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Pregnancy-Associated Exacerbation of Darier's Disease: A Rare Obstetric Case Report

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Case Report

Abstract

Introduction: Darier's disease is a rare inherited disorder of keratinisation. Reports describing its behaviour and clinical impact during pregnancy are limited, particularly in cases with extensive vulval involvement.

Case: A 39-year-old multiparous woman presented at 32 weeks' gestation with a widespread exacerbation of previously diagnosed Darier's disease, involving the vulva, groin, lower abdomen, submammary region, neck, and face. Severe vulval fissuring and reduced skin elasticity were noted. Obstetric evaluation demonstrated normal fetal growth. The patient was managed conservatively with topical therapy under multidisciplinary care.

Conclusion: Although rare, Darier's disease may worsen during pregnancy and present unique obstetric and perioperative challenges. Early recognition and coordinated management are essential to minimise morbidity and guide delivery planning.

Key Words: Darier's Disease, Pregnancy, Kratinization

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Introduction

Darier's disease, also referred to as keratosis follicularis, is a genetically determined disorder characterised by abnormal epidermal adhesion and keratinisation. It arises from pathogenic variants in the ***ATP2A2*** gene, which encodes a calcium pump essential for keratinocyte integrity and intercellular cohesion [1]. The fundamental defect in Darier's disease lies in impaired calcium signalling within keratinocytes due to dysfunction of the SERCA2 pump. This leads to disruption of desmosomal junctions and premature keratinocyte separation [4]. Histopathological features include suprabasal acantholysis and dyskeratosis, which account for the characteristic clinical appearance [4-6]. The disease follows an autosomal dominant inheritance pattern with variable phenotypic expression.

Population-based estimates suggest that Darier's disease is rare, with reported prevalence ranging from approximately 1 in 30,000 to 1 in 100,000 individuals [2,3]. Both sexes are equally affected, and the condition is non-infectious.

Darier's disease is characterised by mal-odorous, pruritic, greasy, hyperkeratotic papules and plaques involving seborrhoeic and intertriginous areas such as the scalp, trunk, groin, and perineum. Nail abnormalities and mucosal involvement may also occur [4,5]. Disease activity is known to fluctuate, with exacerbations triggered by heat, friction, infection, stress, and hormonal changes [5,6]. Despite its chronic nature, the behaviour of Darier's disease during pregnancy has not been extensively studied [7-8]. The available literature consists primarily of isolated case reports, and guidance on obstetric management remains limited [9-12].

Disease Course During Pregnancy

Published case reports demonstrate variable disease behaviour during pregnancy [10]. Some patients experience exacerbation of symptoms, while others show partial improvement or no significant change [6,9,11,12]. Hormonal fluctuations, mechanical stress on the skin, and immune modulation during pregnancy are believed to contribute to this variability.

Distribution and Severity

Cases involving the vulva, perineum, and lower abdomen appear to be associated with greater morbidity. Vulval disease may result in fissuring, maceration, and pain, leading to difficulties with ambulation, hygiene, and sexual activity [9,11]. These manifestations are particularly relevant in the obstetric context.

This report describes a rare pregnancy-associated exacerbation and discusses relevant obstetric considerations.

Case Presentation

A 39-year-old woman, gravida 3 para 2, presented at 32 weeks of gestation with progressive worsening of chronic skin lesions. Her obstetric history included one previous vaginal delivery followed by a caesarean section. She had been diagnosed with Darier's disease several years earlier but reported that the current pregnancy was associated with the most severe flare she had experienced.

Cutaneous examination revealed extensive hyperpigmented, verrucous papules and plaques involving the vulva, groin, perineum, perianal region, lower abdomen, submammary folds, neck, and forehead. Vulval involvement was particularly severe,

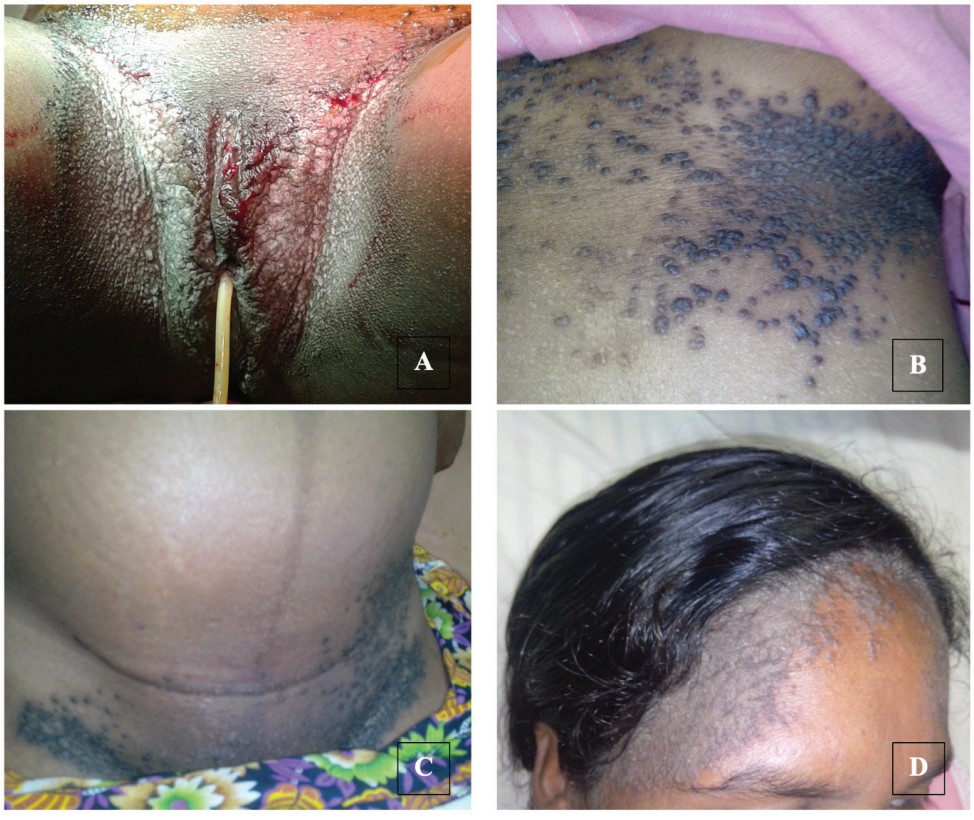

Figure 1. Figure Showing the lesions on Vulva (A), Submammary region (B), Suprapubic and groin area (C), and Scalp (D).

with marked fissuring, maceration, and reduced tissue elasticity, resulting in pain and functional limitation. No secondary infection was evident.

The uterus was appropriate for gestational age, and fetal movements were normal. Ultrasonographic assessment confirmed normal fetal growth and amniotic fluid volume, with no evidence of fetal compromise.

Following dermatological consultation, a diagnosis of pregnancy-related exacerbation of Darier's disease was made based on characteristic clinical features and prior documentation. Skin biopsy was deemed unnecessary.

Management focused on symptom control and prevention of complications. The patient was treated with regular emollients and topical corticosteroids of appropriate potency. Systemic agents, including oral retinoids, were avoided due to known teratogenic risks. Education regarding skin care, avoidance of friction, and early reporting of infection was provided. The patient reported gradual symptomatic improvement with conservative treatment. There was a significant improvement in the lesions following treatment, and

the patient subsequently underwent an uncomplicated caesaran section.

Discussion

This case illustrates an uncommon yet clinically significant exacerbation of Darier's disease during pregnancy. While hormonal influences on keratinocyte function have been proposed as potential triggers, the interaction between pregnancy-related physiological changes and disease severity remains incompletely understood [5,6].

Extensive vulval involvement has particular relevance in obstetric practice. Reduced skin elasticity and fissuring may predispose to traumatic vaginal delivery, impaired wound healing, and postpartum morbidity. Similarly, involvement of the lower abdominal skin may complicate surgical incision healing should caesarean delivery be required. These considerations highlight the importance of anticipatory planning and individualised decision-making regarding mode of delivery.

Anaesthetic implications also merit attention [9]. Widespread cutaneous involvement of the back may complicate neuraxial anaesthesia (Spinal anaesthesia), necessitating early anaesthetic assessment. In addition, disease involvement of the breasts or submammary region may cause pain, fissuring, or secondary infection, potentially interfering with breastfeeding. Supportive care and lactation counselling should be offered when appropriate [6].

Management during pregnancy is largely conservative. Topical therapies remain the cornerstone of treatment, while systemic retinoids are contraindicated. This case underscores the value of multidisciplinary collaboration between obstetricians, dermatologists, and anaesthetists to optimise maternal comfort and obstetric outcomes.

From a genetic standpoint, Darier's disease carries a 50% risk of transmission to offspring. Although prenatal diagnosis is not routinely indicated, affected women should be offered genetic counselling, ideally prior to conception or early in pregnancy, to support informed reproductive choices [1].

Conclusion

Darier's disease is a rare condition that may worsen during pregnancy and pose unique obstetric and perioperative challenges, particularly when vulval and lower abdominal skin are involved. Conservative dermatological management, combined with careful obstetric planning and multidisciplinary care, can minimise morbidity. Awareness of this condition among obstetric clinicians is essential to ensure timely recognition and appropriate counselling.

Conflicts of Interest

The author declares that there are no conflicts of interest.

Ethical considerations and consent for publication

This case report was prepared in accordance with the CARE (CAse REport) reporting guideline. Written informed consent for publication of the patient's clinical information and accompanying images was obtained from the patient after she reviewed the manuscript. All potentially identifying details have been removed from the text and images. Formal ethics committee approval was not required for publication of a single de-identified case report.

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Data Availability Statement

All relevant clinical information supporting this case report is included within the manuscript. No additional datasets were generated or analysed during the current study.

Statement on the Use of Artificial Intelligence

Artificial intelligence tools were used solely to assist with language editing and formatting of the manuscript. The author takes full responsibility for the content, interpretation, and scientific accuracy of the work.

Author Contribution

The author was responsible for the clinical management of the patient, conceptualisation of the case report, literature review, manuscript drafting, and final approval of the manuscript.

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