# A teen with blaschkolinear tumid lupus erythematosus

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## INTRODUCTION

Tumid lupus erythematosus is a subtype of cutaneous lupus erythematosus described by Hoffmann in 1909.[1] Unlike other forms of cutaneous lupus erythematosus, tumid lupus erythematosus lacks systemic involvement and often has negative autoimmune workup. Antinuclear antibody may be positive in about 10% of patients.[2] The classic clinical presentation is characterized by erythematous, succulent, urticaria-like, nonscarring plaques most commonly affecting photosensitive areas. Tumid lupus erythematosus affects mostly adults. There are very few pediatric cases reported; most pediatric cases present on the face and/or upper portion of the body, and none involve the lower extremities. A unilateral blaschkolinear distribution is rare and has only been found in 3 adults and 1 child. We report an additional pediatric case of unilateral blaschkolinear tumid lupus erythematosus with a unique involvement of the lower extremity.

*Abbreviations used:*

C3: complement 3
C4: complement 4
IgA: immunoglobulin A
IgG: immunoglobulin G

## CASE REPORT

A 15-year-old girl presented to the dermatology clinic with a 2-year history of progressive erythematous, edematous papules and plaques in a linear pattern involving the left side of her body. Her previous dermatologist suspected scleredema and referred her to our tertiary care center for further evaluation. She had a history of congenital amniotic band syndrome with multiple digit deformities. She had no family history of autoimmune disease and denied having joint pain and other systemic symptoms. On physical examination, there were violaceous dermal papules coalescing into blaschkolinear plaques extending from the upper portion of the left medial arm to the wrist (Fig 1, *A*), from the left side of the abdomen to the flank (Fig 1, *C*), and from the left posterior thigh to the lower portion of the leg (Fig 1, *E*). Further laboratory workup for autoimmune disease was notable for antinuclear antibody (1:320, nuclear, homogenous) and otherwise all negative for $\alpha_1$-, $\alpha_2$-globulins, antineutrophilic cytoplasmic antibody, antistreptolysin O, $\beta_1$-, $\beta_2$-globulin, cardiolipin (IgA, IgG, immunoglobulin M), centromere B, double-stranded DNA, $\gamma$-globulin, topoisomerase 1, Sjögren's-syndrome-related antigen A, Sjögren's-syndrome-related antigen B, Smith (Sm)/U1-RNP, rheumatoid factor, and C-reactive protein. C3 and C4 were within normal limits. Punch biopsy of the lesions revealed focal vacuolar interface change and mild superficial and deep perivascular and periadnexal lymphoplasmacytic inflammation (Fig 2, *A-C*). Alcian blue and colloidal iron staining showed increased superficial and deep interstitial mucin deposition (Fig 2, *D*). The histologic findings supported the diagnosis of tumid lupus erythematosus. The patient was initially started on hydroxychloroquine (200 mg daily), which was discontinued due to diarrhea. Topical treatment with mometasone (0.1% cream) and tacrolimus (0.1% ointment) for 1 year did not lead to significant improvement. The patient subsequently initiated treatment with oral dapsone

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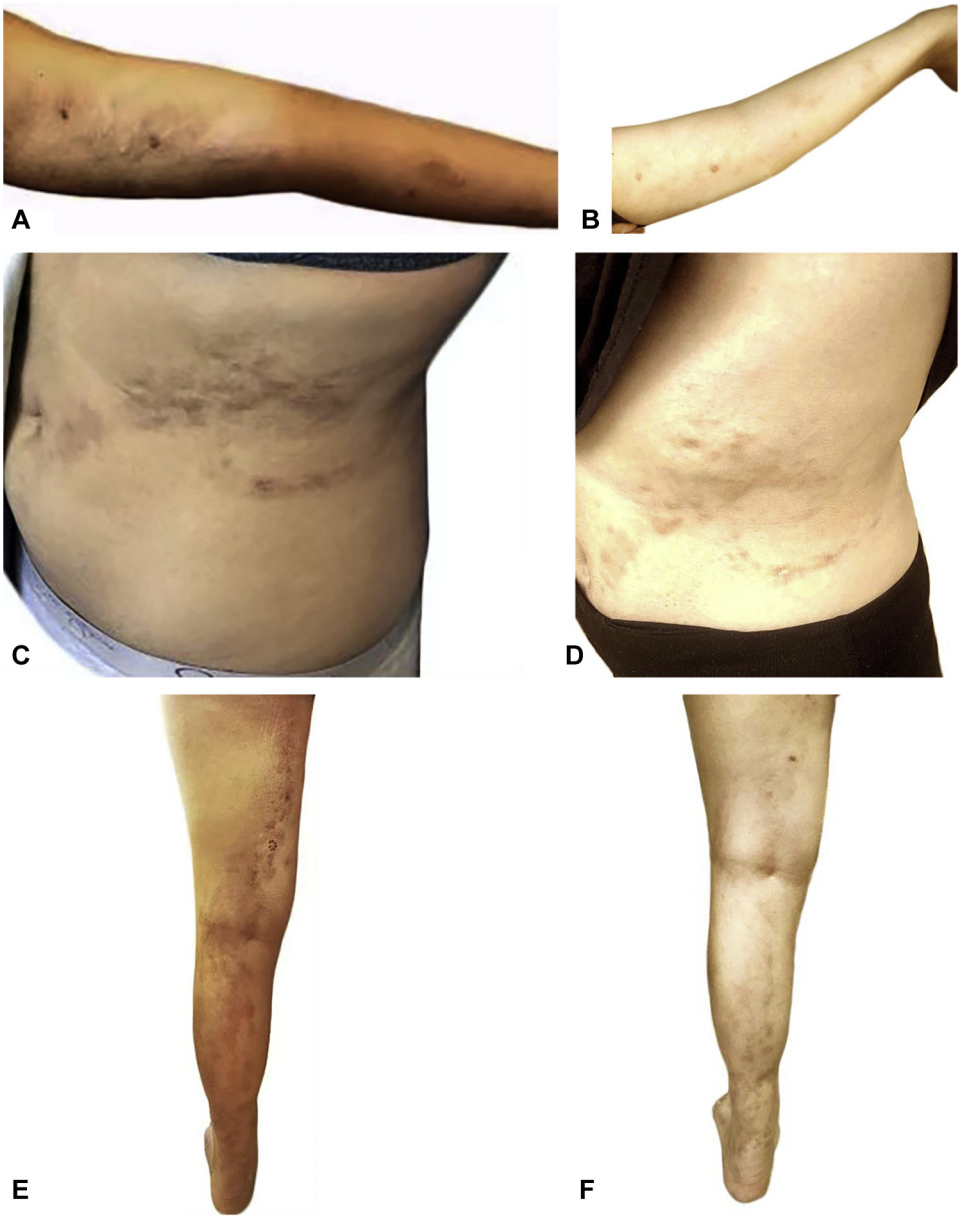


**Fig 1.** Baseline and posttreatment clinic photographs of tumid lupus erythematosus. **A,** Violaceous and skin-colored dermal papules coalescing into a blaschkolinear plaque on the upper portion of the left medial arm extending to the left wrist. **B,** Improvement of skin lesions on the left arm after treatment. **C,** Violaceous and skin-colored dermal papules coalescing into a blaschkolinear plaque on the left side of the abdomen extending to the left flank with midline demarcation. **D,** Improvement of skin lesions on the left side of the abdomen after treatment. **E,** Violaceous and skin-colored dermal papules coalescing into a blaschkolinear plaque on the left posterior thigh to the calf and foot. **F,** Improvement of skin lesions on the left leg after treatment.

(25 mg) daily for 1 week followed by 50 mg daily and noted striking improvement after 5 months of treatment.

## DISCUSSION

The diagnosis of tumid lupus erythematosus is based on clinical presentations confirmed by histopathology. This case demonstrated classic histologic features of tumid lupus erythematosus, including superficial and deep perivascular and periadnexal lymphocytic inflammation, increased interstitial mucin, subtle interface dermatitis, and a notable lack of basement membrane thickening and epidermal change, as would be expected

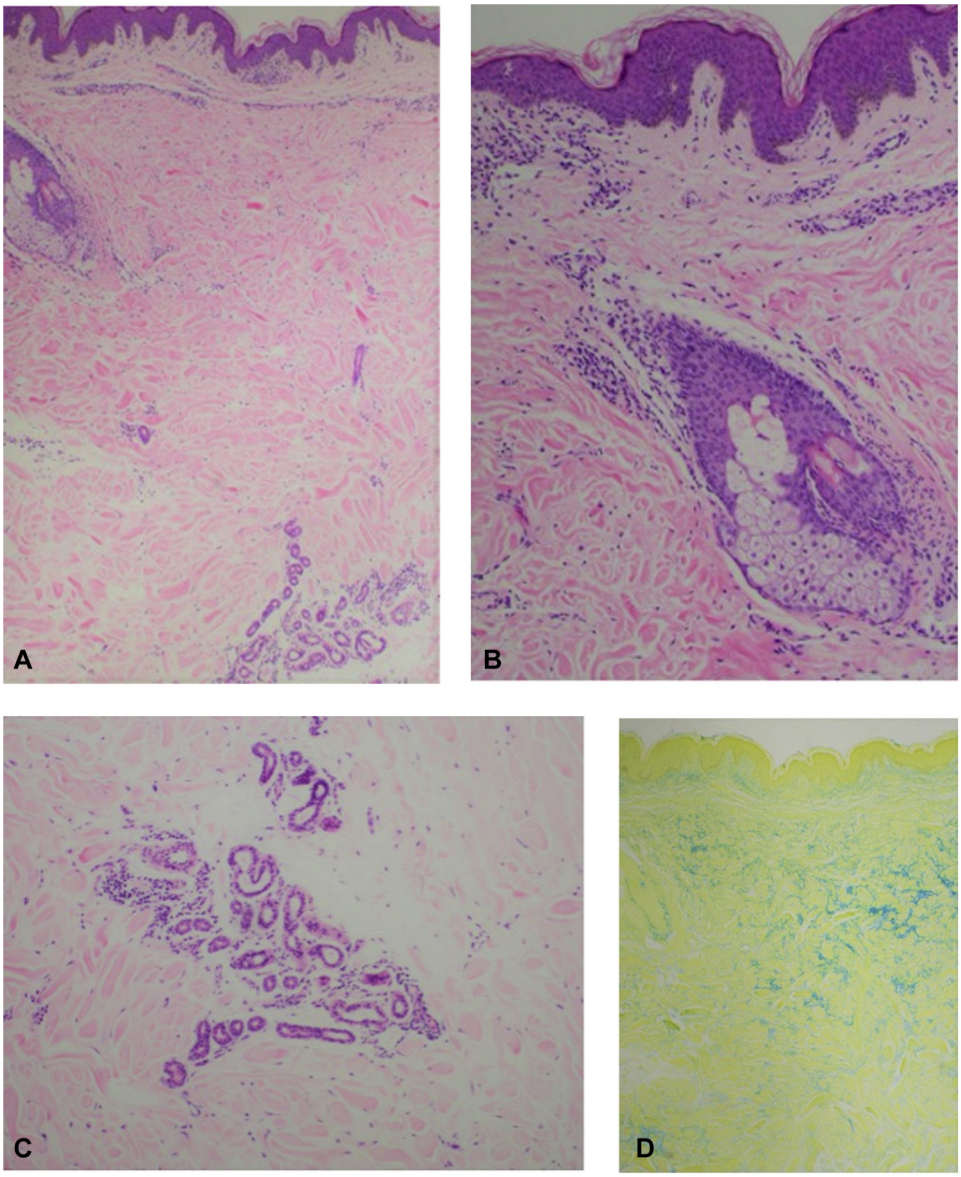


**Fig 2.** Histopathologic images of biopsy specimen from the left posterior thigh. **A**, Mild interface dermatitis and superficial and deep perivascular and periadnexal inflammation. (hematoxylin and eosin stain; original magnification: ×40.) **B** and **C,** Vacuolar interface dermatitis with superficial and deep periadnexal lymphoplasmocytic inflammation. (hematoxylin and eosin stain; original magnification: ×100.) **D,** Increased superficial and deep interstitial mucin. (Colloidal iron stain; original magnification: ×40.)

in other forms of cutaneous lupus. A broad differential diagnosis should include other entities with dermal mucin deposition, in particular morphea, scleredema, and reactive granulomatous dermatitis. Linear morphea is more common in the pediatric population and in the early phase may show a mild increase in dermal mucin. This patient presented with a 2-year history of symptoms, and morphea would likely have progressed to indurated plaques with homogenized, dense collagen entrapping adnexal structures. Scleredema favors the upper portion of the torso and histologically is characterized by prominent mucin deposition between thickened collagen bundles with only sparse inflammation, and therefore it was not favored. Reactive granulomatous dermatitis may present as linear, cord-like bands, but biopsy would demonstrate interstitial and palisaded histiocytes and neutrophils as opposed to the periadnexal lymphocytes seen in this case.[3] In the pediatric population, lichen striatus could also be considered in the differential diagnosis of a blaschkolinear eruption and may

**Table I.** Literature review of blaschkolinear tumid lupus erythematosus cases

| Age group | Age (yr) | Sex | Morphology and chronicity of lesions at initial presentation | Distribution | Treatment(s) with therapeutic effect | Attempted treatment(s) without improvement | Ref. |
|---|---|---|---|---|---|---|---|
| Child | 4 | F | Wheal-like erythematous to brownish plaque in linear pattern for 1 yr | Right upper portion of the back, upper portion of the right chest, right arm and hand | Hydroxychloroquine 50 mg daily (2 mg/kg/day) in addition to intralesional steroid injections and topical steroids | Topical tacrolimus; topical steroids and intralesional triamcinolone injection (2.5 mg/mL) without hydroxychloroquine | Kim et al[8] |
| | 15 | F | Erythematous, edematous papules and plaques in a linear pattern for 2 yr | Left arm, left side of trunk, left leg | Oral dapsone 25 mg daily for 1 wk followed by 50 mg daily | Hydroxychloroquine 200 mg daily (stopped due to side effect); topical mometasone 0.1% cream; topical tacrolimus 0.1% ointment | Our case |
| Adult | 32 | F | Linear papular and erythematous eruption for 3 yr | Forehead | Topical steroids (dexamethasone once a day) and hydroxychloroquine 200 mg daily. Complete resolution achieved after 2 mo* | None reported | Bouzit et al[10] |
| | 27 | M | Erythematous papules and plaques for 3 yr | Right arm, right posterior trunk | Intralesional corticosteroids and oral hydroxychloroquine 400 mg daily | None reported | Pacheco et al[2] |
| | 43 | M | Erythematous papules and plaques for 1 yr | Right side of the forehead to right eyelid | Systemic corticosteroids and hydroxychloroquine 200 mg daily | None reported | Hinz et al[9] |

*mo,* Month; *Ref,* references; *yr,* years.
*The only case reporting complete resolution. All other cases noted marked improvement but did not document complete resolution.

resemble tumid lupus histologically with superficial and deep periadnexal chronic inflammation. However, lichen striatus is composed of numerous coalescing flat-topped scaly papules, and there is a prominent lichenoid infiltrate at the dermoepidermal junction with numerous dyskeratotic keratinocytes and no increase in dermal mucin. Thus, blaschkolinear tumid lupus erythematosus was felt to best characterize this patient's presentation.

At the present time, very few pediatric cases of tumid lupus erythematosus have been reported.[4-8] Most previous pediatric cases involved the face and/or upper portion of the body with variable photosensitivity, and none involved the lower extremities. A blaschkolinear distribution has previously been reported in 3 adult cases[2,9,10] and one pediatric case.[8] In children, the blaschkolinear distribution is more commonly reported in discoid cutaneous lupus erythematosus and lupus panniculitis.[11] In addition, the unique involvement of the lower portion of the body found in our patient shows that tumid lupus erythematosus in children can have a more extensive body involvement than previously reported.

Tumid lupus erythematosus in adults is generally treated similarly to other subtypes of cutaneous lupus erythematosus. First-line therapies are topical or intralesional corticosteroids, topical calcineurin inhibitors, and/or oral antimalarial drugs. Reported second-line therapies include dapsone, methotrexate, acitretin, mycophenolate mofetil, thalidomide, and rituximab.[12] The treatment approach for the blaschkolinear distribution is the same, but it is worth noting that none of the reported blaschkolinear cases was controlled by topical treatment alone. All previous cases were treated with oral antimalarial drugs,

regardless of the patient's age or chronicity of symptoms (Table I). Most pediatric cases of tumid lupus erythematosus require oral antimalarials in addition to or in place of topical treatment (Supplementary Table 1, available via Mendeley at https://data.mendeley.com/datasets/xgwy7bfrrc/1). Our case demonstrates the potential utility of dapsone for the scenario in which antimalarial therapy is poorly tolerated.

The unilateral blaschkolinear distribution is suggestive of underlying mosaicism on the genetic or epigenetic level, leading to the strikingly sharp demarcations of disease along presumed developmental lines. This model would provide the basis for discovery of pathogenic genes in tumid lupus erythematosus (ie, by genome-wide comparison of affected vs normal tissue), as well as future targeted therapy.

**Conflicts of interest**

None disclosed.

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