thickness skin grafts taken from the thigh. Dog-ears occurred at the frontal and left post auricular regions, and a drain exited via a separate stab incision in the same region and close follow up to detect recurrence was mounted post operatively. Two units of blood were transfused intraoperatively. Galeal scoring was omitted. Post operatively there was 98% graft take and 100% flap survival.

The histology report read “Clinical data provided: Marjolin’s ulcer. Scalp mass 15 months. Old burn scar of scalp 11years. Fungating mass left parietal region 8 cm × 6 cm central area covered with slough. Everted edges. Fixed to skull. Surrounding hypopigmented and hyperpigmented skin. Excision biopsy. Calvarial and dural involvement. Report: in

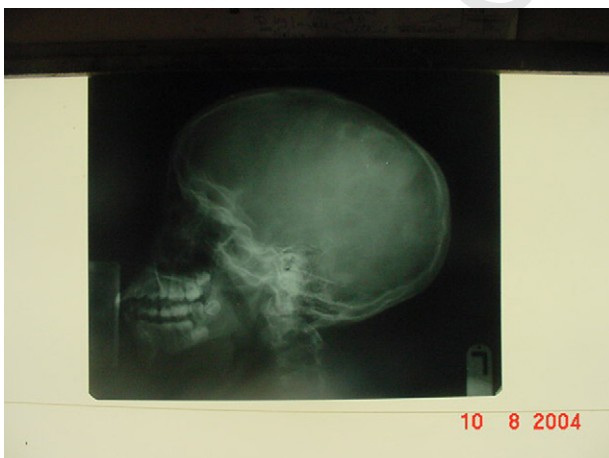
the same container are (A) 20 cm distorted skin with growths, (B) 11 cm bone with irregular growths destroying it centrally, and (C) 4 cm curled up fibrous mass with soft growths. Microscopy—all sections are the seat of squamous cell carcinoma. Incidentally, sections (B) and (C) are highly keratinizing squamous cell carcinoma”.

The patient was not compliant with post operative appointments. Three months later a local recurrence was noted at the edge of the flap. Owing to a combination of the patient defaulting and unable to procure medicament, industrial action in the hospital, and absence of part of the surgical team proper re-excision was not done until six months after first presentation. Excision again exposed the brain which was skin grafted. Combination chemotherapy with cyclophosphamide, adriamycin and cisplatin was ordered, and the patient referred for possible radiotherapy. She developed cervical node metastasis while waiting for it. She was last seen four months after the last excision still with metastases.

### 3. Discussion

The association between thermal burn scars and neoplasia was recognized by Celsius in AD 100 [7]. Although J.N. Marjolin in 1828 reported tumours occurring in burn scars of long duration, it was Dupuytren who noted that it was a malignancy [8]. Da Costa in 1903 first coined the term “Marjolin’s ulcer”, applying it to tumours arising in simple leg ulcers [9]. Several authors have reported that it is not rare [2,10], and some Nigerian workers report that the lesion represents up to 30% of all primary skin cancers seen in our environment [5].

The scalp is an uncommon place for scar cancer; they are most commonly found in the lower limbs. Occasionally they occur around the upper limbs, head region and the trunk [5,11–14]. Previous studies in this centre agree with this [6]. Few reports in children are available. Penetrating lesions limited to the scalp have been reported previously in male children [2],



**Fig. 2 – X-ray picture of skull.**

and that involving bone and dura mater in adults [3,15,16]. The most frequent predisposing lesion in the scalp is a post burn wound, but posttraumatic wounds are infrequently implicated [4,16]. The cause is frequently flame burns, but electrical burns and other thermal burns have been reported [17]. In this case it was flame burns, an unusual cause of burn injury in the infant, which frequently results in full-thickness injury in that age group.

#### 4. Epidemiology

Marjolin's ulcers are more common in males [5,11,18,19]. The mean age is between 50 and 59 years [11,12,20]. The ulcer appears to be affecting younger patients over the years. The mean age and transition time are apparently lowering in Africa [5,6,10,14,19]. In the scalp the age range is from 9 to 74 years [2,15]. This patient is the youngest in our environment, and is female. In the scalp transition time is quite variable depending on the study reviewed, and has varied from 6 months to 65 years [15,21]; similar to reports of 3 months to 67 years for all parts of the body [7,11,22,23].

#### 5. Aetiology

The aetiology is believed to be multifactorial. Slow healing and scar instability characterised by chronic irritation and the induction of a constantly proliferating epidermal unit have been blamed [7]. Repeated cycles of healing and break down, wounds that never healed, ulcers arising at the border between pigmented and depigmented scars and treatment with irritant native medication as in this patient are reported risks [6,18]. It has been postulated that these result in a reduced ability to withstand carcinogens [7,20]. The relatively avascular scar tissue may then act as an immune privileged site that allows the tumour to resist the body's usual defenses against foreign cells [24]. There are similarities also in the formation of scars by fibrosis and tumourigenesis [25].

The rich vascularity of the scalp may be responsible for its relatively low incidence of Marjolin's ulcer [3,4], and may also account in part for the very poor outcomes in lesions that have broken free of the scar, as ready routes for systemic spread are present. Ultraviolet rays which lead to a reduction in Langerhans cell population and cutaneous immuno surveillance against incipient neoplasm [26,27], and alterations in the p53 tumour suppressor gene are implicated in the aetiology [28-30].

#### 6. Presentation

They are commonly present as non-healing ulcers in post burn or other posttraumatic wounds. Even though the lesions are often painless, unprovoked bleeding, offensive discharge and increasing pain are sometimes associated with the ulcer [31]. They may be due to infection but may also indicate that the tumour has escaped the confines of the scar [6]. Bleeding from the primary lesion is associated with recurrent disease, even

in the absence of clinically positive nodes [3,4]. This patient classically complained of pain before the first surgery, and following recurrence. In both instances it was noted that the lesion was infiltrative.

#### 7. Metastasis

When the tumour is confined to the scar the growth is slow and it can be completely cured. Once it breaks free of the scar it metastasises rapidly via the regional nodes [32].

#### 8. Diagnosis

This follows a biopsy. Even though squamous cell carcinoma is the commonest in the scalp; occasionally other malignancies have been reported [3-7,18,33]. Literature seems to suggest histological types other than squamous cell carcinomas are less frequent in the scalp than elsewhere [3,4]. The histological features are characteristic [34].

#### 9. Treatment

The outcome for scalp Marjolin's is often dismal with early recurrence and death [3,35]. There is as yet no consensus on the protocol of managing this aggressive lesion [3,4,17]. Early recognition offers the best chance for cure and wide local excision alone in early lesions may be curative [12,36] since at this stage it is confined to the scar. A margin of 2-5 cm has been advocated [4,25]. In this patient complete excision with a 3 cm margin as well as excision of the scars did not prevent recurrence. Following excision split skin grafts are preferred to cover the defect. This aids early detection of recurrence. Some guidelines have been offered for reconstruction of the scalp [4] postexcision and include:

1. Small defects (<3 cm) closed primarily with a wide undermining of the scalp.
2. If the defect is large and the pericranium is intact, split-thickness skin grafts and later skin expansion.
3. Local flaps for full-thickness scalp defects.
4. In very large defects of the scalp or when the defect includes full-thickness calvarium: free tissue transfer.

The post excision defect in this patient left the brain exposed at the base of the wound. This defect was however covered by modified orthochea flaps. Free tissue transfer had not commenced in our centre or sub-region at this time and was not done. Following recurrence the resulting defect was grafted.

Controversy exists regarding the place and timing of lymph node biopsy [1]. Sentinel lymph node biopsy has been shown to give as high yield as 83% [37] and is recommended to detect occult nodal involvement. Once the sentinel node biopsy is positive the lesion is late.

Early recurrence within four months after excision and death within 10 months of diagnosis has been reported despite apparently adequate excision [3,35,38]. Most patients succumb

within 2 years [3]. Recently gene therapy has shown some promise [39].

## 10. Conclusion

Marjolin's ulcers of the scalp are aggressive tumours. Poverty and ignorance play significant roles in its development, and severely limit the evaluation and management in third world sub-Saharan centres such as ours. Health education is needed to discourage late presentation.

## Conflict of interest

The authors have no financial interests in the publication or otherwise of this article.

## Acknowledgements

**Contributions.** BO assisted in the surgery, and contributed to the case report and references sections. WIBO read the histology slides, gave the histological reports and contributed to the literature review. IIO was the surgeon and conceived the study, contributing to all aspects of the article and is responsible for the format, language, and editing.

All the authors approved the final draft.

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