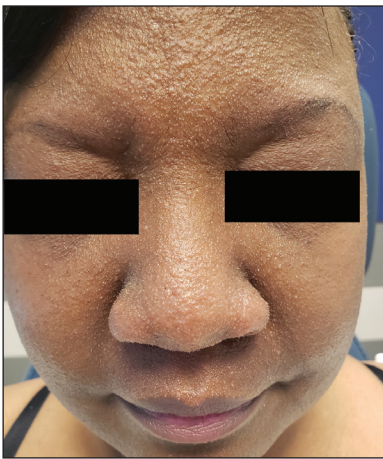


# Flesh-Colored Pinpoint Papules With Fine White Spicules on the Upper Body

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A 54-year-old Black woman presented with a rash that developed 6 months after a renal transplant due to a history of systemic lupus erythematosus with lupus nephritis. She was started on mycophenolate mofetil and tacrolimus after the transplant but was switched to cyclosporine because of BK viremia. The rash developed 1 week after cyclosporine was initiated and consisted of pruritic papules that started on the face and spread to the trunk and arms. Physical examination revealed innumerable follicular-based, keratotic, flesh-colored, pinpoint papules with fine white spicules on the face (top), neck, chest, arms, and back. Leonine facies was seen along the glabella with madarosis of the lateral eyebrows (top) and ears (bottom).



## WHAT'S YOUR DIAGNOSIS?

- keratosis pilaris
- lichen nitidus
- scleromyxedema
- trichodysplasia spinulosa
- trichostasis spinulosa

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The authors report no conflict of interest.

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## THE DIAGNOSIS: Trichodysplasia Spinulosa

A diagnosis of trichodysplasia spinulosa (TS) was rendered based on the clinical presentation—diffuse folliculocentric keratotic papules with spicules and leonine facies—coinciding with cyclosporine initiation. Biopsy was deferred given the classic presentation. The patient applied cidofovir cream 1% daily to lesions on the face. She was prescribed leflunomide 10 mg daily, which was later increased to 20 mg daily, for polyarthritis associated with systemic lupus erythematosus (SLE). Her transplant physician increased her cyclosporine dosage from 50 mg twice daily to 75 mg each morning and 50 mg each evening due to rising creatinine and donor-specific antibodies from the renal transplant. The patient's TS eruption mildly improved 3 months after the cyclosporine dose was increased. To treat persistent lesions, oral valganciclovir was started at 450 mg once daily and later reduced to every other day due to leukopenia. After 3 months of taking valganciclovir 450 mg every other day, the patient's TS rash resolved.

Trichodysplasia spinulosa is a rare condition caused by TS-associated polyomavirus<sup>1</sup> that may arise in immunosuppressed patients, especially in solid organ transplant recipients.<sup>2</sup> It is characterized by spiculated and folliculocentric papules, mainly on the face,<sup>1</sup> and often is diagnosed clinically, but if the presentation is not classic, a skin biopsy can help to confirm the diagnosis. Because of its rarity, treatment options do not have well-established efficacy<sup>1</sup> but include reducing immunosuppression and using the antivirals cidofovir<sup>1</sup> or valganciclovir<sup>3</sup> to treat the polyomavirus. Topical retinoids,<sup>3</sup> photodynamic therapy,<sup>4</sup> and leflunomide<sup>5</sup> also may be effective.

Although the typical approach to treating TS is to reduce immunosuppression, this was not an option for our patient, as she required increased immunosuppression for the treatment of active SLE. Leflunomide can be used for SLE, and in some reports it can be effective for BK viremia in kidney transplant recipients<sup>5</sup> as well as for TS in solid organ transplant recipients.<sup>6</sup> Our patient showed improvement of the TS, BK viremia, renal function, and SLE while taking leflunomide and valganciclovir.

The differential diagnosis includes keratosis pilaris, lichen nitidus, scleromyxedema, and trichostasis spinulosa. Keratosis pilaris is a benign skin disorder consisting of patches of keratotic papules with varying degrees of erythema and inflammation that are formed by dead keratinocytes plugging the hair follicles and often are seen on the extremities, face, and trunk.<sup>7</sup> Our patient's papules were flesh colored with no notable background erythema. Additionally, the presence of leonine facies was atypical for keratosis pilaris. Acids, steroids, and kinase inhibitors are the most frequently used treatments for keratosis pilaris.<sup>8</sup>

Lichen nitidus is a skin condition characterized by multiple shiny, dome-shaped, flesh-colored papules usually found on the flexor surfaces of the arms, anterior trunk, and genitalia. It is mostly asymptomatic, but patients may experience pruritus. Most cases occur in children and young adults, with no obvious racial or gender predilection. The diagnosis often is clinical, but biopsy shows downward enlargement of the epidermal rete ridges surrounding a focal inflammatory infiltrate, known as a ball-in-claw configuration.<sup>9-11</sup> Lichen nitidus spontaneously resolves within a few years without treatment. Our patient did have flesh-colored papules on the arms and chest; however, major involvement of the face is not typical in lichen nitidus. Additionally, fine white spicules would not be seen in lichen nitidus. For severe generalized lichen nitidus, treatment options include topical corticosteroids, topical calcineurin inhibitors, oral antihistamines, or UV light to decrease inflammation.<sup>9-11</sup>

Scleromyxedema is a rare condition involving the deposition of mucinous material in the papillary dermis to cause the formation of infiltrative skin lesions.<sup>12</sup> It is thought that immunoglobulins and cytokines secreted by inflammatory cells lead to the synthesis of glycosaminoglycans, which then causes deposition of mucin in the dermis.<sup>13</sup> The classic cutaneous features of scleromyxedema include waxy indurated papules and plaques with skin thickening throughout the entire body.<sup>12</sup> Our patient's papules were not notably indurated and involved less than 50% of the total body surface area. An important diagnostic feature of scleromyxedema is monoclonal gammopathy, which our patient did not have. Intravenous immunoglobulin is the first-line treatment of scleromyxedema, and second-line treatments include systemic corticosteroids and thalidomide.<sup>14</sup> Our patient also did not require treatment with intravenous immunoglobulin, as her rash improved with antiviral medication, which would not address the underlying inflammatory processes associated with scleromyxedema.

Trichostasis spinulosa is a rare hair follicle disorder consisting of dark, spiny, hyperkeratotic follicular papules that can be found on the extremities and face, especially the nose. The etiology is unknown, but risk factors include congenital dysplasia of hair follicles; exposure to UV light, dust, oil, or heat; chronic renal failure; *Malassezia* yeast; and *Propionibacterium acnes*. Adult women with darker skin types are most commonly affected by trichostasis spinulosa.<sup>15,16</sup> Our patient fit the epidemiologic demographic of trichostasis spinulosa, including a history of chronic renal failure. Her rash covered the face, nose, and arms; however, the papules were flesh colored, whereas trichostasis spinulosa would appear as black papules. Furthermore, yeast and bacterial infections have been

identified as potential agents associated with trichostasis spinulosa; therefore, antiviral agents would be ineffective. Viable treatments for trichostasis spinulosa include emollients, topical keratolytic agents, retinoic acids, and lasers to remove abnormal hair follicles.<sup>15,16</sup>

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