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Jessner lymphocytic infiltration – rare in childhood

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Abstract

We present a 13-year-old girl with Jessner lymphocytic infiltrate of the skin, who has suffered from the disease since the age of 9 years. It is a rare disease in childhood, and we highlight the clinical features and therapeutic response of tacrolimus.

Keywords: Jessner lymphocytic infiltrate

Introduction

Jessner lymphocytic infiltration of the skin (JLIS) is a rare cutaneous condition with unknown etiology, first described by Jessner and Kanof in 1953 [1]. It is a benign disease characterized by single or multiple erythematous papules or plaques on the head, neck, and upper back. The eruption resolves spontaneously after months or a few years [2]. The disease mostly begins in adults and to our knowledge only four other cases in childhood have been described in the literature.

Case Synopsis

A 13-year-old girl was seen with a four-year history of waxing and waning erythematous plaques on both cheeks. The lesions caused pruritus and were cosmetically bothersome (**Figure 1**). She had no photosensitivity. Previous therapy with topical corticosteroids and antihistamines were not effective. Besides a history of atopic dermatitis and asthmatic bronchitis in early childhood, she was otherwise healthy.

On examination, she had reddish, moderately infiltrated plaques with no epidermal changes on her cheeks. Based on her clinical picture, a diagnosis of cutaneous lupus erythematosus was initially considered. A skin punch biopsy showed a dense perivascular and periadnexal lymphocytic infiltration in the superficial and deep dermis with slight mucin deposition between collagen bundles. In addition lymphocytes and few histiocytes infiltrated the interstitial dermis. The lymphocytes were small and mature. Neither neutrophils nor eosinophils were seen. No epidermal changes were found. The histological picture was consistent with JLIS, though the interstitial lymphocytic infiltration in this context was somewhat unusual (**Figure**



Figure 1. Erythematous facial plaques on the cheeks in a 13-year-old girl with Jessner's lymphocytic infiltration.

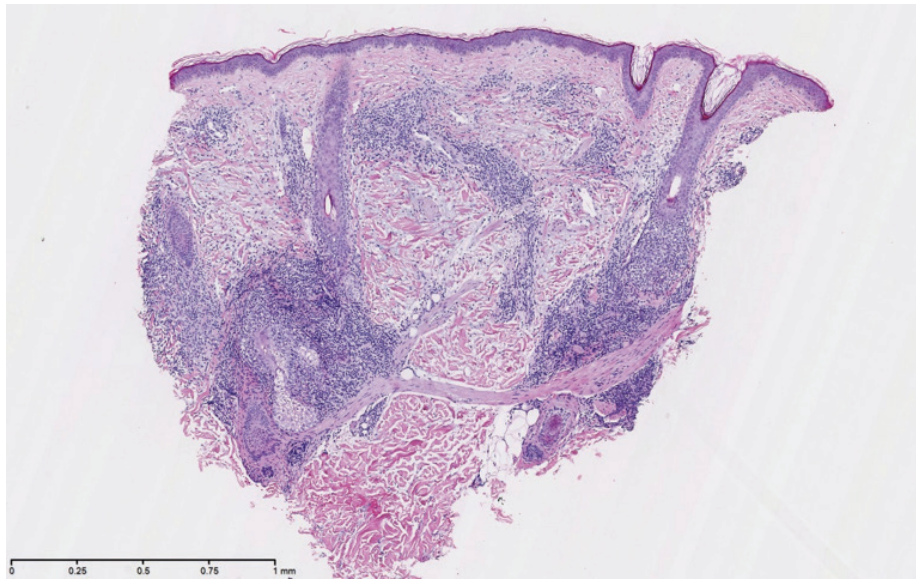


Figure 2. Histology of punch biopsy from the right cheek showing perivascular and periadnexal lymphocytic infiltration and interstitial mucin deposition. H&E, 50 .

2). Immunohistochemical analysis demonstrated lymphocytes positive for pan-T-cell markers admixed with a few B cells. T-cell gene rearrangement analysis showed a polyclonal T cell population of mixed CD4+ and CD8+ subtypes. Full blood cell count, liver, renal, and thyroid function, ANA, SSA/Anti-Ro, and SSB/Anti-La were all negative or normal. She persistently had relapsing erythematous plaques on both cheeks and a reevaluation of her clinical picture, laboratory and histology findings were compatible with JLIS. Treatment with tacrolimus 0.1% daily was prescribed. Six weeks later, her skin had improved significantly and no longer caused pruritus.

Case Discussion

Jessner lymphocytic infiltration of the skin is a rare condition of unknown etiology, characterized by single or multiple erythematous plaques or papules, which are often localized on the head, neck, and upper back. Although JLIS is a photosensitive disorder, there is no correlation with a special season of the year [3]. It has been debated, whether JLIS deserves recognition as a specific disease entity or should be recognized as a variant of cutaneous lupus erythematosus, e.g. lupus erythematosus tumidus (LET), [2]. Erythematous, edematous, non-scarring plaques characterize the clinical picture of LET, which usually starts in the summer months associated with increased sun exposure [4]. The histological features of LET lesions are fairly well described with a dermal lymphocytic infiltrate present in a perivascular and

periadnexal pattern. Abundant interstitial mucin deposition is also detected and dermal edema in the papillary dermis can be observed [5]. Jessner lymphocytic infiltration is histologically characterized by a dense perivascular, predominantly lymphocytic infiltrate with no epidermal changes [6]. It can be very difficult to differentiate LET and JLIS under the microscope. However in this case, prominent lymphocytic infiltration, sparse interstitial mucin, and no dermal edema in the papillary dermis, speak in favour of JLIS [7,8]. The patient was not photosensitive and presented with plaques in the winter months, which also speaks in favour of JLIS.

Jessner lymphocytic infiltration of the skin tends to affect middle-aged patients. Only four other cases of JLIS in childhood have been reported in the literature to our knowledge. These affected two girls and two boys between the age of 4 and 11 years of age, the first case being described in 1988 [9–11]. Usually the skin lesions are asymptomatic, but they can itch or burn. There are no systemic manifestations. Some cases possibly relating to *Borrelia burgdorferi* infection or drugs have been reported [2]. Treatment is difficult, and the response to topical, intralesional or systemic corticosteroids, antimalarials, tetracyclines, penicillin, retinoids, and systemic immunosuppressive drugs is variable. The differential diagnoses include lupus erythematosus, pseudolymphoma, lymphoma cutis, polymorphous light eruption, and cutaneous drug eruption.

Conclusion

We describe the fifth childhood case of JLIS, to our knowledge. The clinical and histological similarities between JLIS and LET may result in diagnostic challenges. However, as our case illustrates there are differences between the two diagnoses. This patient's clinical picture, with no photosensitivity and plaques in the winter months suggests JLIS. In addition, her histology with prominent lymphocytic infiltration, sparse interstitial mucin, and no dermal edema in the papillary dermis speaks in favour of JLIS.

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