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notably improved after 2 weeks. There was nearly complete resolution of those atrophic acne scars like lesions and follicle plugs at the end of the 4th week [Figure 3]. Only some yellow or light-brown pigmentation was left on the forehead.

KFS is actually not rare in China, although it is not yet described in the English literature. This is not a typical KFS as previously reported and should be differentiated from atrophic scars of acne and keratosis pilaris. As she had no history of acne occurrence in the past and atrophic lesions could not disappear by themselves, the former dermatosis can easily be excluded. Keratosis pilaris and keratosis follicularis squamosa share the histological finding of keratotic plugging; however, the KFS lesion is usually clinically and histologically surrounded by fine lamellar scales, while typical eruption of keratosis pilaris looks like a horny papule, rather than a black dot as seen in KFS. Additionally, the genetic contribution in KFS is smaller than in keratosis pilaris. Bacterial infection, irritation by clothing, heredity and hormone imbalance have been proposed as pathogenetic factors for KFS, but our patients had no indications of the above possibilities.

Traditionally, oral antibiotics, such as minocycline and roxithromycin, and topical exfoliative agents have been used for the treatment of KFS. Sadahira et al. reported a case successfully treated with topical tacalcitol. We chose pimecrolimus to treat the patient and the lesions were markedly improved after the therapy. Pimecrolimus (SDZ ASM 981), an ascomycin derivative and calcineurin inhibitor, is a nonsteroid, has anti-inflammatory activity, and has demonstrated efficacy in reducing symptoms of atopic dermatitis in adult and pediatric patients when applied topically. It binds cytoplasmic proteins and the resulting complex binds calcineurin, inhibiting its ability to dephosphorylate the nuclear factor of activated T cells, thus suppressing gene transcription. Rubeqni et al. reported a case of extensive Darier's disease successfully treated with topical tacrolimus, after stopping of oral isotretinoin due to major depression. However, the mechanism of calcineurin inhibitor to treat hyperkeratotic skin disorders remains unclear. We have been following the patient for 6 months with no recurrence.

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Atrichia congenita with papular lesions

Sir,

Atrichia congenita with papular lesions (APL) represents a complex and heterogenous group of genodermatoses characterized clinically by complete and irreversible hair loss shortly after birth, and is associated with the development of keratin-filled cysts over the body resulting from homozygous mutations in the hairless gene (HR).

A 38-year-old lady presented with complete loss of hair on the body. She had scanty scalp hair at birth which was gradually lost. At 1 year of age, she developed multiple skin colored lesions on the face and body, which progressively increased in number. By the
age of 2 years, she had complete loss of scalp and body hair. Her alopecia was unresponsive to therapy [Figure 1]. She did not give a history of delayed milestones, decreased sweating, dimness of vision, decreased hearing, seizures, atopy, or bone pains. She was born of a second degree consanguineous marriage. Her siblings were asymptomatic. None of her family members had similar complaints.

On examination, she had complete absence of hair on the body and scalp. Multiple skin colored smooth papules and nodules of size 0.5–2 cm were present on the entire body. Suture lines on the scalp were hypopigmented. She had no abnormalities related to her mucosae, nails, teeth and sweat glands. Her palms and soles were normal. She had no bony abnormalities, dysmorphic features or systemic involvement.

The differential diagnoses considered for this patient included Alopecia universalis, APL, Vitamin D dependent rickets type II and Ectodermal dysplasia.

A skin biopsy from a nodule on the right forearm revealed multiple dermal cysts containing keratinous material. No normal hair follicles were visualized. There was no evidence of inflammation [Figure 2].

Based on the clinical presentation and histopathology, a diagnosis of APL was made.

APL is a rare, autosomal recessive form of total alopecia of the scalp, eyebrows, eyelashes, axillary and pubic hair, characterized by hair loss soon after birth and the development of keratin-filled cysts over extensive areas of the body. The patients have normal development, hearing, teeth and nails. There are no abnormalities of sweating. Heterozygous individuals have normal hair and are clinically indistinguishable from genotypically normal persons.[2] This condition has been noted for decades among gypsies known as Irish Travelers, who have existed as a distinct indigenous ethnic minority within Ireland. It was first referred to as congenital atrichia by Ahmad et al.[3]

The hair matrix cells in APL appear to undergo a premature and massive apoptosis, together with a concomitant decline in Bcl-2 expression, a loss of Neural Cell Adhesion Molecule (NCAM) positivity, and a disconnection from the overlying epithelial sheath essential for the movement of the dermal papilla. As a consequence, the hair bulbs and dermal papillae remain stranded in the dermis, and indispensable messages between the dermal papillae and stem cells in the bulge are not transmitted, so no further hair growth occurs.[2]

The molecular basis of this disease remains obscure. Mutations in the human hairless gene located on chromosome 8p21.2 which encodes a putative single zinc-finger transcription factor protein are believed to regulate catagen remodeling in the hair cycle. A missense mutation (C622G) in the zinc-finger domain of the human hairless gene, a single base pair deletion (3434del C) in the hairless gene and deletion mutations in exons 2 and 8 of the human hairless gene have been described in APL. Phenotypic heterogenicity suggests different roles for different regions of the hairless gene mutated in APL.[2]

Miller et al.[4] reported a patient with Vitamin D

![Figure 1: Papular lesions on the face associated with universal alopecia. Patient is wearing a wig](image1)

![Figure 2: Histopathology of a nodule on the right forearm revealed multiple dermal cysts containing keratinous material. No normal hair follicles were visualized. There was no evidence of inflammation (H and E, x40)](image2)
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resistant rickets type IIA, a compound heterozygote for mutations in the Vitamin D receptor gene (VDR) in which the phenotype of atrichia with papular lesions was identical to that seen in patients carrying mutations in the HR gene. It is hypothesized that the VDR and HR genes, which are both zinc-finger proteins, may be in the same genetic pathway that controls postnatal cycling of the hair follicle.[4]

Yip et al.[5] proposed revised clinical criteria for APL based on their personal observation and have made a retrospective analysis of cases described in literature. These features are listed in Table 1.

Published estimates of the prevalence of APL remain surprisingly low, considering that pathogenetic mutations in HR have been found in distinct populations around the world.[2] APL is more common than previously thought and is often mistaken for the putative autoimmune form of alopecia universalis.[5]

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**ACKNOWLEDGMENTS**

We would like to acknowledge the contributions of Dr. Priva Aley Joseph MD, Dr. Joyce Ponnaiya MD and Dr. Mary Jacob DVD MNAMS.

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**REFERENCES**


**Treatment of severe nail psoriasis with etanercept**

Sir,

Nail involvement is common in psoriasis and has been reported to occur in up to 50% of patients.[1] It can cause significant physical impairment, severe distress and pain.[1,2] There is a broad spectrum of nail dystrophies associated with psoriasis, ranging from the common pitting and loosening of the nail plate to the less frequent discoloration and splinter hemorrhages seen in the nail bed.[1] It is frequently refractory to treatment and there is no standardized therapy regimen.[3] Etanercept is a fully humanized, soluble tumor necrosis factor (TNF)-alpha receptor approved for the treatment of plaque psoriasis. It has been shown to be safe and to have long-term efficacy for treatment of moderate to severe psoriasis resistant to other modes. Some reports show that etanercept may have significant benefit in the treatment of psoriatic nail disease, although not approved for its treatment (as only manifestation).[1-4]