

## Pediatric Tumid Lupus Erythematosus: Two Case Reports

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### ABSTRACT

Tumid lupus erythematosus (TLE) is a rare variant of cutaneous lupus erythematosus. The clinical diagnosis can be difficult to ascertain as it often presents with negative serology and without associated systemic symptoms. We present two pediatric cases of TLE: a young girl with erythematous facial papules and a second with purple and grey facial plaques. Biopsies displayed a lymphocytic infiltrate consistent with TLE and both improved on antimalarials. These cases highlight the varied morphologies and bring greater awareness of TLE and its treatments.

**Key words:** Pediatric; Tumid; Lupus; Cutaneous; Erythematosus; TLE

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### INTRODUCTION

Tumid lupus erythematosus (TLE) is a rare variant of chronic cutaneous lupus erythematosus, typically with a favorable prognosis. The clinical diagnosis can be difficult since TLE often presents without associated systemic systems and the lesions are not characteristic of those found in other forms of cutaneous lupus<sup>[1]</sup>. Pediatric cases can be more challenging to diagnose as the condition is exceedingly rare in this population with only a few reported cases in the literature<sup>[2]</sup>. A biopsy is often necessary to definitely diagnose TLE.

We present two pediatric cases of TLE: a 6-year-old and an 11-year-old girl with multiple erythematous and edematous plaques on the face. In both cases, a biopsy was performed in order to confirm the diagnosis of TLE. Both were started on hydroxychloroquine with positive response to treatment.

### CASE REPORTS

#### Patient 1

A 6-year-old girl presented to our pediatric dermatology clinic with three well-demarcated and indurated pink plaques on the face and one on the left ear present for two months duration (Figure 1). As per her parents' description, these lesions started as flat pink macules and patches on the forehead, nose, and right cheek that became gradually raised over time. Prior to her presentation at our clinic, she had been given and failed treatment with a topical steroid, topical antibiotic, and an oral antifungal for presumed bacterial and fungal infections respectively. Bacterial culture of the nares and fungal scraping of the skin were both negative. Laboratory work-up including antinuclear antibody (ANA) was unremarkable. A biopsy of the plaque on her left ear revealed a lichenoid, superficial and deep perivascular lymphocytic infiltrate consistent with cutaneous lupus erythematosus (Figure 3). After evaluation and clearance from a pediatric ophthalmologist, the patient was started on a weight-based dose of hydroxychloroquine 100mg daily, topical mometasone, and instructed to wear sunscreen daily along with sun protective clothing and hats. Within four weeks, all the lesions resolved with only minimal erythema remaining on the nose and barely noticeable post-inflammatory hyperpigmentation (Figure 2). Mometasone was discontinued, and she was maintained on daily hydroxychloroquine which she continued to tolerate without any side effects.

**Patient 2**

A 11-year-old female presented with multiple purple and grey dermal plaques on the cheeks, malar area, eyebrows, and postauricular regions present for eight months (Figure 4). This started in conjunction with a non-productive cough and a history of exposure to a sibling with a positive PPD test. A diagnosis of cutaneous tuberculosis (lupus vulgaris) was suspected. Laboratory evaluations yielded a urine culture that was negative for tuberculosis organisms but a positive ANA serology with titers of 1:1280. A biopsy of a lesion behind the left ear exhibited superficial and deep periadnexal lymphocytic infiltration with a hint of mucin deposition consistent with tumid lupus erythematosus (Figure 5). After evaluation and clearance from an ophthalmologist, the patient was started on a weight-based course of hydroxychloroquine 200mg daily and instructed to apply sunscreen regularly. On two week follow-up, the lesions appeared to be improving and she denied any medication side effects.

Continuing her regimen, she presented two months later with scattered erythematous papules and hyperpigmentation following a trip to Peru. Given the acute symptoms, her hydroxychloroquine was increased to 400mg every other day alternating with 200mg on the subsequent days for a duration of two months. On follow-up, she displayed improvement with resolving lesions and hydroxychloroquine was tapered back down to a daily dose of 200mg. ANA serum titers at that time had decreased to 1:640 and after another two months at 200mg daily, the patient began a 10-month slow taper off the hydroxychloroquine. Once the taper was completed, she was instructed to take daily hydroxychloroquine only during the spring and summer months as prophylaxis which she continued to do for two years and then stopped. Since then, she has maintained strict sun protection and has remained free of any TLE-associated lesions and repeat ANA serology testing has been within normal limits.

**COMMENT**

TLE is a chronic form of cutaneous lupus erythematosus. Clinically, patients develop erythematous, edematous, non-scarring, violaceous plaques on sun-exposed skin. Patients can have solitary or multiple lesions, and symptoms can develop rapidly or progress slowly over weeks to months. Cutaneous findings lack surface changes such as scaling, atrophy, scarring, follicular plugging, or telangiectasias that one may see with other forms of cutaneous lupus such as discoid lupus erythematosus<sup>[3]</sup>. Kuhn et al. conducted a case study of forty adult patients and long-established that patients exhibited extreme photosensitivity and had marked improvement with antimalarials and in some cases topical steroids. Histological changes such as perivascular and periadnexal lymphocytic infiltration and interstitial mucin deposition are diagnostic<sup>[4]</sup>.

Given the rarity of TLE in children and the spectrum of possible morphological presentations, diagnosis can be challenging. While cutaneous lupus was highly suspected on first dermatologic examination of our 6-year-old patient, other more common pathologies for her age group such as fungal and bacterial infections needed to be ruled-out. Only after her tests were negative and she had failed adjunctive treatments was a biopsy performed in order to confirm the diagnosis. The 11-year-old patient was first diagnosed with lupus vulgaris based on her clinical history and exposure to a positive PPD sibling. However, negative urine tests and a positive ANA led to a differential that included TLE which was confirmed on biopsy. These two cases demonstrate the challenges in recognizing and diagnosing TLE.

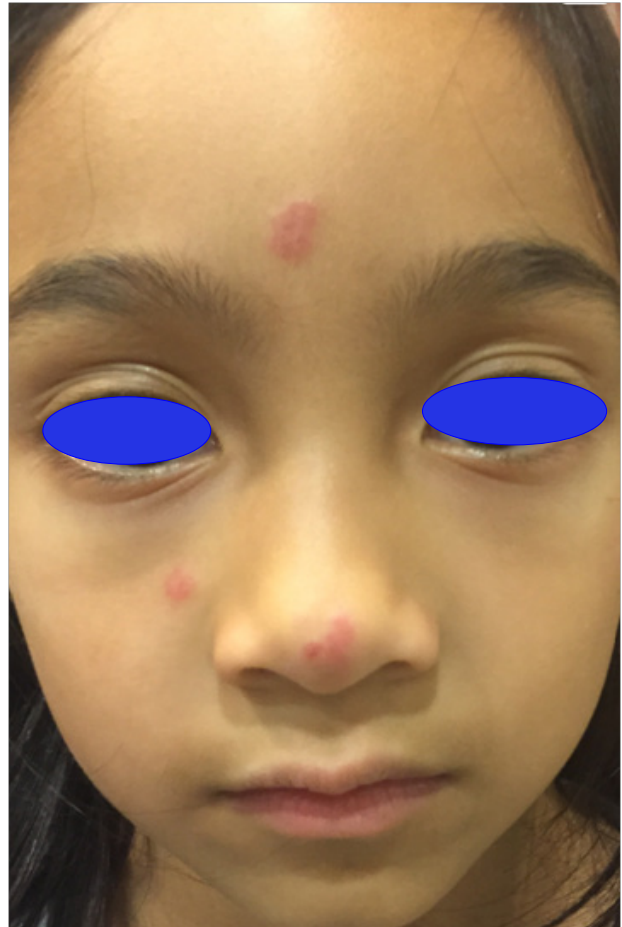


Figure 1 Patient #1, Well-demarcated, annular pink papules on the forehead, tip of nose, and right cheek.

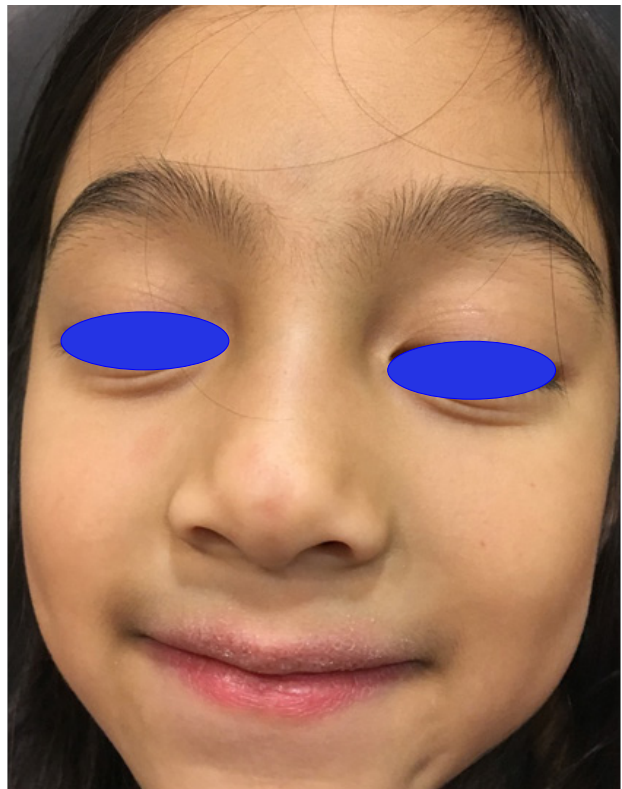
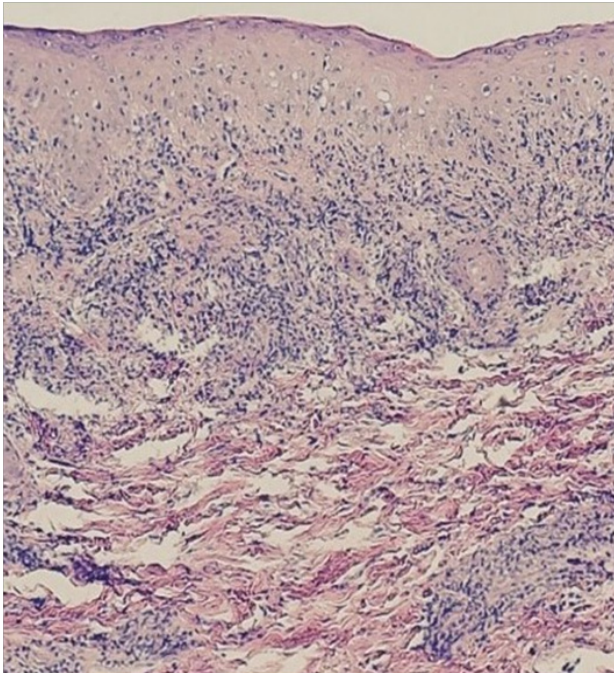


Figure 2 Patient #1, Minimal erythema on the tip of the nose and mild post-inflammatory hyperpigmentation on the forehead and right cheek that was barely noticeable after four weeks of treatment.

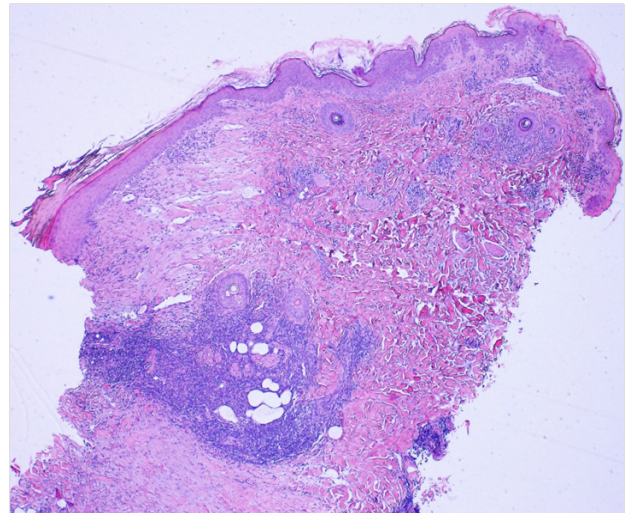


**Figure 3** Patient #1, Biopsy of lesion on left ear. Histology exhibits an interface pattern with lichenoid superficial and deep perivascular and periadnexal lymphocytic infiltrate consistent with cutaneous lupus erythematosus [hematoxylin and eosin stain, magnification x200].



**Figure 4** Patient #2, Multiple purple and grey dermal plaques on the cheeks, malar area, and eyebrows.

Treatment efficacy can vary among patients. In adults, first line treatment options include antimalarials, sun protection and avoidance, and topical steroids. While pulsed dye laser has been used successfully to treat active lesions, it has not been shown to prevent future recurrences<sup>[5]</sup>. Topical tacrolimus has been used in chronic cutaneous lupus; however, there are no cases in the literature describing its use in TLE. While promising new treatments for lupus are on the horizon, clinical trials are typically aimed at adults with systemic or discoid lupus erythematosus and not TLE. In children, TLE is extremely rare and treatment protocols



**Figure 5** Patient #2, Biopsy of the left postauricular area. Histology exhibits Superficial and deep periadnexal lymphocytic infiltration with a hint of mucin deposition consistent with tumid lupus erythematosus [hematoxylin and eosin stain, magnification x40].

are based upon adult protocols, case reports, and anecdotes.

While improvement of TLE symptoms with hydroxychloroquine is expected, it can take up to three months to achieve clinical significance. It is imperative that patients understand this chronicity so that they are patient and compliant with their treatments. Both of our patients had very rapid responses to the treatment regimen which included a combination of hydroxychloroquine, aggressive sun protection, and for one patient, a topical steroid. They experienced improvement of their cutaneous symptoms in only a few weeks.

These cases highlight the variable clinical presentations of tumid lupus erythematosus and the importance of clinicopathologic correlation.

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