CASE REPORT

Recurrent lupus profundus-like panniculitis associated with streptococcal throat infections in an immunocompetent child

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Key words: infection; lupus erythematosus profundus; panniculitis; Streptococcus.

INTRODUCTION

We report the unusual case of an immunocompetent child in whom recurrent lupus profundus-like panniculitis associated with group A Streptococcus throat infections developed. Her skin findings were independent of the presence of bacterial organisms and resistant to antibiotics alone, suggesting an inflammatory, immune-mediated process. Surprisingly, histopathology testing found characteristics of lupus erythematosus profundus in an acute panniculitis stage.

CASE REPORT

A previously healthy 7-year-old girl with no family history of autoimmune disorders presented with a 2-week history of an 8.5-cm x 4.5-cm annular, erythematous, indurated plaque with central hyperpigmentation on the right side of the chest (Fig 1). Three days before, the patient had fevers up to 101°F, sore throat, vomiting, and difficulty walking. Of note, the patient had presented to clinic 1 year previous with similar lesions on the face, chest, and arm, associated with positive anti-DNAse B strep antibody.

Throat culture identified moderate growth of Streptococcus pyogenes. Laboratory values were significant for positive anti-DNAse B strep antibody (3 times the reference range), antinuclear antibody titer of 1:80 (speckled pattern), and mildly elevated erythrocyte sedimentation rate and C-reactive protein. Negative or normal tests included complete blood

Abbreviations used:
EN: erythema nodosum
LEP: lupus erythematosus profundus
SLE: systemic lupus erythematosus

...count, complete metabolic panel, anti—double-stranded DNA, complement levels, anti—neutrophil cytoplasmic antibodies, Lyme titer, rheumatoid factor, Sjogren antibodies, and immunoglobulin levels.

Biopsy found mild interface dermatitis with a mixed suppurative and lymphocytic panniculitis (Fig 2). A mild perivascular infiltrate of predominantly mononuclear cells surrounded vessels of the superficial plexus. The epidermis showed scattered foci of vacuolar changes, suggesting cellular injury. Neutrophils and lymphocytes were found replacing adipocytes, and colloidal iron highlighted mucin in the deeper dermis. Period acid-Schiff and Grocott methenamine silver stains were negative for organisms; no basement membrane zone thickening was detected on period acid-Schiff stain. Biopsy result was indicative of an acute panniculitis with features of lupus erythematosus profundus (LEP).

In the initial episode, lesions resolved within 4 weeks on oral prednisolone acetate, 1 mg/kg for 5 days followed by a 3-day taper. Because of concerned for side effects, systemic steroids were avoided during the second episode. When oral...

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amoxicillin failed to improve cutaneous sequelae, intralesional triamcinolone was given with minimal improvement in 1 week. Complete resolution occurred 1 month later. The patient was referred to a rheumatologist who did not identify a systemic autoimmune disorder. The patient did not have other rashes, oral ulcers, photosensitivity, or arthritis. She is doing well months later.

**DISCUSSION**

To our knowledge, only a few cases of panniculitis unrelated to erythema nodosum have been linked to *S. pyogenes*, or group A *Streptococcus*, in immunocompetent children. All patients had bacteremia, with upper respiratory infection being the most common source. In 2 cases, panniculitis persisted despite resolution of upper respiratory symptoms or antibiotic treatment, eventually self-resolving in 3 to 5 months. Interestingly, organisms were not detected on histology in these latter cases, whereas they were visible in the cases that rapidly responded to antibiotics. No case showed lupus panniculitis-like features on pathology.

We propose that bacterial antigens trigger an immune response, resulting in panniculitis. In antibiotic-resistant cases, such as with our patient, group A *Streptococcus* appears to be the trigger for inflammatory processes, even remote from the site of bacterial infection including in skin, joints, and other organs. It is hypothesized that bacterial infections expose adipocytes, increasing vulnerability to autoimmune processes. Autoimmunity would explain the treatment effectiveness of immunosuppressant therapies, such as steroids, as opposed to antibiotic monotherapy. A similar phenomenon is encountered with erythema nodosum (EN), in which nearly half of cases in children are linked to streptococcal infections. EN is believed to be a type IV delayed hypersensitivity reaction to a variety of antigens. These reactions are also reminiscent of poststreptococcal reactive arthritis, associated with HLA-DRB1*01, further suggestive of an autoimmune mechanism. It is unclear why certain individuals are more susceptible to one remote reaction versus another.

The interesting histopathology of our case requires the consideration of 3 forms of panniculitis: acute panniculitis, EN, and LEP. Acute panniculitis is described by neutrophilic infiltrates of fat lobules, as in our case, and is often accompanied by white cell necrosis and variable adipose necrosis. Although acute panniculitis is most frequently associated with EN, most, if not all, forms of panniculitis likely have an acute inflammatory phase in which neutrophils predominate. Particularly, this neutrophilic panniculitis phenomenon can be seen as an id reaction to bacterial infection. Therefore, the presence of neutrophilic infiltrates in fat lobules in itself is not predictive of EN. Classically, EN presents on the lower extremities; histopathology testing finds a septal panniculitis, neutrophilic infiltrates around proliferating capillaries, and actinic radial granulomas. Besides neutrophilic infiltrates with a pattern consistent with acute panniculitis, our patient did not have findings of EN.

Unlike EN, LEP tends to present on the arms, shoulders, face, and buttocks, as in our patient. In half of LEP, atrophy of the epidermis, vacuolar change at the dermoepidermal junction, thickened basement membrane, interstitial mucin between collagen bundles of the dermis, and superficial and deep perivascular inflammatory infiltrate of lymphocytes involving the dermis are seen. In other cases, lobular panniculitis with predominantly lymphohytic infiltrate is identified in the subcutaneous layer of the skin. Because of the presence of mild interface dermatitis, lobular panniculitis, and dermal mucin in our case, we favored the diagnosis of LEP-like

![Clinical image of an annular, erythematous plaque with indurated borders on the chest of a 7-year-old girl.](https://example.com/image.png)
panniculitis, attributing the presence of neutrophils to an acute inflammatory phase. Other investigators have also described a mixed neutrophilic and lymphoid panniculitis in lupus panniculitis. Most children with LEP undergo biopsies months to years after initial presentation, and we are likely missing the acute inflammatory phase we suspect we captured. The presence of interface changes and dermal mucin disfavors acute Sweet syndrome in spite of the presence of neutrophils in the dermis.

We share our experience with an immunocompetent child who had a recurring Streptococcus-induced lupus panniculitis-like eruption. The unusual histopathologic findings are suggestive of an acute panniculitis favoring LEP. Because acute panniculitis is not diagnostically predictive, clinical pathologic correlation is imperative. Because of the lack of organisms on biopsy and response to steroids rather than antibiotics, we presume our patient’s panniculitis to be an immunologic reaction. For such a patient, we recommend close follow-up, workup for autoimmune diseases, and referral to the rheumatology department.

REFERENCES

1. Kakourou T, Theodoridou M, Stefanaki K, et al. Two cases of panniculitis complicating Neisseria meningitidis serogroup A and Streptococcus pyogenes bacteremia, respectively. J Eur Acad Dermatol Venereol. 2006;20:1165-1167.

2. Randle SM, Richter MB, Palmer RG, et al. Panniculitis: a report of four cases and literature review. Arch Dis Child. 1991;66:1057-1060.

3. Villasenor-Park J, Davis A, Singh S, et al. Group A streptococcal panniculitis. Pediatr Dermatol. 2014;31:256-258.

4. Pao W, Duncan KO, Bologna JL, et al. Numerous eruptive lesions of panniculitis associated with group A streptococcus bacteremia in an immunocompetent child. Clin Infect Dis. 1998;27(3):430-433.

5. Black JM, Le L, Altmeyer M, Luster W, Shrum J, Wang A. Sandpaper dermatitis on a 17-month-old girl. Bacterial infectious panniculitis. Arch Dermatol. 2011;147(4):499-504.

6. Schwarz RA, Nervi SJ. Erythema nodosum: a sign of systemic disease. Am Fam Physician. 2007;75:695-700.

7. Shulman ST, Ayoub EM. Poststreptococcal reactive arthritis. Curr Opin Rheumatol. 2002;14:562-565.

8. Forstrom L, Winkelmann RK. Acute panniculitis: a clinical and histopathologic study of 34 cases. Arch Dermatol. 1977;113:909-917.

9. Magro CM, Dyrsen ME, Crowson AN. Acute infectious id panniculitis/panniculitis bacterid: a distinctive form of neutrophilic lobular panniculitis. J Cutan Pathol. 2008;35:941-946.

10. Raquena L, Sanchez Yus E. Panniculitis. Part II. Mostly lobular panniculitis. J Am Acad Dermatol. 2001;45:325-361. quiz 362-364.

11. Brinster NK, Nunley J, Pariser R, et al. Nonbullous neutrophilic lupus erythematosus: a newly recognized variant of cutaneous lupus erythematosus. J Am Acad Dermatol. 2012;66:92-97.

12. Weingartner JS, Zedek DC, Burkhart CN, et al. Lupus erythematosus panniculitis in children: report of three cases and review of previously reported cases. Ped Dermatol. 2012;29:169-176.