

A recalcitrant case of Jacquet erosive diaper dermatitis after surgery for Hirschsprung disease in a boy with Waardenburg-Shah syndrome

Goncagul Babuna Kobaner, Gizem Pinar Sun, Esen Ozkaya

Affiliations: Department of Dermatology and Venereology, Istanbul Faculty of Medicine, Istanbul University, Istanbul, Turkey

Corresponding Author: *Goncagul Babuna Kobaner MD, Department of Dermatology and Venereology, Istanbul Faculty of Medicine, Istanbul University, Istanbul, Turkey. 34093 Çapa-Istanbul, TURKEY, Tel: 90-212-4142238, Fax: 90-212-4142526, Email:*

goncagulbabuna@yahoo.com

Abstract

We herein present a four-year-old boy with Waardenburg-Shah syndrome who developed Jacquet erosive diaper dermatitis following a total colectomy and ileoanal anastomosis procedure for Hirschsprung disease. The diagnosis was made according to history and typical clinical findings. Complete resolution of the recalcitrant lesions after an ileostomy procedure supported the diagnosis. This case highlights the importance of being familiar with the predisposing factors and clinical presentation of this rare and severe form of chronic irritant dermatitis, since it may easily be misdiagnosed as other diseases in children and may lead to unnecessary diagnostic procedures, treatments, and anxiety due to suspicion of child abuse.

Keywords: Jacquet erosive diaper dermatitis, diaper dermatitis, Waardenburg-Shah syndrome, Hirschsprung disease

Introduction

Jacquet erosive diaper dermatitis is a rare and severe form of chronic irritant contact dermatitis of the perianal and genital regions [1]. We herein report a boy with Waardenburg-Shah syndrome who developed Jacquet erosive diaper dermatitis after surgery for Hirschsprung disease. The dermatitis was

resistant to several topical therapies, but completely resolved following an ileostomy procedure.

Case Synopsis

A four-year-old boy with Waardenburg-Shah syndrome presented with recalcitrant lesions in the diaper area. The lesions appeared after he started to wear diapers for chronic diarrhea following a total colectomy and ileoanal anastomosis procedure for the accompanying Hirschsprung disease. Dermatological examination revealed multiple well-demarcated papules and nodules, some of them showing central erosions on an erythematous base in the perianal region (Figure 1a). There was no preceding use of high-potency fluorinated corticosteroids. Various topical therapies including antifungals, antibiotics, low-potency corticosteroids, and barrier creams were not beneficial. A histopathological examination was planned but denied by the parents. Based on the history and clinical features, a diagnosis of Jacquet erosive diaper dermatitis was made. Protective measures and topical zinc oxide yielded only a partial response, whereas the lesions resolved completely shortly following an ileostomy operation to repair an ileal perforation (Figure 1b).

Case Discussion

Waardenburg syndrome is a rare genetic disorder causing congenital hearing loss and pigmentation



Figure 1. A) Multiple, bilateral, well-demarcated papules and nodules with a diameter of 2-5 mm in the perianal region on an erythematous base. Some of the lesions show central erosions. B) Complete resolution following an ileostomy procedure.

changes. If accompanied by Hirschsprung disease, which is extremely rare, it is called Waardenburg-Shah syndrome [2].

Jacquet erosive diaper dermatitis is a severe form of irritant contact dermatitis, which has been rarely reported in the literature [1, 3]. Predisposing factors include urinary and fecal incontinence, presence of detergent remnants in the diaper, infrequent diaper change, rough toilet paper usage, and chronic diarrhea [3]. Surgical procedures with ileoanal or coloanal anastomosis for Hirschsprung disease may result in watery diarrhea and activation of fecal enzymes by high pH; this may lead to various forms of chronic irritant diaper dermatitis including Jacquet erosive diaper dermatitis, granuloma gluteale infantum, and perianal pseudoverrucous papules and nodules (PPPN) [3]. The histopathology of these entities may show different findings such as epidermal hyperplasia, a dense mixed superficial and deep inflammatory infiltrate, and variable dilation and proliferation of blood vessels as in granuloma gluteale infantum [4]. Spongiotic psoriasiform dermatitis or acanthosis without a significant dermal inflammation may be observed in PPPN [4]. Nonspecific inflammatory findings are seen in **Jacquet's erosive diaper dermatitis** [4]. Yet, an

overlap of these above-mentioned histopathological findings has also been reported suggesting that there are no clear-cut histopathological criteria among these three entities [4]. Since these three disorders have common predisposing factors and overlapping clinical and histopathological features, it has been proposed that they represent the same spectrum of chronic and severe irritant contact dermatitis and therefore may be collected under **more practical terms like "severe diaper dermatitis" [3] or "erosive papulonodular dermatosis" [4].**

Typical history and clinical features are usually sufficient for the diagnosis of Jacquet erosive diaper dermatitis. However, it should be differentiated from other conditions [3, 4], (Table 1). Especially in children, it is crucial to distinguish Jacquet erosive diaper dermatitis from genital condyloma [5], since the implications of sexual abuse will create unnecessary anxiety in the patient and family.

Jacquet erosive diaper dermatitis may pose a therapeutic challenge owing to its chronic and recalcitrant course. Prevention of secondary infections, protecting the diaper area from irritation, and limiting the use of diapers may provide partial relief, but treatment of the underlying disease and

Table 1. *The differential diagnosis of Jacquet erosive diaper dermatitis [3, 4].*

Differential diagnostic condition	Characteristic Features
Granuloma gluteale infantum	Purple-red granulomatous nodules of a few centimeters following chronic candidial infection or prolonged use of fluorinated topical corticosteroids
Perianal pseudoverrucous papules and nodules	Shiny, moist, bright red, flat-topped round papules and nodules of a few millimeters
Intertriginous candidiasis	Bright red plaques on the intergluteal area with satellite papules and pustules at the periphery
Langerhans cell histiocytosis	Persistent diaper rash; greasy, scaly plaques; erosions and ulcerations; coexistence of seborrheic dermatitis-like lesions of the scalp. In systemic involvement: organomegaly, pancytopenia, lymphadenopathy and bone lesions
Acrodermatitis enteropathica	Ecematous, scaly, or bullous lesions on periorificial areas including the perianal region (triad of periorificial dermatitis, alopecia, and diarrhea due to zinc deficiency)
Condylomata lata	Macerated, flat, moist, wart-like papules/plaques of secondary syphilis
Condyloma acuminata	Flat or pedunculated vegetative papules or plaques induced by genital human papilloma virus infection

eradication of the predisposing factors remain the mainstay of therapy [1, 3].

Conclusion

Jacquet erosive diaper dermatitis should be

considered in the differential diagnosis of erosive papulonodular lesions of the diaper area, especially in the setting of chronic diarrhea and diaper usage. Being aware of this entity will help to prevent invasive diagnostic procedures and unnecessary treatments.

References

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