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1° INCONTRO 2024 Dermatology Update

Casistica Clinica - Match

U.O.C. di Dermatologia & U.O. Anatomia Patologica Generale Fondazione Policlinico Universitario A. Gemelli, IRCCS Università Cattolica del Sacro Cuore, Roma



Caso # 1





- BMG, 20aa
- No comorbidità
- Febbraio 2024
- Visita ortopedica:
 «lesione distrattiva 3c
 secondo classificazione
 BAMIC ai muscoli flessori
 del ginocchio»
- Prime lesioni comparse dopo l'estate del 2023



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1. La diagnosi corretta è tra le seguenti: micosi fungoide, morfea, eritema anulare centrifugo, tinea corporis?



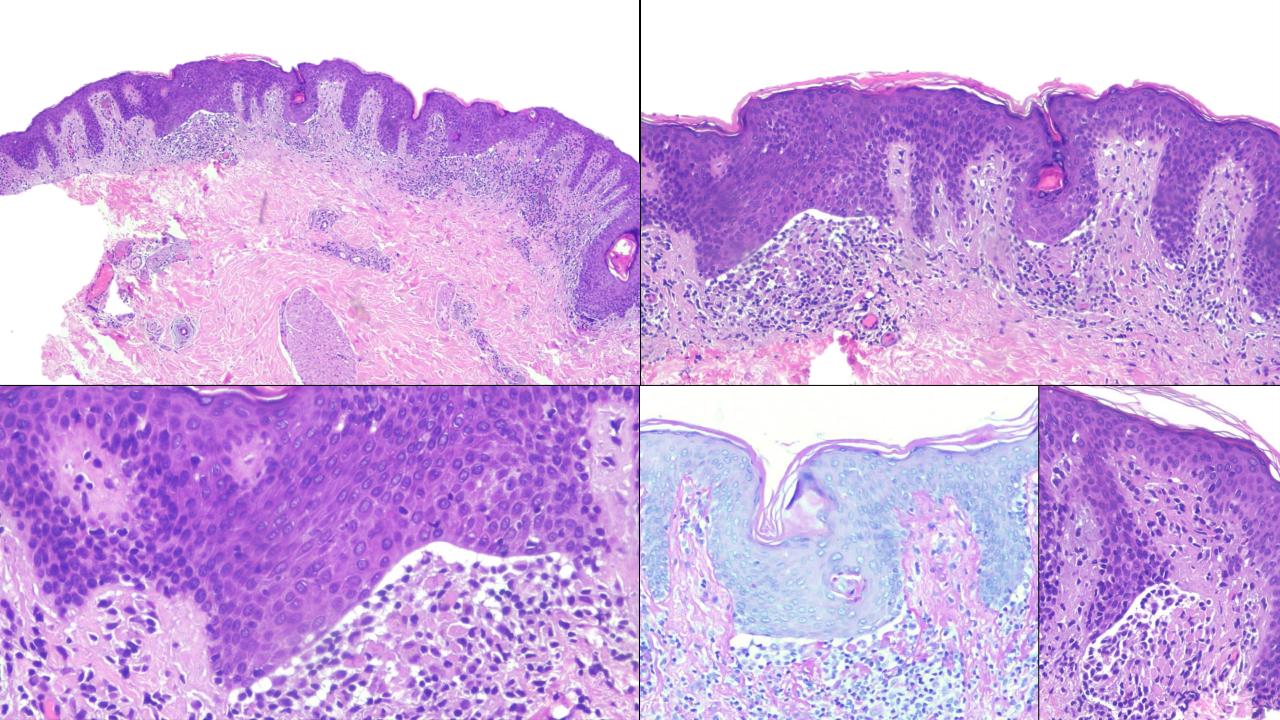
- 1. Sì
- 2. No













Annular lichenoid dermatitis of youth

Giorgio Annessi, MD, Mauro Paradisi, MD, Corrado Angelo, MD, Marie Perez, PhD, Pietro Puddu, MD, and Giampiero Girolomoni, MD Rome, Italy

Background: Lichenoid dermatoses are composed of a wide spectrum of disorders with a common histopathologic interface pattern but diverse causes and pathophysiology.

Objective: We describe a series of young patients with a peculiar annular lichenoid dermatitis, the clinical appearance of which initially suggested diagnoses of morphea, mycosis fungoides, or annular erythema.

Results: The study involved 23 patients (median age 10 years; age range 5-22 years). Lesions consisted of persistent asymptomatic erythematous macules and round annular patches with a red-brownish border and central hypopigmentation, mostly distributed on the groin and flanks. Histology revealed a peculiar lichenoid dermatitis with massive necrosis/apoptosis of the keratinocytes limited to the tips of rete ridges, in the absence of dermal sclerosis and epidermotropism of atypical lymphocytes. The infiltrate was composed mainly of memory CD4⁺ CD30⁻ T cells with few B cells and macrophages. Analysis of T-cell receptor-γ-chain gene rearrangement in skin biopsy specimens revealed polyclonality in all the 15 cases studied. Topical and systemic corticosteroids or phototherapy were effective in most patients with relapse after treatment withdrawal.

Conclusions: We suggest that this is a distinctive inflammatory condition, and we propose to term it "annular lichenoid dermatitis of youth." (J Am Acad Dermatol 2003;49:1029-36.)

ichenoid dermatoses are composed of a wide spectrum of disorders characterized histologically by vacuolar alteration and necrotic/apoptotic keratinocytes in the basal layer of the epidermis together with a bandlike lymphohistiocytic infiltrate obscuring the dermoepidermal junction. These histologic changes are associated with disparate clinical lesions, including erythematous macules, flat-topped violaceous papules, papulovesicles, and plaques that can be arranged in linear or, more rarely, annular pattern.¹

During the last 6 years we have observed a series of young patients with peculiar skin changes consisting of persistent erythematous macules and annular patches mostly localized on the groin and flanks. In all cases the clinical picture was suggestive of inflammatory morphea, patch-/plaque-stage my-

From the Istituto Dermopatico dell'Immacolata, IRCCS. Supported by the Italian Ministry of Health.

Conflicts of interest: None.

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cosis fungoides, or annular erythema. However, all 3 of these diagnoses were excluded histologically with a distinctive superficial lichenoid dermatitis with massive necrosis/apoptosis of the keratinocytes situated at the tips of rete ridges.

In this study we describe the clinical, histologic, immunohistochemical, and molecular characteristics of this condition, which we have termed "annular lichenoid dermatitis of youth" (ALDY), and discuss the differential diagnosis with morphea, patch-/plaque-stage mycosis fungoides, and annular erythemas.

PATIENTS AND METHODS Patient selection

We reviewed the history, clinical photographs, and histologic slides of 23 cases of ALDY, which have been seen during the last 6 years at our institution

Histology and immunohistochemistry

From 23 patients, 32 biopsy specimens were collected for histologic study. In each case 2 hematoxylin and eosin sections were prepared. In all, 2 biopsy specimens from lesions at different stages of evolution were acquired from 6 patients; in another 3 patients, skin samples were obtained from both initial lesions and lesions recurring after 6 to 12

1020

ALDY

- Clinical appearance initially suggested morphea, MF or annular erythema
- 23 patients (median age 10 years; age range 5-22 years)
- persistent asymptomatic erythematous macules and round annular patches with a red-brownish border and central hypopigmentation, mostly distributed on the groin and flanks
- Histology revealed a peculiar lichenoid dermatitis with massive necrosis/apoptosis of the keratinocytes limited to the tips of rete ridges, in the absence of dermal sclerosis and epidermotropism of atypical lymphocytes

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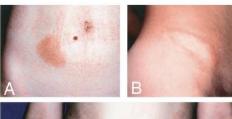
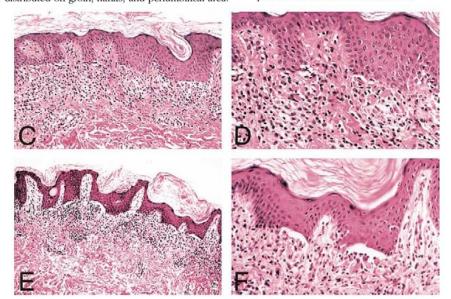




Fig 1. A, Early lesion presenting as round-oval, well-circumscribed erythematous macule with smooth surface. **B**, Annular patch with slightly raised erythematous border and central clearing. **C**, Erythematous macules together with vaguely and overt annular patches are characteristically distributed on groin, flanks, and periumbilical area.

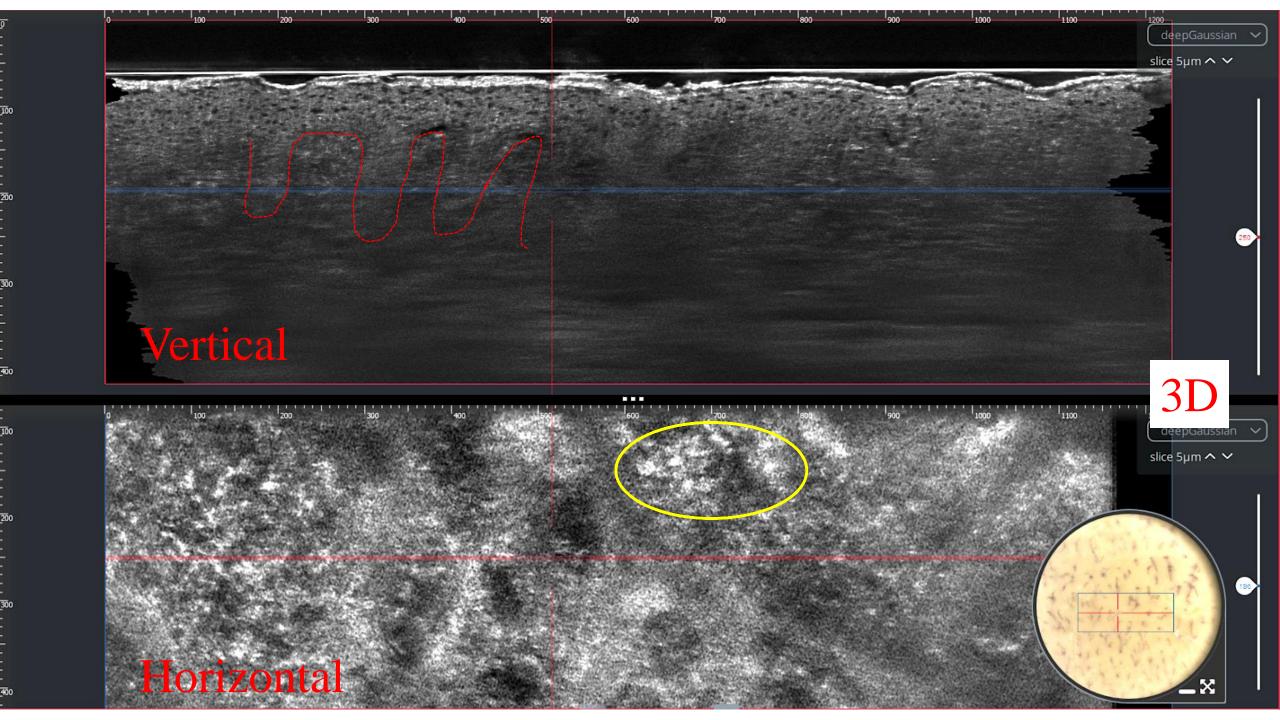


Fig 2. Late lesions appearing as annular patches with brownish nonscaling border delimiting central nonatrophic and nonindurated whitish area.



- Clinical appearance initially suggested morphea, MF or annular erythema
- 23 patients (median age 10 years; age range 5-22 years)
- persistent asymptomatic erythematous macules and round annular patches with a red-brownish border and central hypopigmentation, mostly distributed on the groin and flanks
- Histology revealed a peculiar lichenoid dermatitis with massive necrosis/apoptosis of the keratinocytes limited to the tips of rete ridges, in the absence of dermal sclerosis and epidermotropism of atypical lymphocytes





2. La terapia steroidea è di solito efficace nell'ALDY?

- 1. Sì
- 2. No













lournal of **Cutaneous Pathology**

Annular Lichenoid Dermatitis of Youth ... and Beyond: A Series of 6 Cases

Anna Maria Cesinaro, MD.* Pamela Sighinolfi, MD.* Antonietta Greco, MD.† Lorella Garagnani, BsPhD, * Andrea Conti, MD,† and Fabrizio Fantini, MD†

Abstract: Annular lichenoid dermatitis of youth (ALDY) is a clinicopathologic entity described in children and young patients, clinically reminiscent of morphea, annular erythema, vitiligo or mycosis fungoides. We report on six patients presenting single or multiple lesions, distributed particularly on the flanks and abdomen, with clinical and histologic features consistent with ALDY. Two patients were young girls and four were adults males. Three patients received topical therapy and four showed complete resolution of the lesions after a 24-65 months follow-up. Analogously to the cases reported so far, immunohistochemistry showed a T cell infiltrate with a predominance of CD8+ lymphocytes, while T cell receptor rearrangement was absent in all cases. It seems appropriate to include annular lichenoid dermatitis of youth among the dermatoses with a lichenoid pattern. For the first time, we found that it can affect also adult patients, therefore we propose to rename the disease annular lichenoid dermatitis. The differential diagnosis with mycosis fungoides, especially in adult patients, is particularly crucial for their proper management and treatment.

Key Words: annular lichenoid dermatitis, age, histology, immunohistochemistry, molecular analysis

(Am J Dermatopathol 2009;31:263-267)

INTRODUCTION

Annular lichenoid dermatitis of youth (ALDY) is a clinicopathologic entity described in 2003 by Annessi et al1 in children and young patients. It is characterized by clinical features suggesting inflammatory morphea, mycosis fungoides, vitiligo, or annular erythema, whereas histologically the main differential diagnosis includes mycosis fungoides, particularly the hypopigmented variant.

The differential diagnosis in most cases is not difficult. Clinically, morphea is characterized by indurated plaques and histologically by thickened collagen fascicles and lymphoplasmacytic perivascular inflammatory infiltrate.2 Classic erythema annulare presents as lesions with scaling

erythematous borders, whereas histologically it shows foci of parakeratosis and a superficial or superficial and deep perivascular lymphocytic infiltrate.3 Vitiligo can present as inflammatory lesions mimicking ALDY both clinically and histologically, but unlike to vitiligo, no loss of melanocytes leading to permanent depigmentation has been observed in ALDY.^{1,4} The main problem is the differential diagnosis between ALDY and mycosis fungoides, although histopathology, immunohistochemistry, and molecular analysis seemed to be able to differentiate the 2 diseases.1

MATERIALS AND METHODS

The cases were collected in a 6-year period, from 2003 condition, whereas patient 1 had a sister suffering from atopy. remission of the lesions was observed during and after therapy.

Few more cases have been reported so far, always in young patients.⁴⁻⁷ We describe herein 6 patients with clinicopathologic features consistent with ALDY. For the first time, we found that the disease can develop also in adulthood; the differential diagnosis with mycosis fungoides in these patients is particularly crucial for their proper management

Patients

throughout 2008. Two patients were young girls, aged 10 and 12 years, and 4 were adult males, age ranging from 33 to 45 vears. Lesions consisted of asymptomatic rounded or oval patches, developed by initial erythematous macules with peripheral spreading. Most lesions showed the characteristic annular shape, with a raised erythematous border and a hypopigmented center. In 2 patients, only 1 lesion was present, whereas 4 patients had multiple lesions. In 5 patients, the lesions developed on the trunk, particularly on the flanks and the abdomen; in the last patient, the lesions were localized on the trunk, axillary regions, groin, and neck (Figs. 1A-C). The duration of the lesions before the biopsy ranged between 1 and 15 months. The clinical diagnosis included "eczema," morphea, mycosis fungoides, granuloma annulare, and vitiligo. The patients were otherwise healthy. Testing for autoimmunity, infections, and allergy and collection of relevant clinical information about their families disclosed no putative etiologic factors. Patient 5 revealed an atopic A 1 or 2-month course of topical therapy was given to 3 patients (2 patients with corticosteroids and 1 patient with tacrolimus), whereas the others did not receive any therapy. No Spontaneous resolution was observed at follow-up in 4 of 6 patients after 24-65 months (Fig. 1D). Clinical data are

and review of the literature Annular lichenoid dermatitis (ALDY) is a rare dermatosis that is Viktorvia Kazlouskava¹. Jonathan D.K. Trager² and

Annular lichenoid dermatitis of youth:

a separate entity or on the spectrum

of mycosis fungoides? Case report

most often seen in children and young adults and is characterized by annular patches with raised borders, most frequently on the trunk and the groin. A distinct lichenoid tissue reaction involving the base of the rete, resulting in squared-off rete ridges, helps to differentiate this from other lichenoid dermatoses and mycosis fungoides (MF). Herein, we report an additional case of this condition in a 7-year-boy, whose biopsy exhibited the typical quadrangular rete alteration and also contained distinct aggregates of CD8+ lymphocytes, Langerhans cells and colloid bodies within the involved rete. A literature review with emphasis on the clinical and histopathological differential diagnosis reveals additional clinical features of ALDY to potentially help differentiate this entity from annular presentations of mycosis

Keywords: dermatitis, lichenoid reaction, mycosis fungoides

Kazlouskaya V, Trager JDK, Junkins-Hopkins JM. Annular lichenoid dermatitis of youth: a separate entity or on the spectrum of mycosis fungoides? Case report and review of the literature. I Cutan Pathol 2015; 42: 420-426. © 2015 John Wiley & Sons A/S Published by John Wiley & Sons Ltd

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Annular lichenoid dermatitis of youth (ALDY) is a recently described condition, initially reported in young patients, with a median age of 10 years (range, 5-22 years). The modified designation annular lichenoid dermatitis was later suggested after this entity was described in older patients.² It is not clear whether the condition is rare or under-recognized. It is possible that, given the clinical and histopathologic similarities between ALDY and mycosis fungoides (MF), it may be erroneously diagnosed and managed as MF. Herein, we present an additional case of ALDY and review the literature, with a focus on the possible relationship to MF and inflammatory skin conditions.

Case report

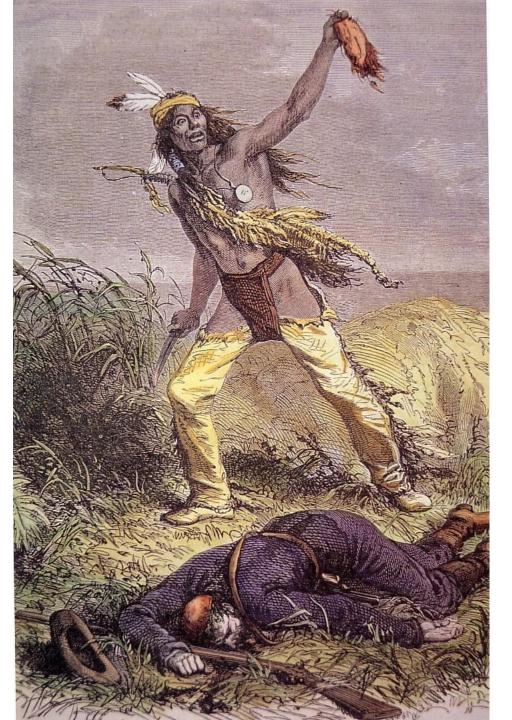
A 7-year-old boy presented to the dermatologist with asymptomatic skin lesions, which had persisted for about 1 year. On examination, there were annular and round, salmon color patches and thin plaques 6-8 cm in diameter, with a slightly raised border. Small pinpoint papules were noted at the periphery. These were located bilaterally in the hips and the left shoulder (Figs. 1 and 2). Central clearing was seen in one

fungoides.

From the *Departments of Pathology; and †Dermatology, Azienda Ospedaliero-Universitaria Policlinico di Modena, University of Modena and Reggio Emilia, Modena, Italy.

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Caso # 2





- DT, 89aa
- Comorbidità: ipertensione, scompenso cardiaco, cardiopatia ischemica, Insufficienza tricuspidale moderatasevera, fibrillazione atriale, portatore di PMK, storia di sepsi da MSSA. Ha subito ricoveri recenti ripetuti per infezione da Clostridium Difficile, a causa della quale ha perso circa 15kg e tuttora in trattamento, e per polmonite con addensamenti polisegmentali nel LSD edema polmonare e versamento pleurico



- DT, 89aa
- Visita Chirurgia Plastica:
- ampia neoformazione cutanea della fronte/cuoio capelluto, di circa 10x6cm compatibile con SCC
- Su pregressa cicatrice di asportazione di lesione cutanea (precedente istologico: cheratosi attinica)
- In considerazione
 dell'età, delle
 comorbidità associate e
 delle terapie
 farmacologiche in corso,
 si ritiene l'intervento
 chirurgico gravato da un
 alto rischio di
 complicanze
- Si richiede valutazione per radioterapia



- DT, 89aa
- Visita Radioterapia:
- Paziente affetto da neoplasia della cute del cuoio capelluto con aspetti compatibili con SCC
- Un trattamento radiante potrebbe essere indicato come trattamento alternativo alla chirurgia previa esecuzione di
- RM cranio e collo con mdc con studio della lesione ed eventuale conferma istologica.





3. La diagnosi corretta è tra le seguenti: SCC, BSC, BCC, carcinoma sebaceo?



- 1. Sì
- 2. No



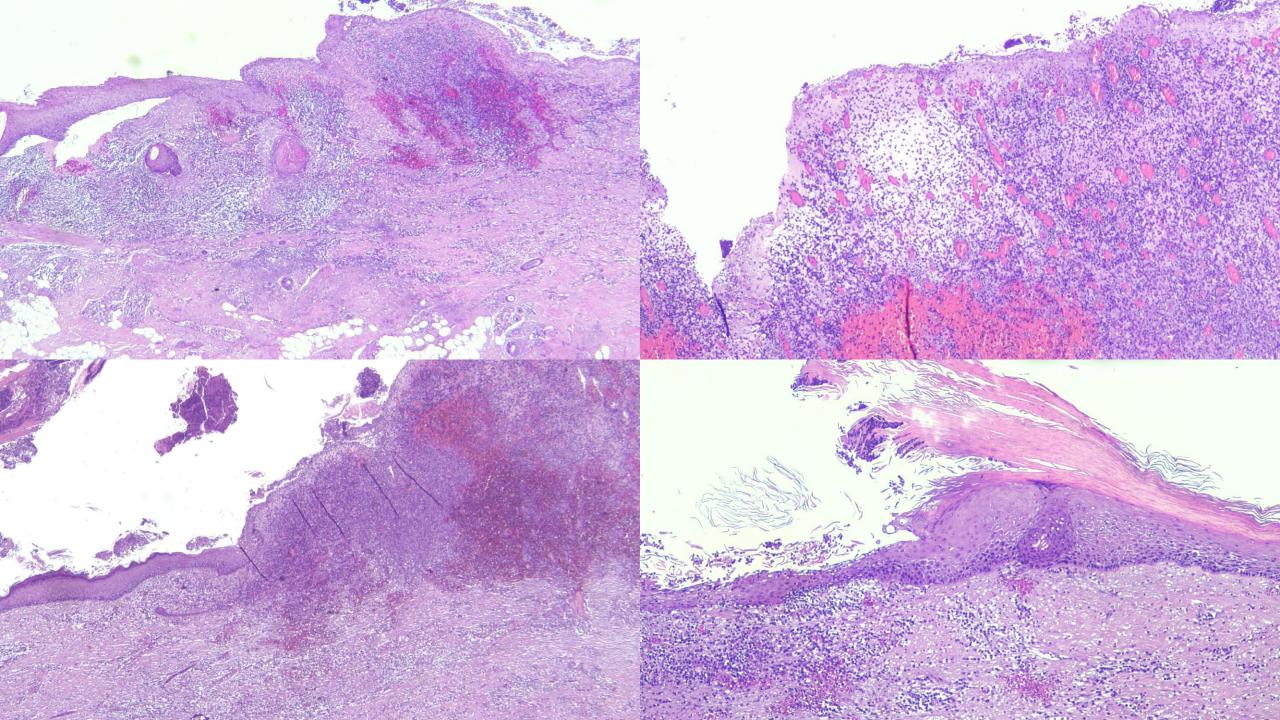


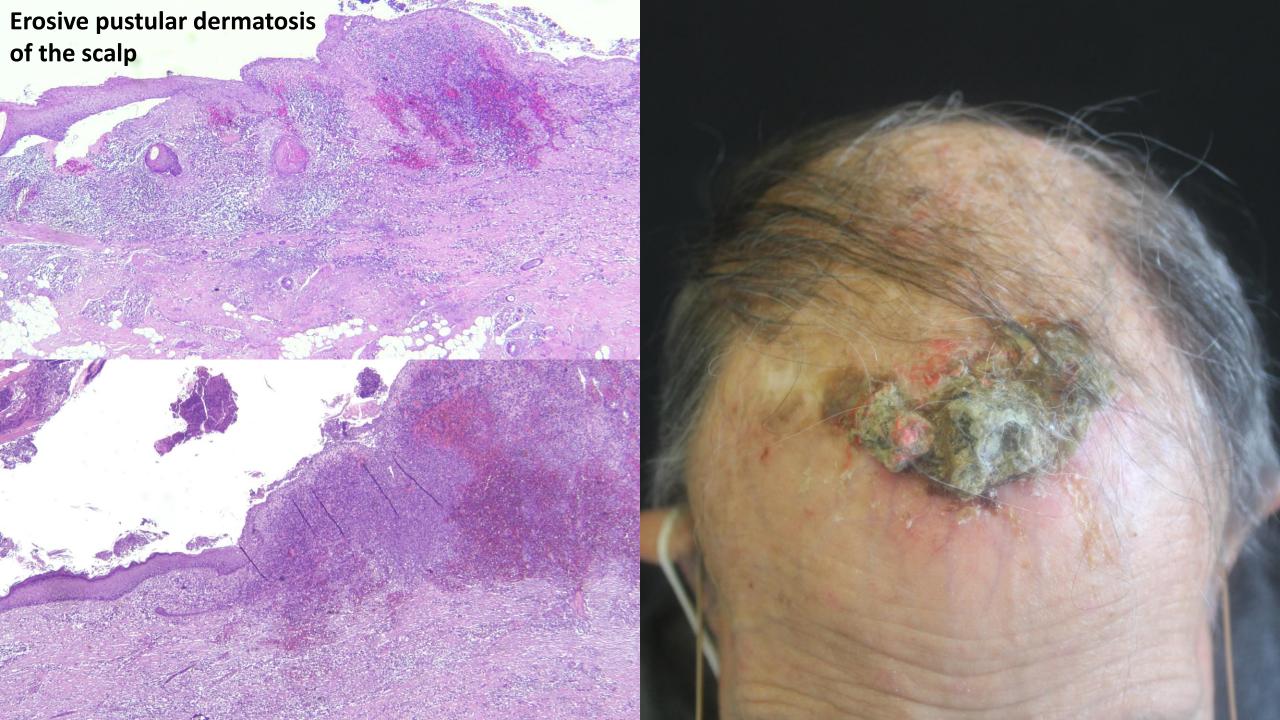












Erosive pustular dermatosis

Christopher Sladden MB BCh, Rebecca Afford BSc

■ Cite as: CMAJ 2019 June 17;191:E667. doi: 10.1503/cmaj.181685

73-year-old man presented to a dermatology clinic with a 10 x 9 cm erosive plaque covered with a thick adherent yellow-brown crust of the scalp (Figure 1). The mass had been enlarging for more than 3 years, without diagnosis or treatment. The patient's history included tinea capitis at age 8 years, treated with radiotherapy (the historical gold standard treatment), and multiple nonmelanoma skin cancers of the head, which had been treated surgically. Removal of the crust revealed a friable, eroded, tender, somewhat vascular plaque. The differential diagnosis included squamous cell carcinoma; other nonmelanoma skin cancers; inflammatory dermatoses, including pemphigus vegetans and Brunsting-Perry type pemphigoid; kerion, pyoderma gangrenosum; and erosive pustular dermatosis. Bacterial skin swabs were negative. Potassium hydroxide and dermatophyte fungal cultures were negative, as was rapid plasma reagin testing. Three punch biopsies from representative areas were taken for histopathologic evaluation, including direct immunofluorescence studies. Histologic findings of fibrosis; chronic mixed inflammation, including plasma cell aggregation; reactive epidermal cytological atypia; serum crust; hyperkeratosis; and parakeratosis were noted on all specimens. There were no findings of malignancy. Histopathology was compatible with erosive pustular dermatosis.

Erosive pustular dermatosis has a striking clinical presentation and may be mistaken for cutaneous malignancy. Clinicopathologic correlation is critical because histopathology is nonspecific; thus, erosive pustular dermatosis is a diagnosis of exclusion supported by a positive response to treatment. It is a noninfectious inflammation of the skin, generally the scalp or legs, associated with chronic ultraviolet damage (in this case, previous radiotherapy may have been involved) and subsequent skin trauma. ²

Systemic antibiotics (to treat any associated infection) and corticosteroids may be required;³ however, in this patient, topical treatment with clobetasol 0.05% and fusidic acid 2% ointments (1:1) used twice daily for 1 month produced excellent improvement (Figure 2). This was followed by hydrocortisone acetate 2.5% and fusidic acid 2% ointments (1:1) applied 3 times daily to complete resolution over the following 2 months.

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Figure 1: Erosive plaque covered with a thick adherent crust of the scalp in a 73-year-old man, consistent with erosive pustular dermatosis.



Figure 2: The patient's scalp after 1 month of treatment with clobetasol 0.05% and fusidic acid 2% ointment applied twice daily. The patient did not follow written instructions for suture removal; however, the sutures serve as a useful marker for re-epithelialization.

Competing interests: None declared.

This article has been peer reviewed.

The authors have obtained patient consent.

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- Uncommon, pustular, idiopathic disorder described in 1979 by Burton and Pye
- typically occurs on the scalp of the elderly
- It presents with variably thickened grey or yellow-brownish crusts, covering shallow, inflamed erosion(s), with scanty, barely detectable, small pustules
- The disease has a chronic and progressive course without spontaneous remission and may involve large scalp regions, ultimately leading to scarring alopecia

4. Il trattamento di scelta è antibiotico?



- 1. Sì
- 2. No











Erosive Pustular Dermatosis of the Scalp: A Clinicopathologic **Study of Fifty Cases**

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Abstract: Erosive pustular dermatosis of the scalp (EPDS) is an uncommon, pustular, idiopathic disgeneris skin disorder.

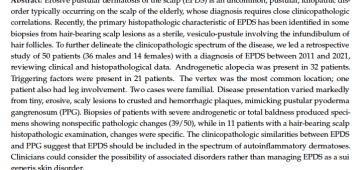
Keywords: erosive pustular dermatosis; histology; pustular spongiotic infundibular folliculitis; scalp; neutrophilic dermatoses; pyoderma gangrenosum; pathergy

Received: 30 August 2021

Erosive pustular dermatosis of the scalp (EPDS) is a rare pustular, idiopathic, inflammatory condition first described in 1979 by Burton and Pye [1]. The disorder typically occurs on the scalp of elderly males and predisposing factors include androgenetic alopecia or sun-damaged skin and/or a history of scalp trauma. However, some cases of EPDS have been reported in younger individuals [2] or even children [3,4]. This dermatosis may also affect other skin sites, including the face [5,6] and extremities [7].

Clinically, this condition is characterized by recurrent small and barely detectable sterile pustules, erosions and variably thickened grey or yellow-brown crusts. The course is chronic, recurrent and progressive, ultimately leading to scarring alopecia [8]. Topical high-potency corticosteroids (clobetasol propionate 0.05% ointment) are the main line of

The true disease incidence is unknown, and it is not clear whether EPDS is a rare entity or, more likely, an underdiagnosed condition. Indeed, underdiagnosis is frequent and may be attributed both to clinical features mimicking other conditions and the purported lack of distinctive histologic findings. Recently, the primary histopathologic characteristic of EPDS has been identified in some biopsies from hair-bearing scalp lesions as a sterile,





updates

Citation: Michelerio, A.; Vassallo, C.

Fiandrino, G.; Tomasini, C.F. Erosive

Pustular Dermatosis of the Scalp: A Clinicopathologic Study of Fifty

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colonization Frequent association with local physical

Cultures are usually negative; when

isolated, microorganisms are a secondary

injuries, mostly accidental mechanical trauma, or inflammation, suggesting a tissue damage-induced immunologic response or immune dysregulation, with the production of a chronic inflammatory reaction

1. Introduction

Erosive pustular dermatosis of the scalp: A neutrophilic folliculitis within the spectrum of neutrophilic dermatoses



A clinicopathologic study of 30 cases

Carlo Tomasini, MD, and Andrea Michelerio, MD *Pavia, Italy*

Background: It is general opinion that histopathology is nonspecific and of little value in diagnosing erosive pustular dermatosis of the scalp (EPDS).

Objectives: Clinicopathologic correlation of erosive pustular dermatosis of the scalp.

Methods: We reviewed the clinical and pathologic records of patients with a clinicopathologic diagnosis of EPDS between 2011 and 2016 at the Dermatopathology Unit of Turin University.

Results: Thirty elderly patients with EPDS were identified (22 men and 8 women). Androgenetic alopecia was present in 19 of 30 patients. Triggering factors included mechanical trauma in 10 of 30 cases, surgical procedures in 4 of 30 cases, and herpes zoster in 1 of 30 cases. Three patients were affected by autoimmune disorders. The vertex was the most common location. Disease presentation varied markedly from tiny, erosive, scaly lesions to crusted and hemorrhagic plaques, mimicking pustular pyoderma gangrenosum. The pathologic changes differed according to lesion type and disease duration. Interestingly, a spongiotic and suppurative infundibulo-folliculitis was observed in 8 of 30 cases.

Limitations: This was a retrospective study.

Conclusions: We believe that the primary lesion of erosive pustular dermatosis of the scalp is a spongiotic, pustular superficial folliculitis. The clinicopathologic similarities with other neutrophilic dermatoses, such as pustular pyoderma gangrenosum, suggest this condition should be included in this spectrum, where pathergy plays a pathogenetic role. (J Am Acad Dermatol 2019;81:527-33.)

Key words: erosive pustular dermatosis; histology; neutrophilic dermatoses; pathergy; pustular spongiotic infundibular folliculitis; pyoderma gangrenosum; scalp.

B rosive pustular dermatosis of the scalp (EPDS) is an uncommon, pustular, idiopathic disorder that was first described in 1979 by Burton and Pye.¹ Since its initial description, about 100 cases have been reported in literature to date.^{2,3} However, it remains to be clarified whether the disease is truly rare or simply underdiagnosed. This disorder

typically occurs on the scalp of the elderly, mostly in women. It presents with variably thickened grey or yellow-brownish crusts, covering ≥ 1 shallow, inflamed erosion(s), with scanty, barely detectable, small pustules.

Although historically referred to as scalp of the elderly, this dermatosis may occasionally occur in

- histopathology is nonspecific and of little value in diagnosing EPDS
- It is a neutrophilic superficial folliculitis, with some clinicopathologic similarities with other pathergic neutrophilic dermatoses, such as pyoderma gangrenosum
- Above all a biopsy is often needed to rule out other conditions







EPDS may be mistaken for cutaneous malignancy



is it truly rare or simply underdiagnosed?!?...



Caso # 3





- EC, 32aa
- Comorbidità: -
- Familiarità per carcinoma cervice uterina
- Recente ingrandimento di una macchia pre-esistente a livello del piccolo labbro di destra
- al rientro dopo una vacanza al mare
- Visita Ginecologia Oncologica:
- Escludere neoplasia cutanea





5. La diagnosi corretta è tra le seguenti: BCC, pemfigoide delle mucose, Paget extramammario, angiocheratoma diffuso di Fordyce?

1. Sì

2. No



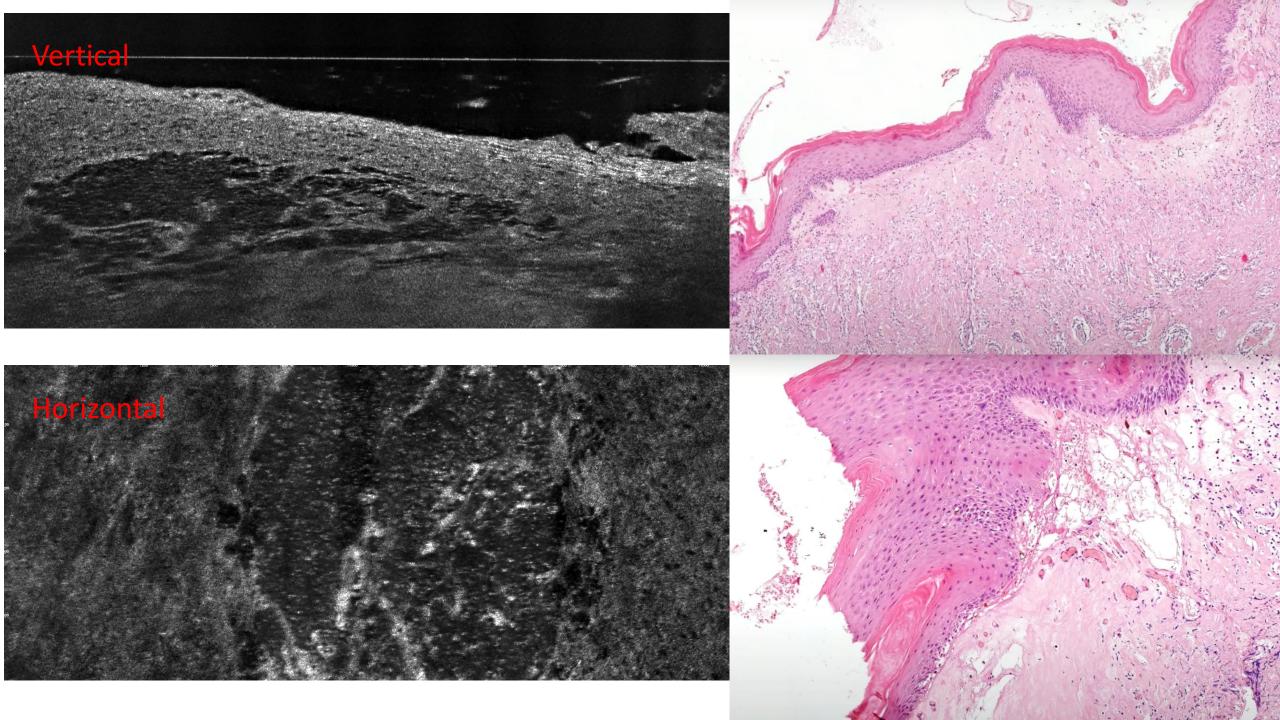












Dermatology Online Journal | Letter Volume 24 Number 2 | February 2018 24(2): 14

Bullous lichen sclerosus: isolated vulvar involvement

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Abstract

We present a patient with the bullous form of lichen sclerosus of the vulva. She had no lesions in other cutaneous and mucosal areas. We used topical tacrolimus and topical clobetasol propionate. The patient was lesion free at the first-year follow-up.

Keywords: bullous, lichen sclerosus, vulva

Introduction

The bullous type is a rare variant of lichen sclerosus

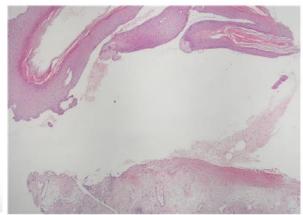


Figure 2. Subepidermal vesiculation with blister formation,. H&E,



Figure 1. Hemorrhagic bulla $(\mathbf{1})$, vesicle $(\mathbf{1})$, erosions and ivorywhite plaques.

TSH was $5.94\,\mu$ lU/mL (normal range 0.27-4.2). Anti TG was $382.5\,$ IU/ml (normal range <115). Bacterial culture of bulla was negative.

Open Access Case Report

Dermal Hemorrhage: A Clue to Lichen Sclerosus et Atrophicus

DOI: 10.7759/cureus.9343

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Abstract

Lichen sclerosus et atrophicus (LSA) may present in a rare bullous and hemorrhagic form that is often difficult to recognize both clinically and histopathologically. Clinically, the lesions may be characterized by atrophic and ivory-white sclerotic plaques in both genital and extragenital regions. Histologically, fully developed lesions of LSA are characterized by a thinned, effaced epidermis with interface change, a wide band of hyalinization in the upper dermis, and a lymphohisticytic infiltrate below the hyalinized area. Extensive vacuolar degeneration weakens the integrity of the dermoepidermal junction, which contributes to the development of marked edema in the papillary dermis and subepidermal vesiculation. With increased fragility of dermal capillaries, hemorrhage can accumulate within the bullae. Recognizing prominent upper dermal hemorrhage as a secondary change may lead to a prompt diagnosis of LSA. We present a case of



FIGURE 1: Clinical image of right medial breast demonstrating a welldemarcated fragile hemorrhagic plaque with thin cigarette paper-like texture, with surrounding atrophic skin.

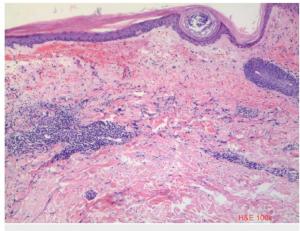


FIGURE 2: H&E ×100 histology demonstrating hyperkeratosis, epidermal atrophy, papillary dermal edema, and band-like lymphocytic infiltrates directly below. Notice the subepidermal vesiculation and homogenization of the papillary dermis surrounding the hemorrhage.

6. Si può osservare anche nella morfea, configurando il quadro della c.d. morfea bollosa emorragica?



- 1. Sì
- 2. No



ROMA 17-18 Maggio 2024









CASE AND RESEARCH LETTERS

Bullous Morphea: Description of a New Case and Discussion of Etiologic and Pathogenic Factors in Bulla Formation[☆]



Un nuevo caso de morfea ampollosa y discusión de los factores etiopatogénicos en la formación de las ampollas

To the Editor:

Bullous morphea is a rare form of localized scleroderma. Since the description of the first case, numerous theories have been put forward on the origin of the blisters, but the actual mechanism by which they develop remains unclear.

Our patient, a 63-year-old woman, presented 2 lesions that had arisen some months earlier, localized symmetrically in the pretibial regions (Fig. 1). The lesions had an atrophic appearance, an erythematous base, and a central blister that contained a blood-stained fluid. The patient described no history of trauma.

Biopsy showed subepidermal separation with preservation of the basal layer. A mild inflammatory component and collagen fibrils were observed in the superficial dermis, but extensive sclerosis with a dense lymphocytic infiltrate was found in the reticular dermis and subcutaneous cellular tissue, though with no vasculitis or significant presence of eosinophils (Fig. 2). No lymphangiectasia was observed after staining with hematoxylin-eosin or D2-40 (Fig. 3). The findings were compatible with a diagnosis of bullous morphea. Blood tests were within normal limits and *Borrelia* serology was negative. Topical therapy with betamethasone and fusidic acid was started, leading to resolution of the blisters, but the sclerotic plaques deteriorated and systemic treatment was therefore commenced with oral methotrexate, achieving a good response.

Bullous morphea is a rare form of localized scleroderma. ¹ The first case was described by Morrow in 1896. ² Since that time, numerous theories on its origin have been proposed. ³⁻⁵

The most widely accepted theory on the origin of the blisters



75

Figure 1 Bilateral atrophic plaques with overlying blisters.

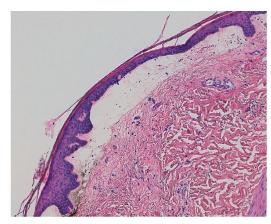
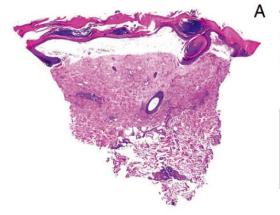


Figure 2 Subepidermal blister with preservation of the basal layer. Hematoxylin and eosin, original magnification $\times 20$.

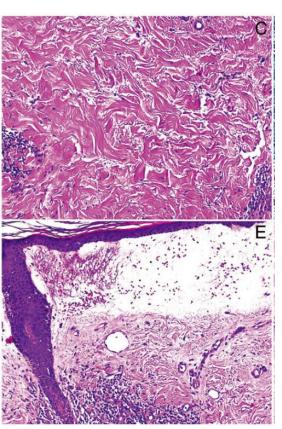


Three cases of bullous morphea: histopathologic findings with implications regarding pathogenesis

Bullous morphea is a rare variant and is not frequently reported. We present three cases of bullous morphea. Although lymphangiectases have been suggested as the most likely mechanism for the development of the bullae in cases of morphea, none of the cases presented with lymphangiectases. To the contrary, all of our cases showed hemorrhagic content in the bullae, which suggests local trauma as a mechanism involved in bulla formation.

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7. Brutti i casi?

1. Sì

2. No









