Hong Kong J. Dermatol. Venereol. (2013) **21**, 134-138

Case Report

Fatal volcanic squamous cell carcinoma of the scalp

頭皮上致命的火山狀鱗狀細胞癌

A Tomizawa 殷衍伶, S Lee 李啓焱, R Cocciolone

Cutaneous squamous cell carcinoma of the head and neck is an invasive skin cancer with significant morbidity and mortality, chiefly among older and immunocompromised patients. We present a case report of a patient without obvious immune suppression developing multiple, eruptive cutaneous squamous cell carcinomas of the scalp. Despite multiple excisions, radiotherapy and adjuvant oral retinoids, the patient developed regional parotid metastasis and died of his disease within thirteen months. This case serves as a stark and tragic reminder that painful, eruptive cutaneous squamous cell carcinoma of the head and neck must be managed actively due to the potential for rapid progression and a fatal outcome.

頭頸的皮膚鱗狀細胞癌是一種有顯著發病及死亡率的侵入性皮膚癌，多見於年老及免疫力低下的患者。我們報告一名沒有明顯免疫力受抑制的病人在其頭皮叢生的發疹性皮膚鱗狀細胞癌。儘管多次的切除手術、放射治療並輔以口服維甲酸治療，病人仍然發生癌細胞的局部腮腺轉移，並於發病後十三個月離世。這宗病例實在是一個悲切的提示，我們必須積極治理頭頸上伴有痛感的發疹性皮膚鱗狀細胞癌，以對抗其潛在的快速惡化和致命特質。

Keywords: Acitretin, fatal outcome, radiotherapy, squamous cell carcinoma

關鍵詞：依曲替酸、致命結果、放射治療、鱗狀細胞癌

Royal North Shore Hospital, NSW, Australia
A Tomizawa, BA, MBBS(Hons)

Royal Prince Alfred Hospital, Central Clinical School, The University of Sydney, Camperdown, NSW, Australia
S Lee, AM, DDM(Syd), FACD

Dubbo Dermatology, Dubbo, NSW, Australia
R Cocciolone, MBBS(Hons), FACD

Correspondence to: Dr. A Tomizawa

Royal North Shore Hospital, NSW, Australia

Introduction

Cutaneous squamous cell carcinoma (SCC) of the head and neck is not uncommon, particularly amongst the elderly and immunocompromised. Its aggressive biological activity is based on damaged host immune status as well as tumour size, tumour cell type, depth and anatomical sites. Tumours typically appear on sun-exposed areas of the scalp, ears, face and the neck. Despite its well-recognised metastatic potential, transformation in an explosive, painful and fatal

way is fortunately rare. We describe a case of rapidly progressing cutaneous squamous cell carcinoma of the scalp in an apparently immunocompetent patient that was ultimately fatal.

Case report

An 83-year-old Caucasian male presented with an anterior scalp tumour that was suggestive of a keratoacanthoma clinically. His past medical history was significant for chronic obstructive pulmonary disease, renal insufficiency, thromboembolic disease, gout, osteoarthritis and chronic polymyalgia rheumatica, for which he took prednisone 3 mg daily for twelve years without adverse effects. The initial scalp tumour was excised, and histopathology confirmed an ulcerated, moderately-differentiated squamous cell carcinoma which extended into the subcutis, with clear margins.

The patient presented six months later with a crop of recurrent scalp tumours. Clusters of crateriform tumours measuring between five and ten centimetres in diameter appeared on the anterior and lateral aspect of the scalp (Figure 1). The tumours were inflamed with purulent discharge and contained large keratin cores. A subsequent scalp biopsy revealed poorly-differentiated, deeply penetrating SCC with focal infundibulocystic differentiation on haematoxylin & eosin (H & E) stain (Figures 2a & 2b). Computed tomography (CT) imaging demonstrated tumour abutting the skull table without bony involvement and excluded metastatic disease. The patient was admitted to a tertiary referral-centre for multi-disciplinary care and commenced on oral acitretin 50 mg daily as well as oral antibiotics.

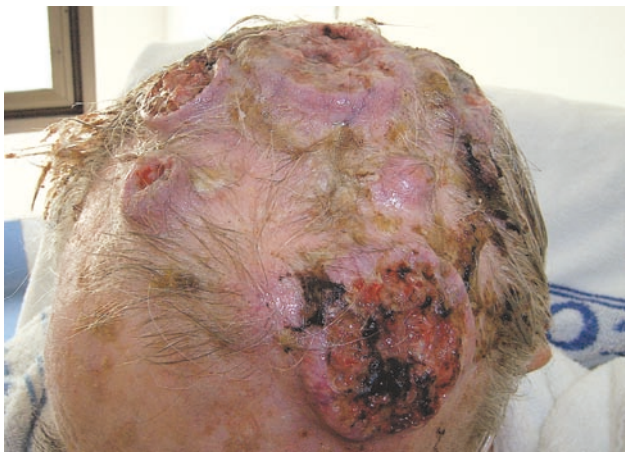

Figure 1. Cutaneous squamous cell carcinomas of the scalp.

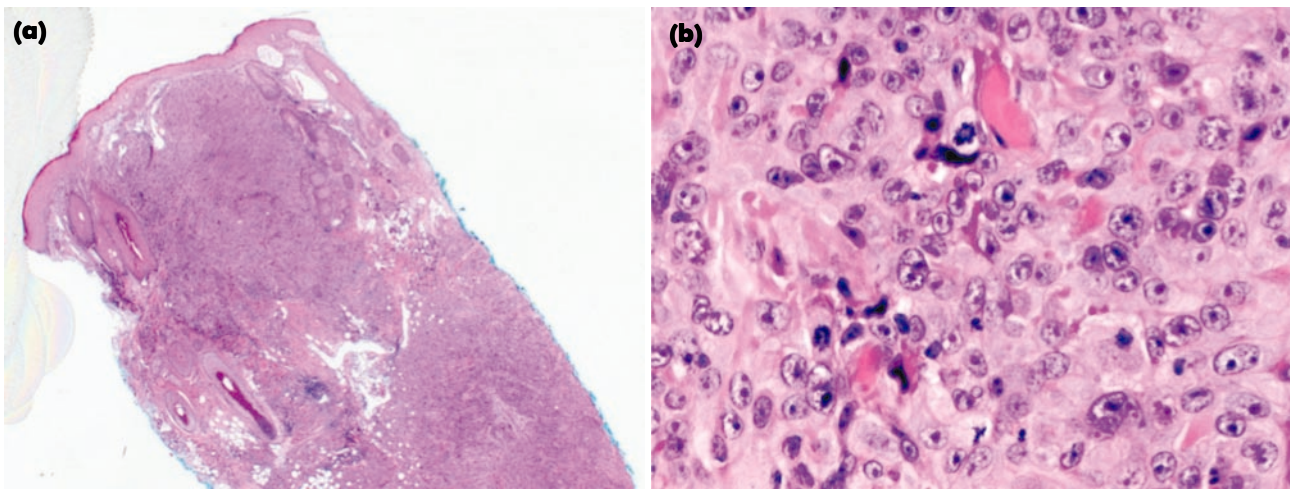

Figure 2. (a) Punch biopsy specimen demonstrating a deeply penetrating squamous cell carcinoma (haematoxylin & eosin (H&E) stain, original magnification x 1.25). (b) High-power view of poorly-differentiated squamous cell carcinoma (H&E stain, original magnification x 40).

Given the extensiveness of the lesions and the patient's anaesthetic risk, he was deemed unsuitable for surgery. Therefore, radiotherapy was administered to the involved site at 45 Grays in 15 fractions for three weeks with improvement in swelling and a reduction in both diameter and height of the lesions (Figure 3). Following radiotherapy adjuvant oral acitretin was recommenced, but the dose was limited to 25 mg daily due to mucous membrane side-effects, predominantly eye irritation.

Six weeks following radiotherapy, the patient presented with nausea and malaise, and was found to have a firm mass in the right parotid region on CT imaging (Figure 4). Histopathological examination of a fine-needle aspiration biopsy specimen from the parotid mass demonstrated malignant keratinising cells with keratin pearls and background necrosis. Tumour cells had irregular nuclei, abnormally distributed chromatin and increased nuclear-to-cytoplasmic ratio, consistent with metastatic, poorly-differentiated SCC. Staging CT imaging excluded distant metastases.

Radiotherapy was administered to the right parotid lymph node and the patient was recommenced on oral acitretin following discharge. He re-presented to hospital six months later with increasing scalp pain and general malaise. Palliative radiotherapy was administered to the left anterior scalp lesion for palliative pain relief. Unfortunately, the patient passed away from medical complications of active palliative care treatment thirteen months following initial presentation of his index scalp lesion.

Discussion

This report describes a rare case of aggressive cutaneous squamous cell carcinomas of the scalp in a patient without significant immunosuppression. Risk factors for cutaneous SCC of the head and neck include Caucasian background, male gender, age over 65 years, cumulative sun exposure and immunodeficiency.[1] Other cutaneous malignancies and squamoproliferative tumours, such as "atypical fibroxanthoma" share

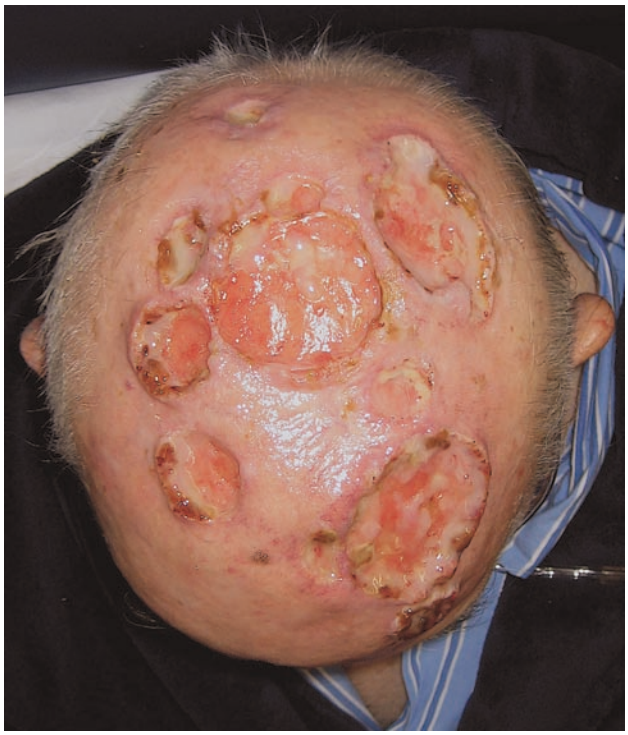

Figure 3. Cutaneous squamous cell carcinomas of the scalp, after radiotherapy.

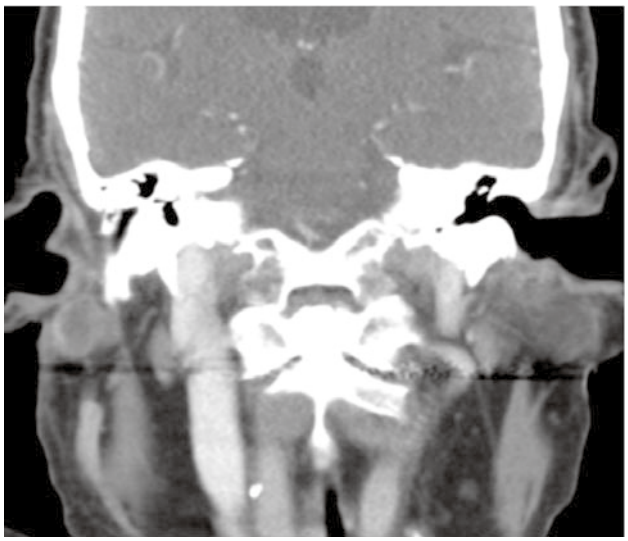

Figure 4. CT image of a local metastasis of cutaneous SCC to the right parotid lymph node.

these risk factors.[2] Although our patient did have multiple risk factors, it is unclear whether his taking 3 mg daily of oral prednisone was sufficient to increase his risk of developing advanced cutaneous malignancy. Indeed, many patients with chronic inflammatory diseases such as asthma have taken long-term oral steroids of up to 5 mg daily prednisone without a demonstrable increase in the risk of cutaneous SCC. While dermatological data is scarce, a recent study reported in the rheumatology literature indicated that 10 mg daily of low-dose oral prednisone had minimal side-effects in patients with rheumatoid arthritis.[3] Further research in this therapeutic area may clarify this issue in the future.

It has been thought that SCC of the head and neck greater than 2 cm in diameter are twice as likely to recur locally and three times as likely to metastasise as smaller tumours.[4] Other risk factors include tumours located in sun-exposed sites, tumour depth over 4 mm, histological subtype of poorly-differentiated SCC and compromised host immune status.[5] While our patient had multiple lesions over 2 cm, the question of why he developed so many eruptive SCCs that grew explosively but without bony involvement, remains unanswered. A recent case report described multiple synchronous SCC on the head and neck less than 1.5 cm in diameter occurring in an elderly Caucasian patient with a history of high-grade non-Hodgkin's lymphoma.[6] Despite multiple excisions with clear margins, over 40 primary SCC continued to develop over the patient's scalp and face, and the patient died of his disease. To our knowledge, our patient is the first case of multiple, large, volcanic cutaneous SCC of the head and neck reported in a patient without a significant past history of immunosuppression.

The current recommendations for management of aggressive cutaneous SCC of the head and neck with nodal metastases include both surgical treatment and adjuvant radiotherapy. Single modality treatment alone is associated with a worse therapeutic outcome.[7] In our case, surgery was contraindicated because of his age and the extent of disease. Accordingly, radiotherapy was administered with the aim of retarding disease progression. While doses for adjuvant radiotherapy of 60 Grays have been advocated for nodal metastases, our patient received a lower dose of 45 Grays to his primary lesions, as recommended by the radiation oncology team in order to limit cutaneous side-effects.[8] Although no standardised recommendation for adjuvant chemotherapy currently exists, there is emerging data for platinum-based chemotherapy combined with adjuvant radiotherapy in mucosal SCC of the head and neck.[9]

A striking feature of this case was the rapid evolution from presentation to development of regional metastasis, culminating in the fatal outcome within thirteen months. Moreover, the patient complained of unrelenting pain, which is not a common feature of cutaneous SCC.[10] The senior author notes that in his experience, this demoralising symptom bears a poor disease prognosis.

While we know that cutaneous SCC of the head and neck may invade surrounding structures and metastasise, transformation in such an eruptive, painful and fatal way is fortunately rare. This case raises awareness of three key issues: the guarded prognosis of large and painful SCC of the scalp, particularly in the elderly and including those who are not significantly immunosuppressed; the use of acitretin as an ancillary therapy to impede progression of this form of skin cancer; and the importance of high-quality palliative care in looking after dermatological patients with advanced and metastatic malignancy. In clinical medicine, compassion is vital!

Acknowledgements

We are indebted to Associate Professor Steven Kossard and Dr. Penelope Lim for assistance with preparing the histopathology slides.

No funding sources supported this work. No sponsors had a role in the design and conduct of the study, in collection or interpretation of data, or in the preparation or approval of the manuscript.

References

1. D'Souza J, Clark J. Management of the neck in metastatic cutaneous squamous cell carcinoma of the head and neck. Curr Opin Otolaryngol Head Neck Surg 2011;19:99-105.
2. Avolio AM, Lee S, Mann S. Sixteen photogenic wonders from Down Under: a case series of Atypical Fibroxanthoma from Australia. Hong Kong J Dermatol Venereol 2011;19:167-74.
3. Conn DL. Resolved: Low-dose prednisone is indicated as a standard treatment in patients with rheumatoid arthritis. Arthritis Rheum 2001;45:462-7.
4. Rowe DE, Carroll RJ, Day CL Jr. Prognostic factors for local recurrence, metastasis and survival rates in squamous cell carcinoma of the skin, ear and lip. J Am Acad Dermatol 1992;26:976-90.
5. Motley R, Kersey P, Lawrence C; British Association of Dermatologists; British Association of Plastic Surgeons; Royal College of Radiologists, Faculty of Clinical Oncology. Multiprofessional guidelines for the management of the patient with primary cutaneous squamous cell carcinoma. Br J Dermatol 2002;146: 18-25.
6. HyunBaek J, Abu-Serriah M, Ameerally P. Multiple synchronous cutaneous squamous cell carcinomas of the head and neck: a case report. J Oral Maxillofac Surg 2011;69:e317-e23.
7. Veness MJ. Treatment recommendations in patients diagnosed with high-risk cutaneous squamous cell carcinoma. Australas Radiol 2005;49:365-76.
8. Howle JR, Morgan GJ, Kalnins I, Palme CE, Veness MJ. Metastatic cutaneous squamous cell carcinoma of the scalp. ANZ J Surg 2008;78:449-53.
9. McNab AA, Francis IC, Benger R, Crompton JL. Perineural spread of cutaneous squamous cell carcinoma via the orbit. Clinical features and outcome in 21 cases. Ophthalmology 1997;104:1457-62.
10. Cooper JS, Pajak TF, Forastiere AA, Jacobs J, Campbell BH, Saxman SB, et al. Postoperative concurrent radiotherapy and chemotherapy for high-risk squamous cell carcinoma of the head and neck. N Engl J Med. 2004;350:1937-44.