Introduction

The association between thermal burn scars and neoplasia was initially recognized by Celsus in AD 100 at the sites of chronic pressure ulcers. Jean-Nicolas Marjolin described in 1828 the formation of ulcers specifically in chronic burn scars. It was De Costa in 1903 who coined the term “Marjolin’s ulcer”, applying it to tumours arising in simple leg ulcers. Although he did not specifically describe cancer, the term Marjolin’s ulcer has been used to name a carcinoma originating in any type of scar tissue and is now synonymous with malignant transformation, usually ectodermal and rarely mesenchymal, of chronic ulcers, sinus tracts, and burn scars. The most common form of malignancy arising from a chronic wound varies in the literature and appears to be dependent on the aetiology of the wound. Squamous cell carcinoma is a well-known and rare complication of chronic ulcers, sinuses, chronic osteomyelitis, radiotherapy, and burn scars, and may complicate chronic pressure ulcers as well as cystostomy sites and Fournier’s gangrene scars. Basal cell carcinoma, on the other hand, is well documented in ulcers associated with venous insufficiency. The majority of Marjolin’s ulcers, however, arise from burn scars. Although they consist of all kinds of skin cancer, squamous cell carcinoma is the main cancer type reported in the literature.

The pathogenesis of carcinoma arising in a burn scar is not completely known. It could be due to chronic irritation of the affected area, and mostly involves the extremities and scalp area. Repetitive trauma and a prolonged healing phase may predispose an area to malignant transformation. Most Marjolin’s ulcers have been described as occurring in full-thickness burn scars that remain ungrafted or suffer graft failure. In most cases, only a focal portion of the scar undergoes malignant changes. This increases the risk of obtaining a false-negative diagnosis. Moreover, the latency from primary thermal trauma to the malignant transformation is about 30 years.

Particularities of this tumour reported in the literature include the high rate of lymphatic metastasis and recurrence, as well as the poor prognosis. Squamous cell carcinomas resulting from Marjolin’s ulcers can have an aggressive course and a much greater tendency to metastasize than squamous cell carcinomas arising from other causes, which makes early diagnosis imperative.

Case report

A 68-yr-old man presented with chronic ulceration of a burn scar over the scalp 20 years after sustaining an electrical burn injury. The patient related that the original injury resulted in a full-thickness burn wound that was treated by sequential soft tissue and bone debridements, burring of the skull, a prolonged period of dressing changes, and repeated skin grafts. Complete healing was only achieved several months later, mainly by secondary intention. Two years prior to presentation he started to note small scar ulcerations that would heal with local topical treatment, only to recur later. Serious ulcerations developing six months earlier failed to heal despite continuous oral antibiotics and topical therapy (Fig. 1a). Multiple punch biopsies performed in Montreal, Canada, failed to demonstrate a frank malignancy; however, the pathology report described necrotic tissues with few cells with signs of anaplasia. There was no evidence of any distant metastasis. A skull CT scan revealed, however, lytic bony lesions involving the skull near the vertex (Fig. 2).

The extent of the scalp lesion and its location over the vertex necessitated a major reconstruction post-resection. Owing to the patient’s history of coronary artery disease and previous bypass surgery, lengthy microvascular reconstruction was precluded. Excluding the random multistaged pedicled jump flaps described several years ago, which at present are rather of historical interest, none of...
cisional defect. Donor sites were covered by split-thickness skin grafts. All wounds healed uneventfully (Fig. 3).

Discussion

In general, risk factors for cutaneous squamous cell carcinoma include long-term exposure to ultraviolet or ionizing radiation or arsenic and repeated contact with polycyclic hydrocarbons. When such cancers occur in chronic lesions caused by burns, ulcers, or infection of traumatic wounds they are called Marjolin’s ulcers. These are rare tumours in which various aetiological factors have been incriminated, including repeated irritation, poor lymphatic regeneration, antibodies, mutations, and local toxins. More recently, a theory of immunological isolation has been suggested, whereby lymphatic channel obliteration at the site of injury may decrease the delivery of antigen or specifically stimulated small lymphocytes to the regional lymph nodes from that site. This renders the site “immunologically privileged”, allowing the development and progression of antigenically foreign tumour cells to go unchecked. Such cells may initially arise by spontaneous mutation or develop under the influence of viral or chemical carcinogens. Tumour antigen recognition may then be delayed long enough for tumours to reach “critical size”, when immune mechanisms are no longer sufficient to prevent continued neoplastic progression. Nevertheless, confusion continues about the precise pathophysiology and clinical behaviour of Marjolin’s ulcer. The reported mortality and morbidity rates are also conflicting.

The majority of reported cases of Marjolin’s ulcers (75–96%) are squamous cell carcinoma, but basal cell carcinoma should be considered as well (1–25%); melanoma and sarcoma have been reported less frequently. There are two variants of this type of cancer: one is an acute form, in which the cancer occurs within one year of the date of injury, and the other a chronic form in which malignant changes are seen much later. In the former, the average latency is four weeks to one year, while in the latter the average period is 36 years (range, 1–75). Burn scar carcinoma has a propensity for the extremities, specifically for flexion creases, where blood supply may be decreased and trauma increased. The high vascularity of the scalp may explain why scalp Marjolin’s ulcers have a relatively low incidence, pointing to the possible role of diminished blood supply in this lesion’s aetiology.

Since Marjolin’s description, management of this carcinoma has remained controversial. When following patients with chronic ulcers and wounds, it is important to evaluate any changes immediately with biopsies and further imaging studies if indicated in order to treat them effectively. A biopsy should be performed without delay in cases of suspicious degeneration. Awareness of the malignant potential associated with chronic ulceration in scar
tissue may allow early diagnosis and a decrease in the morbidity associated with advanced disease, such as radical node dissection or amputation.

As would be expected, there is a wide variety of suggested treatment protocols for this disease. However, early diagnosis and radical excision are essential requirements. A multitude of options and recommendations exists for the management of both primary lesions and regional nodal metastasis. Radical excisional surgery is the most appropriate and optimal treatment, but there is no consensus on the indications for lymph node dissection. Given its multifocality, aggressive resection of the entire burn scar as of the tumour is advocated, with precise margin control in order to prevent the development of further neoplasms within the affected scar. However, even the most aggressive surgical intervention will sometimes be inadequate in the treatment of these tumours. Initially aggressive radical treatment may eventually be associated with radiotherapy. This treatment must be integrated in an elaborated therapeutic strategy, taking into consideration the evolutive potentiality of such tumours in order to improve chances of recovery and survival. Reconstruction, on the other hand, is critical for improved quality of life and can be accomplished by skin grafting whenever feasible as this method allows early detection of any recurrence. Some, however, prefer primary skin closure with the variation of a complex linear closure or employing a flap technique.

Locally advanced cutaneous malignancy of the scalp and forehead requires an aggressive approach to resection and reconstruction. Fortunately, the scalp is a rather uncommon site for Marjolin’s ulcers. The unique anatomy of scalp tissue makes it difficult to close excisional defects unless the defect is very small. Not infrequently, scalp reconstruction after ablative surgery can be challenging. Important tenets for successful management of

Fig. 3 - Reconstruction result at two weeks with bilateral anteriorly based parieto-occipital scalp flaps. Donor site covered with STSG.
sculpt defects are durable coverage, adequate debridement, preservation of blood supply, and proper wound drainage. However, a useful reconstructive algorithm is still lacking.

A wide array of coverage options is available. Techniques for reconstruction include split- or full-thickness skin grafting, local or regional flaps, tissue expansion, and free tissue transfer with variations and combinations thereof. Many of these options involve multiple procedures, significant donor site morbidity, complex surgical techniques, attendant blood loss, and other associated surgical risks. Not infrequently, skin grafting fails or simply may not be applicable or possible. In such cases, complex flaps or the importation of vascularized tissue may be required. Optimal techniques for the management of most scalp defects include one to two flap rotation-advancement flaps for small to moderate-sized defects and microvascular free tissue transfer or tissue expansion for larger defects.

The multiple-flap reconstruction method as described by Ortolocohea may be useful in a small subset of patients such as older, severely debilitated patients who would be optimally treated with microvascular tissue transfer but cannot tolerate lengthy general anaesthesia and young patients who will not accept the significant area of alopecia that might result from other techniques, such as secondary intention, skin grafts, or free flaps. Pre- and post-operative radiation therapies, as also the need for expedient tumour resection and immediate flap coverage, are issues that make free tissue transfer attractive for the oncology patient who needs scalp reconstruction. In order to minimize the risk of free flap failure in difficult cases, a preliminary arteriovenous fistula with subsequent free tissue transfer can be an extremely valuable reconstructive option. Cranioplasty is not usually required but, if needed, alloplastic or autologous cranial reconstruction does not appreciably increase morbidity in the patients selected.

The patient we are describing presented some significant challenges due to the large extent of the lesion and the presence of bony erosion overlying the sagittal sinus. Exposure of brain tissue and possible sagittal sinus injury following tumour ablation should not be overlooked. These are two extremely serious complications that are potentially lethal. It was hence essential, prior to tumour excision, to secure safe and adequate tissues for coverage capable of salvaging the worst of situations. Any flap necrosis, or any shortage of tissues for complete coverage, may prove to be disastrous for the patient. The concept of “flap prefabrication” is the ultimate option for this particular patient, tissue expansion being safest though not without complications.

RÉSUMÉ. En 1828 Jean-Nicolas Marjolin a décrit la formation d’ulcères en manière spécifique dans les cicatrices chroniques des brûlures, et en 1903 De Costa a inventé le terme «ulcère de Marjolin», qu’il a appliqué aux tumeurs qui se produisent dans les simples ulcères de la jambe. Les Auteurs décrivent les causes de cette condition, comme aussi les protocoles du traitement, et présentent l’étude d’un cas particulier.

BIBLIOGRAPHY