Trichodysplasia spinulosa: A rare complication of immunosuppression

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INTRODUCTION

Trichodysplasia spinulosa (TS) is a recently described rare disease of immunosuppressed patients characterized by folliculocentric papules and keratin spicules. The condition is highly associated with the TS-associated polyomavirus (TSPyV); however, the causal mechanism remains undiscovered. Currently, there is a dearth of case reports on this condition, and treatment for TS is not well established. We report a case of TS that successfully responded to reduction of immunosuppression.

Abbreviations used:

TS: trichodysplasia spinulosa
TSPyV: TS-associated polyoma virus

CASE REPORT

A 35-year-old man with a history of end-stage renal disease secondary to hypertension status post allogeneic renal transplant presented to the dermatology clinic with complaints of asymptomatic skin-colored lesions on the trunk and extremities and a decreased hair density of the eyebrows and eyelashes. The lesions started about 6 months after kidney transplantation, and he was seen in the clinic after the rash progressed for 6 additional months. Immunosuppressive medications at the time of onset included tacrolimus and mycophenolate mofetil. After onset, the patient's nephrologist discontinued tacrolimus and started the patient on alternative immunosuppression comprising cyclosporine and mycophenolate mofetil; also, he initiated treatment for the rash with daily low-dose oral prednisone. Despite the medication changes, the patient's rash and hair loss continued to worsen over the next few months. Clinical examination found scattered 1- to 2-mm follicular spiny skin-colored papules on the face, trunk, arms, and ears (Fig 1). Additionally, alopecia of the eyebrows and eyelashes was seen (Fig 2, *A*). A biopsy of a papule from the helix found dilated follicles with abnormally large trichohyalin granules occupying most of the follicular epithelium (Fig 3), consistent with TS. Our patient underwent a slow taper of his immunosuppression and showed improvement after 4 months with marked improvement of his eyebrow and eyelash alopecia at 6-month

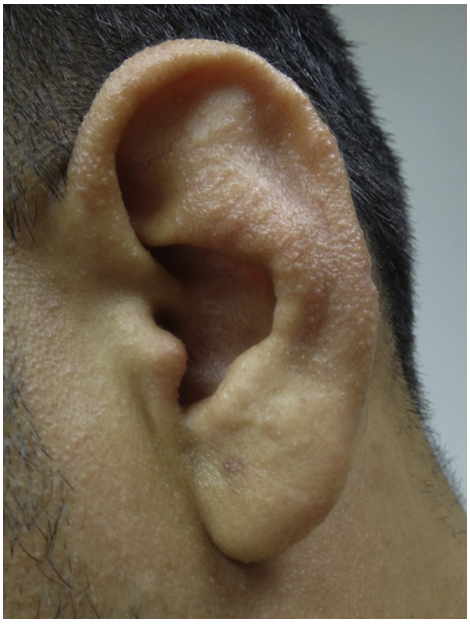

Fig 1. Trichodysplasia spinulosa. Spiny keratotic papules in a follicular distribution were also present on the upper extremities, trunk, and face.

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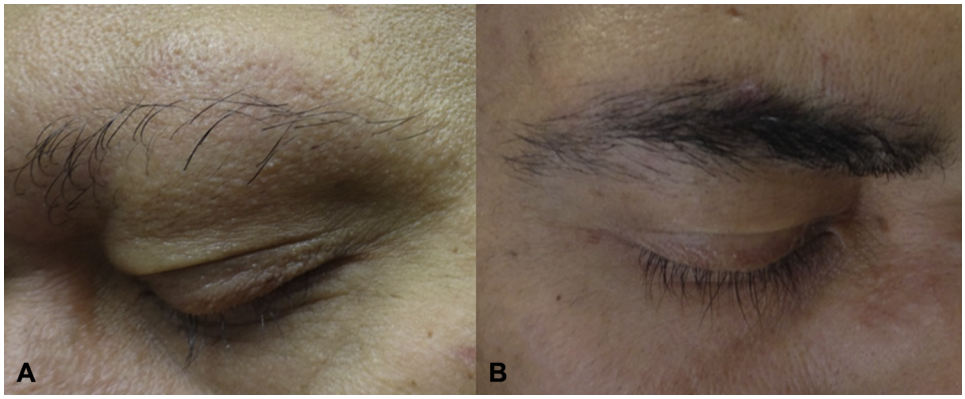

Fig 2. Trichodysplasia spinulosa. **A**, Alopecia of eyebrows and eyelashes upon presentation to clinic. Also note the scattered follicular papules. **B**, Markedly increased hair density of eyebrows and eyelashes at 6-month follow-up with absent follicular papules.

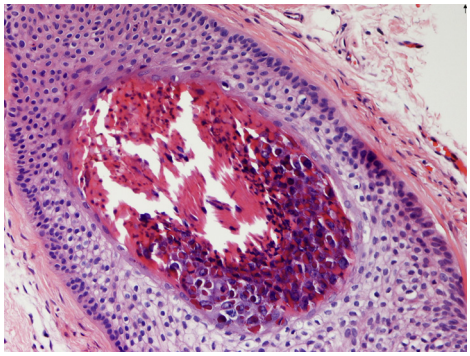

Fig 3. Trichodysplasia spinulosa. Histologic examination of a papule from the right helix found a dilated follicle with abnormally large trichohyalin granules occupying most of the follicular epithelium. Periodic acid–Schiff stain was negative for fungi. (Hematoxylin-eosin stain; original magnification: ×100.)

follow-up (Fig 2, *B*). At 2-year follow up, the patient exhibited complete regrowth of his eyebrows and eyelashes and a significant decrease in the number of keratotic papules.

DISCUSSION

TS is a rare disease that was first described in 1999 by Haycox et al.[1] A new human polyoma virus named *trichodysplasia-associated polyomavirus* (TSPyV) was isolated using rolling-circle DNA amplification by van der Meijden et al[2] in 2010. Diagnosis of TS is made with a combination of both clinical and pathologic evidence. In contrast, a causal relationship hinges on confirming TSPyV presence with further studies such as polymerase chain reaction or a newly created immunohistochemical stain for the polyomavirus middle T antigen.[3] A recent prevalence study found that 70% of adults exhibit seroconversion, with about one-third showing seroconversion by early childhood.[4] The ubiquitous nature of TSPyV parallels the widespread prevalence of other human polyomaviruses such as the BK and JC polyomaviruses.[5]

TS is characterized by an eruption of asymptomatic to pruritic folliculocentric papules with keratotic spiculations classically involving the mid portion of the face and ears and are found less frequently on the trunk and extremities.[1] TS is associated with variable degrees of nonscarring alopecia, most severely affecting the eyelashes and eyebrows. Progression to an infiltrated appearance or leonine facies has been described in some patients.[6]

Histopathology finds dilated and dysmorphic hair follicles and proliferation of inner root sheath cells with enlarged trichohyalin granules.[7] Kazem et al[7] found that TSPyV disrupts the retinoblastoma regulatory pathway, a probable cause of inner root sheath cell hyperproliferation; however, other cell cycle regulatory pathways remain unexplored.[7]

The long-term consequences of TSPyV infection are unknown. The JC and BK polyomaviruses are known causes of disease in immunosuppressed patients. Additionally, the Merkel cell polyomavirus has been isolated from most Merkel cell carcinoma samples.[8] Although there are no reported cases of clinically relevant tumors caused by TSPyV, the oncogenic potential warrants continued close observation of patients with the disease and may justify more aggressive treatment. There is no established treatment for TS, although topical cidofovir 1% to 3% cream and oral valganciclovir have shown clinical improvement.[9] In patients with organ transplants like our patient, management typically begins with reducing immunosuppression while avoiding graft rejection. The patient discussed here underwent successful treatment of his TS with only a decrease in immunosuppression—additional therapy was not necessary.

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