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IMAGES IN CLINICAL PRACTICE

LEG ULCER IN A CHILD WITH SICKLE CELL DISEASE - IS IT A COMPLICATION OF THE DISEASE OR A CUTANEOUS SIDE EFFECT OF HYDROXYUREA TREATMENT?

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An 8-year-old girl with sickle cell disease (SCD) is under the care of a Pediatric Hematology Unit at a tertiary hospital. She has a history of multiple bone vaso-occlusive crises, some of which required hospitalization and two episodes of acute chest syndrome. The patient has been receiving hydroxyurea treatment at 20 mg/kg/day. After three months of therapy, she presented with a non-traumatic superficial leg ulcer on the internal malleolar region of her right foot. There were no reported cutaneous side effects aside from this ulcer and she did not experience other symptoms such as pain crises, anemia or jaundice. Physical examination revealed a superficial leg ulcer in the internal malleolar region of the right foot, clinically resembling an abrasion. It measured 2.0 cm by 1.5 cm and had irregular hyperpigmented edges, a pinkish base and peri-ulcer induration, as shown in Figure 1.

Figure 1. Leg ulcer in a sickle cell patient›s internal malleolar region of the right foot.

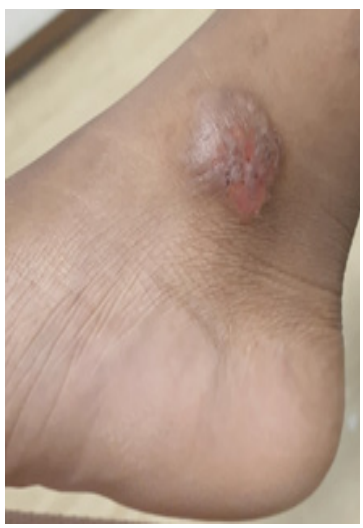

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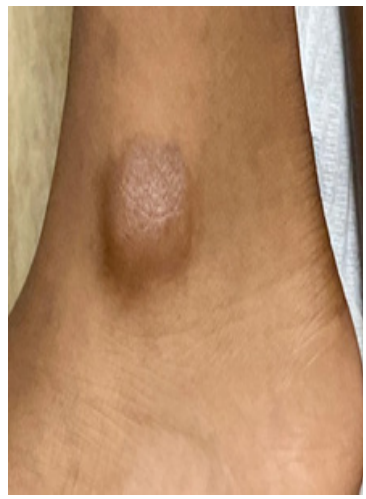
Figure 2. The ulcer's healing progress four weeks after discontinuing hydroxyurea treatment.



What is the diagnosis?

A leg ulcer is a classic and common complication of SCD, but it is rare before age ten.[1] Local factors and systemic dysfunction, such as vasculopathy and chronic inflammation, seem to impact its development.[2] Common locations for these ulcers involve the peri-malleolar area, with less frequency in the distal third of the lower extremity and rarely on the foot or digits.[2,3] Hydroxyurea (also known as hydroxycarbamide) is an oral chemotherapeutic medication employed to alter SCD treatment, mitigate organ-related issues and decrease acute complications, specifically pain events and the need for blood transfusions.[4] Although hydroxyurea-related leg ulcers have been infrequently reported, the mechanism behind the development of these ulcers remains unclear. Distinguishing a leg ulcer caused by hydroxyurea from one caused by SCD can pose a challenge, as both conditions can result in similar skin manifestations. Lesions associated with hydroxyurea often appear around the malleolar area, exhibiting a fibrinous appearance of inflammatory edges but without necrosis.[5] In this case, the diagnosis of leg ulcers as a cutaneous side effect of hydroxyurea treatment was considered, leading to the suspension of the treatment. Local

care was administered twice daily for four weeks, resulting in visible improvement in skin regeneration, as shown in Figure 2. After two months, it was possible to gradually resume hydroxyurea therapy while maintaining skin care to prevent the appearance of new lesions. Currently, the patient is taking a lower dose of hydroxyurea and has not experienced ulcers or crises. While a skin biopsy was not performed, the characteristics of the leg ulcer around the malleolar area, presenting with a fibrinous appearance but without necrosis, coupled with the rapid healing of the ulcer following the suspension of hydroxyurea treatment, strengthened our hypothesis. The advantages of using hydroxyurea to treat SCD generally surpass the risks associated with side effects. Nevertheless, hydroxyurea-induced ulcers are a rare complication and addressing this issue may require dosage reduction or discontinuation of the drug. Close monitoring and open communication with healthcare professionals are crucial to ensure a safe and effective course of treatment.

Compliance with Ethical Standards
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Conflict of Interest : None

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