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A 7-Year-Old Girl from West Africa With Two Skin Ulcers and a Contracture of Her Right Wrist

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Clinical Presentation

History

A 7-year-old girl is presented to a district hospital in the tropical region of a West African country. After an insect bite, she developed an itchy papule on the back of her right hand, which enlarged over a period of 3 months. A traditional healer had prescribed herbal remedies. When the lesion ulcerated a few weeks later, diclofenac and dexamethasone were administered at the local health post. When a second lesion appeared, treatment with oxacillin was initiated with no effect. The girl became increasingly unable to use her right hand. There is no history of relevant trauma or systemic symptoms.

Clinical Findings

A 7-year-old, anxious girl in good general condition holding her right wrist in a 45° flexion and 20° abduction position. Pulse 108 bpm (normal 70–110), blood pressure 100/70 mmHg, temperature 37.9°C (100.2°F).

Two skin ulcers are present on the back of her right hand (3 × 3 cm) and on the medial side of her right wrist (0.5 × 1 cm). The larger ulcer is filled with necrotic tissue (Fig. 41.1) and surrounded by hypo- and hyperpigmentation, lichenification and desquamation. An ill-defined induration surrounds the ulcers (12 × 8 cm) with oedema extending from the lower arm to the fingers. A 0.5 × 0.5 cm nodule is noticed above the medial right elbow.

Questions

1. What is the most likely diagnosis and what is the differential diagnosis?
2. What is the appropriate clinical approach in the given context?

Discussion

A 7-year-old West African girl presents with two progressive cutaneous ulcers linked by an area of altered skin. The movement in the associated joint is restricted. General symptoms are limited to a mildly elevated temperature.

Answer to Question 1

What is the Most Likely Diagnosis and What is the Differential Diagnosis?

The geographical region of West Africa, the young age of the patient, the location of the lesion on the extremities, the absence of major trauma and the clinical picture lead to a suspected diagnosis diagnosis of Buruli ulcer (BU). The history of an insect bite is an incidental finding, the mode of transmission of the causative organism, *Mycobacterium ulcerans,* remains unknown.

Pain and low-grade fever as seen in this case may also be explained by bacterial superinfection. A careful history and physical examination will provide guidance in differentiating numerous other infectious (bacterial, viral, fungal, parasitic) and non-infectious (trauma, envenoming, autoimmune, haematological, neoplastic) causes of ulcers in tropical countries. In areas endemic for BU, the accuracy of clinical diagnosis in experienced hands is remarkably high.

• **Fig. 41.1** Large skin ulcer on the back of the right hand filled with necrotic tissue; second smaller ulcer on the medial side of the wrist.

• **Fig. 41.2** At the end of specific treatment, the ulcer on the back of the hand had healed almost completely. However, the second, smaller ulcer had increased in size.

Answer to Question 2

What is the Appropriate Clinical Approach in the Given Context?

Adequate wound care, including pain relief, according to WHO guidelines should be instituted immediately. The desired sterile, moist atmosphere of the wound can be achieved with saline-soaked gauze changed daily. Colonization with other bacteria may be controlled by povidone-iodine. Antitetanus coverage must be secured. Written documentation of pain control and picture documentation of wound progress is helpful in achieving or maintaining a high-quality wound management standard.

Laboratory confirmation of BU by *M. ulcerans* using PCR is desirable but limited by the availability of reliable laboratory capacity and by its cost, which must be balanced with important supportive measures, such as improved nutrition.

The indication for surgical debridement depends on the clinical picture, but also on the availability of adequate anaesthetic and surgical care.

BU-specific antimycobacterial therapy should be commenced as soon as possible. In the absence of systemic symptoms, the toxicity of the recommended antibiotic treatment may justify a few days of delay until diagnostic results have been obtained.

The Case Continued…

PCR from a wound swab confirmed the presence of *M. ulcerans.* During the 8 weeks of specific treatment and wound care the ulcer on the back of the hand healed almost completely, but the second smaller ulcer increased to 8 × 10 cm in size (Fig. 41.2).

Debridement and skin grafting were performed, and the patient made an uneventful recovery. The restriction of movement was corrected by physiotherapy.

SUMMARY BOX

Mycobacterium ulcerans Disease (Buruli Ulcer)

M. ulcerans disease is a necrotizing infection mainly of the subcutaneous tissue. It is most prevalent among children and adolescents living in rural communities of West Africa; but other continents and temperate regions of Australia, China and Japan are also affected. Endemic areas are associated with water bodies such as rivers and lakes. However, the exact mode of transmission remains poorly understood.

M. ulcerans is characterized by its particular sensitivity to heat, its propensity to develop local and distant satellite lesions and its production of a necrotizing, locally immunosuppressive and analgesic macrolide exotoxin called mycolactone.

Clinically, *M. ulcerans* disease presents with skin lesions ranging from papules, nodules and plaques to the eponymous ulcers. The latter may involve most of a limb surface or trunk and can simultaneously occur at different body sites. All lesions share skin alterations including induration, hypo- and hyperpigmentation, lichenification, desquamation and possibly local oedema. Ulcers are characterized by undermined edges surrounding a 'cotton wool' like necrosis. Lesions are classically described as painless, but evidence has emerged questioning this doctrine. The diagnosis can be confirmed most reliably by PCR from a wound swab; however, a test for the presence of mycolactone suitable for district-level laboratories is under investigation.

Culture is difficult because of the slow growth rate of the mycobacteria.

The WHO recommends combination therapy with rifampicin (10 mg/kg per day PO) plus either streptomycin (15 mg/kg per day IM) or clarithromycin (7.5 mg/kg twice daily PO) for 8 weeks, which achieve high specific cure rates. Local heat application at temperatures >40°C for several weeks has also been shown to be curative. Special attention must be paid to patients co-infected with HIV, who are at increased risk for complications.

Careful wound management, pain relief, surgical excision and skin grafting as well as physiotherapy for restricted movements remain indispensable cornerstones of BU treatment. Extensive necrosis of subcutaneous tissue at diagnosis may cause a significant increase in ulcer size under treatment. Once secondary bacterial infection has been ruled out, this must not be mistaken for treatment failure. Local or distant new lesions may become evident during or after treatment. This so called 'paradoxical reaction' is thought to be caused by a local host immune reconstitution syndrome because of a fall in mycolactone levels. The distinction between bacterial secondary infection, recurrence and paradoxical reactions remains challenging and should involve expert advice.

Further Reading

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