

An eruption of numerous spiny papules in a pediatric transplant patient



School of Medicine

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SYNOPSIS

- Viral associated trichodysplasia spinulosa (VATS) is a rare skin eruption characterized by folliculocentric papules and keratotic spicules on the central face and ears, with concomitant alopecia of the eyebrows and eyelashes. (1)
- VATS occurs in the setting of immunosuppression, putting patients undergoing long term immunosuppressive therapy, such as transplant patients, at increased risk. (1)
- When possible, VATS should be treated by reducing or reversing the immunosuppression. If this is not safe, as in this case, topical 1% or 3% cidofovir is an effective alternative. (2,3) (4,5)
- Here, we describe a case of VATS in a renal transplant patient with a history of antibody mediated transplant rejection, necessitating a creative treatment strategy.

OBJECTIVES

- Familiarize clinicians with the cutaneous and histological findings associated with VATS.
- Describe the pathogenic process and infectious agent responsible for VATS.
- Describe the ideal treatment strategies for VATS and offer effective alternatives if the first line option is contraindicated.

CASE PRESENTATION

- An eight-year-old African American female presented to the dermatology clinic with a three-month history of numerous, tiny, asymptomatic, white folliculocentric papules and spicules on the face, with concomitant alopecia of the eyebrows and eyelashes (Figures 1).
- The patient had a past medical history of congenital renal dysplasia.
- The patient had a history of antibody-mediated rejection four months post-transplant which was treated with intravenous methylprednisolone and rituximab.
- Her current immunosuppressive medications included oral tacrolimus, mycophenolate mofetil, and prednisolone, no adjustments had been made to this regimen for 7 months prior to presentation.
- Laboratory testing revealed a negative serum BK virus PCR.
- A 3mm punch biopsy of the left ear revealed a dilated follicular canal, underlying cyst formation with a granular layer, and keratinous material and inflammatory debris in the lumen. The surrounding dermis showed fibrosis, dilated vessels and sparse perifollicular inflammatory cell infiltrate. Abnormal eosinophilic inclusions were noted surrounding the inner root sheath of a hair follicle (Figure 3).
- After 8 months of treatment with compounded topical 1% cidofovir, the patient showed improvement of the perifollicular papules, facial spicules, and eyelash and eyebrow regrowth, without experiencing any adverse effects (Figure 4).

RESULTS



Figure 1



Figure 2

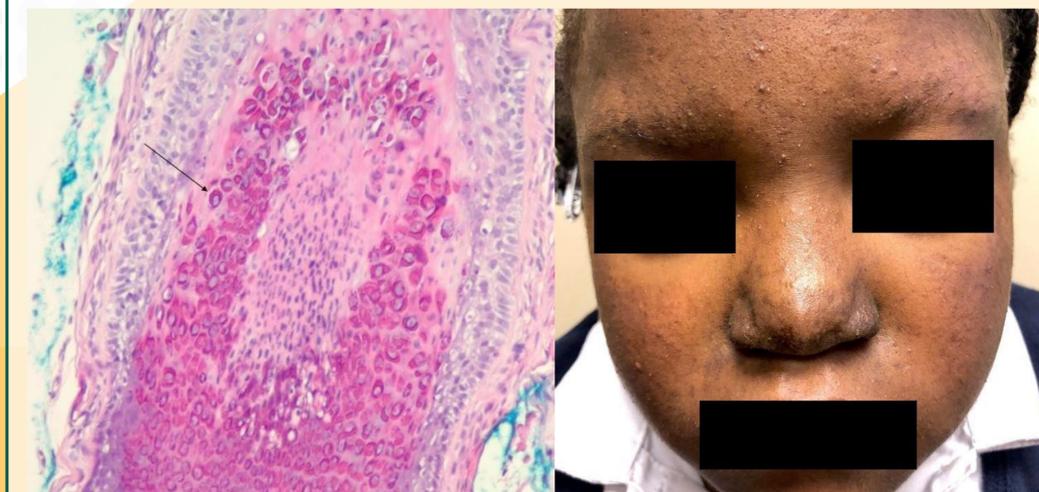


Figure 3



Figure 4

Figure 1: 8-year-old child status post renal transplant with numerous tiny white folliculocentric papules and spicules on the face with concomitant alopecia of the eyebrows and eyelashes.

Figure 2: Closer view emphasizing the morphology of the spicules on the nose.

Figure 3: Dermatopathology of a hair follicle at 100X magnification showing abnormal cytoplasmic eosinophilic inclusions (arrow) in the inner root sheath.

Figure 4: 8-month follow-up visit showing improvement of the folliculocentric papules and spicules on the face with associated post-inflammatory hyperpigmentation and regrowth of the eyebrows and eyelashes.

DISCUSSION AND CONCLUSION

- We present a rare case of viral associated trichodysplasia spinulosa in a pediatric transplant patient. (1)
- VATS is caused by trichodysplasia spinulosa polyomavirus (TSPyV) – a ubiquitous virus whose pathologic effects are only seen in the context of immunosuppression.(4)
- VATS presents with folliculocentric papules and keratin spicules on the central face and ears, with concomitant alopecia of the eyebrows and eyelashes.(1)
- TSPyV proliferates in the inner root sheath of hair follicles, resulting in abnormally increased cytoplasmic collections of eosinophilic keratin protein (trichohyalin) on histology.(1) These collections were apparent in our patient (Figure 2).
- TSPyV PCR can confirm viremia, but given this test's lack of availability, BK viral loads have been suggested as a surrogate marker for TSPyV viral load.(3)
- Electron microscopy may aid in diagnosis as it allows for visualization of intranuclear viral particles within keratinocytes of hair follicles, though it is often unnecessary due to the clinically distinct presentation.(1, 3-5)
- While VATS prevalence is roughly equal between adults and children, transplant patients are at particular risk for VATS due to the immunosuppressive regimens required to maintain graft viability.(5)
- When possible, VATS is treated by reducing or reversing the immunosuppression. If not safe for the patient, as in this case, topical 1% cidofovir is an effective alternative. (2,3)

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