with deep breathing and an abnormal Valsalva response. Higher mental functions, muscle tone and power, as well as both superficial and deep reflexes were normal. Routine investigations (including blood sugar), ultrasonography of the abdomen, electrocardiogram, magnetic resonance imaging of the brain, and nerve conduction velocity studies of the peripheral nerves did not reveal any abnormalities. Cutaneous histopathology was normal with preservation of the sweat glands.

A large number of cutaneous or systemic conditions may underlie anhidrosis. These include neurologic disorders such as Guillain-Barre syndrome, heatstroke, diabetes, congenital disorders including ectodermal dysplasia, drugs, autonomic neuropathy, infections of or trauma to the sweat glands, burns, and excessive dehydration.

Localized anhidrosis is of limited clinical importance apart from its diagnostic value in leprosy. On the other hand, generalized or extensive anhidrosis (such as in the case of anhidrotic ectodermal dysplasia) may lead to hyperpyrexia with its associated complications. Disorders of the autonomic nervous system may result from pathology of either the central or peripheral nervous systems. Pure pandysautonomia is clinically characterized by some combinations of anhidrosis, orthostatic hypotension, paralysis of papillary reflexes, loss of lacrimation and salivation, impotence, impaired bladder and bowel function, flushing, and heat intolerance. Somatosensory and reflex functions are usually spared. This patient showed an absence of sweating along with other features of autonomic failure in the form of urinary and fecal urgency, postural hypotension, asymmetry of the pupils with an absence of a light reflex, as well as an abnormal Valsalva response. Moreover, a histopathological examination of the skin did not reveal any abnormalities, indicating that the anhidrosis was due to autonomic failure. The neuroimaging and nerve conduction velocity were normal; hence, a diagnosis of idiopathic generalized anhidrosis was made.

Patients with generalized anhidrosis have a dangerous inability to tolerate heat. Therefore, when the weather is dry and hot, the inability to sweat can be life-threatening due to the potential to develop heat stroke. Anhidrosis may go unrecognized until a substantial amount of heat or exertion fails to cause sweating. This forms the background of the hyperpyrexia seen in our patient. In rare cases, family members of such patients may have anhidrosis and anisocoria, but our patient did not have any such family history. In conclusion, anhidrosis can be present in some stages of autonomic failure, and a dermatologist must be aware of this fact so as to be able to routinely rule out the possibility of autonomic dysfunction when dealing with a case of anhidrosis.

Intertriginous granular parakeratosis persisting for 20 years

Sir,
We read with interest the article “Granular parakeratosis presenting with facial keratotic papules” by Joshi and Taneja. We report here a case of granular parakeratosis that involves multiple intertriginous areas.

A 25 year-old male presented with a pruritic eruption in the intertriginous areas involving the axilla, inner thighs, and perineum. The lesions were characterized by interruped scales and biopsies showed intertriginous granular parakeratosis persisting for 20 years.
flexures that had been prevalent since he was five years old. The eruption was persistent throughout this period except for brief intervening periods, during which it resolved completely when he lived in cooler climatic conditions. He denied the use of deodorants, antiperspirants, and body sprays. No other family member had similar skin lesions.

Examination revealed multiple, hyperpigmented, linear, verrucous papules coalescing to form well-delineated, annular plaques in the vault of the axillae [Figure 1]. A well-defined, semiannular, hyperpigmented, macerated plaque was seen along with erosions and crusting over the groin, ventral surface of the penile shaft and the glans penis [Figure 2]. A well-defined, moist, macerated, grayish-white plaque was present between the buttocks and the perianal region. The clinical differential diagnoses considered were Hailey-Hailey disease and pemphigus vegetans.

A potassium hydroxide wet mount preparation from the lesion did not show any fungal hyphae. Skin biopsy showed a markedly thickened and compact stratum corneum. The nuclei were preserved throughout the thickened stratum corneum [Figure 3]. Instead of the expected absence of keratohyaline granules from the cytoplasm of horny cells, these organelles were present throughout the stratum corneum [Figure 4]. A thickened stratum granulosum was observed. There was no evidence of neutrophils in the epidermis. Spongiosis and acantholysis were absent and direct immunofluorescence did not show any immune deposits in the epidermis. Correlating the distribution and morphology of the lesions with the histology, a diagnosis of intertriginous granular parakeratosis was made. The patient did not respond to topical steroids, antibiotics, antifungals, tretinoin or oral isotretinoin.

In 1991, Northcutt, Nelson, and Tschen described a clinicopathological entity from a series of biopsy specimens of axillary lesions that showed an unusual and unique form...
of parakeratosis which they designated as “axillary granular parakeratosis”. In 1998, Mehregan, Thomas, and Mehregan described four more cases of granular parakeratosis, including a patient with isolated involvement of the inguinal region and suggested that the term “axillary granular parakeratosis” may be re-designated as “intertriginous granular parakeratosis.” Although more common in middle-aged and elderly women, granular parakeratosis can occur in both sexes and in all age groups. It has been described in infants as linear and geometric scaly plaques over the pressure points beneath their diapers. Rarely, as reported by Joshi and Taneja, it can also occur over non-intertriginous areas such as the face. Contact allergens and physical factors such as heat, moisture, friction, obesity, and occlusion from diapers were suspected to play a role. The resolution of the lesions in cooler climates points to sweating and friction as two possible factors responsible for the parakeratosis seen in our case.

It is presumed that granular parakeratosis is a reaction pattern secondary to various stimuli rather than a distinctive disease. The disturbance in keratinization in the form of a blockade of the conversion of profilaggrin into filaggrin units is suspected to be a basic pathogenic abnormality. Other than the characteristic distribution of lesions and unique histopathological features, granular parakeratosis has been observed as an incidental histopathologic finding in various unrelated conditions such as dermatomyositis, dermatophytosis, and molluscum contagiosum.

The therapeutic response of granular parakeratosis to various modalities of treatment has been inconsistent and frequently disappointing, and no controlled therapeutic trials have been conducted. Individual case reports have shown response to topical tretinoin[6] and to isotretinoin[7]. There are reports in which these eruptions responded to topically applied glucocorticoids,[8] antifungals, and antibiotics, as well as reports which showed no response to the above medications.

A review of literature confirmed that prevalence of these eruptions for 20 years (as observed in our patient) is the longest of all such reported cases. Involvement of multiple intertriginous areas has been published earlier[9] and this patient is the second such case to be reported. To conclude, this case is presented for its rarity, extreme chronicity, and the involvement of multiple intertriginous areas.

I. S. Reddy, G. Swarnalata, Tejal Mody
Departments of Dermatology and Pathology, Apollo Hospitals, Jubilee Hills, Hyderabad 500 033, India

REFERENCES