Background and aims

Atypical wounds comprise approximately 20% of all chronic wounds. With an aging population and increasing rates of comorbidity, these numbers are expected to rise. Atypical wounds are those wounds that do not fall into a typical wound category (i.e., venous, arterial, mixed venous and arterial, pressure or diabetic foot ulcers). They are a broad spectrum of conditions or diseases caused by inflammation, infection, malignancy, chronic illnesses or genetic disorders.

An atypical wound can be suspected if the wound has an abnormal presentation or location, pain out of proportion to the size of the wound and does not heal within 4 to 12 weeks following a good treatment plan. Unfortunately, the diagnostic delays associated with these wounds can be considerable, which leads to higher mortality rates.

With the above in mind, it is important that every healthcare professional treating these wounds is familiar with this concept, or at least has the knowledge to suspect an atypical wound and knows when to refer the patient to an expert. A multidisciplinary team is needed to manage this group of patients, but it is mainly community healthcare practitioners who manage these patients on a day-to-day basis. Early diagnoses and referrals to dermatologists are important, as they are experts in diagnosing and treating these wounds. After an exact diagnosis is made, a holistic assessment and interdisciplinary plan of care is essential for cost-effective management and to prevent recurrence.

From the patient perspective, atypical wounds can be very painful and have prolonged healing times, which lead to impaired quality of life. Many patients suffer daily with physical challenges such as odour, exudate, pain and reduced mobility, and psychologically with negative emotions, loneliness and depression. Mortality rates are higher not only due to their comorbidities, but also due to lower socio-economic circumstances, which lead to higher rates of suicide.

Still, there is a dearth of literature reporting on quality of life or health economy in this patient population. In response to this lack of uniform data, EWMA has established a working group to gather the best available knowledge on atypical wounds.

This document is targeted at increasing awareness of the clinical picture, diagnosis and treatment of...
these wounds among healthcare professionals and to provide practical advice on some of the chal-
gen  lenges that typically arise, such as delays in diagnosis of inflammatory and vasculopathy wounds (such as pyoderma gangrenosum (PG), an inflammatory neutrophilic disorder, and cutaneous vasculitis). We hope that a systematic approach will improve the care and quality of life for this patient group. Lastly, it is also hoped that this document will act as a catalyst in the management of atypical wounds and fill the void that currently exists in clinical decision making.

The document focuses on atypical wounds caused by inflammation, malignancy and chronic illnesses.

The aim of the document is to:

- Present the diagnostic criteria, comorbidities and diagnostic tools for wounds defined as ‘atypical’, including practical hints for healthcare professionals.
- Present the best available documented current treatment options. High-quality evidence is sparse, but there are retrospective and observational studies and some randomised prospective studies upon which we may draw.
- Present some newer treatment options for atypical wounds.
- Reduce the diagnostic delays of these wounds by providing up to date, evidence-based literature on atypical wounds and an algorithm to aid clinicians in assessing these wounds in a systematic way.

**Document content**

The document contains different chapters for the following atypical wounds: Pyoderma gangrenosum (Figures 1a and b), Vasculitides (Figure 2), Occlusive vasculopathy, Martorell HYTILU (Figure 3) and calciphylaxis, Hidradenitis suppurativa, malignant wounds, artefactual ulcers, Ecthyma and ecthyma gangrenosum. For these atypical wounds, pathophysiology, clinical presentation, diagnosis and treatment are described. In addition, several clinical pictures are presented, in order to make these wounds more recognisable. Other types of atypical wounds are covered briefly in one chapter. There are also separate chapters for the histology of atypical wounds, the topical treatment of atypical wounds, the patient perspective and health economy and organisation.

As atypical wounds consist of heterogeneous diseases, the author group considered it important to have one chapter that summarises the most important aspects related to suspecting, diagnosing and treating these challenging wounds. Therefore, one chapter is titled ‘Practical aspects of diagnosing and treating atypical wounds’. The chapter highlights the importance of a patient’s history in diagnosing these wounds; a table for important comorbidities and medications is included. In addition, important clinical signs, such as hypergranulation, pathergy, violaceous and undermined borders, necrosis, atypical location, severe pain (out of proportion which is normally associated with wounds), livedo racemosa and reticularis and rapidly progressive ulceration are described. An algorithm of different types of atypical wounds is introduced. It is also stressed that, if the wound does not show signs of healing after 4–12 weeks despite optimal treatment (topical treatment, offloading, compression therapy), a biopsy should be taken from the wound edge. Drawings to show this are provided. However, it is important to remember that a negative histology does not exclude an atypical wound. A clinical assessment is required, and the patient should be referred to a dermatologist.

Once a diagnosis has been established, an interdisciplinary approach should be adopted to reach a successful outcome. This might include one or more of the following healthcare professionals: dermatologist, vascular and plastic surgeon, nephrologist, rheumatologist, tissue viability nurse or podiatrist. The exact nature of the treatment relies on the specific type of the atypical wound and will be presented in the individual chapters of this document. Where patients with a wound are being treated by immunosuppressive agents, negative pressure wound therapy (NPWT) and skin grafting can also be considered, but only after the inflammation has been reduced by immunosuppressive therapy. Also, topical corticosteroids may help to reduce excessive inflammation that impairs wound healing in atypical wounds, such as Pyoderma gangrenosum, vasculitides and Martorell HYTILU.

As the immunosuppressive agents can cause delayed wound healing, advanced therapies, epidermal grafting and surgical procedures are also important in terms of reducing healing time, it is possible to perform punch grafting, a traditional method for obtaining thin split-thickness skin grafts containing epidermis and papillary dermis.
Compression therapy (as provided by bandages, hosiery or compression wraps) can be beneficial for most leg ulcers with oedema, even if the cause of the wound is not venous. After exclusion of advanced peripheral arterial occlusive insufficiency, compression therapy should be used in all patients with lower leg ulcers. However, compression therapy may be painful in patients with inflammatory PG or vasculitides, therefore lower pressures of 20 mmHg should be used initially.

Conclusions and future perspectives
The key observations and recommendations of the document are presented in the final combined ‘Conclusions and future perspectives’ chapter, which addresses, among other topics:

- The need for prospective multicentre clinical trials with well-defined outcomes.
- The need for more precise understanding of the inflammatory and occlusive mechanisms of atypical wounds.
- The need for further studies of the benefits of traditional treatments, such as topical corticosteroids and tacrolimus.
- That the time of recurrence for ulcers, as well as their frequency, should also be taken into consideration when examining the effectiveness of treatments.
- That studies on diagnostic delays are important both from the patient’s and the organisational point of view.
- There is evidence of a positive outcome to early skin grafting in ulcers in the context of arteriopathy, with a breakdown in the vicious cycle necrosis–inflammation and consequent pain reduction and epithelialisation promotion.
- That organised multidisciplinary teams consisting of dermatologists, vascular and plastic surgeons, rheumatologists, diabetologists, infectious disease specialists, psychiatrists, tissue viability nurses, psychologists, nutritionists, physiotherapists and social care workers should be included in care pathways for these wounds.
- The importance of early suspicion and expert consultation with these wounds; a ‘wait and see’ attitude can lead to devastating outcomes.
- The proposal for an algorithm, presented in the chapter ‘Practical aspects of diagnosing and treating atypical wounds’ that will hopefully help with the daily practice, systematic assessment of these wounds.

We hope that the document will be disseminated and used among all healthcare professionals treating chronic wounds. We also hope that the document stimulates practitioners and scientists to adopt a translational research approach in the future.

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