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vulgaris is still obscure. Hodgson et al. reported a 29-year-old HIV infected male with pemphigus vulgaris with little response to corticosteroids. There was marked improvement with thalidomide, but despite continued therapy there was recurrence of ulceration. Stray reports are available in the literature of occurrence of autoimmune vesiculobullous disease in HIV infection. Chou et al. reported epidermolysis bullosa acquisita in a patient with AIDS. Mitsubashi and Hohl reported a case of dermatitis herpetiformis in an HIV infected HLA-B8 positive patient. Levy et al reported an HIV infected patient with bullous pemphigoid who responded to ritodrine.

The clinical characteristics and response to therapy as far as pemphigus vulgaris is concerned appear not to be modified by HIV infection. Therapy of pemphigus vulgaris with immunosuppressive drugs may cause HIV disease progression and may necessitate initiation of antiretroviral therapy which may not be feasible in resource restricted set up. The impact of antiretroviral therapy and HIV infection on pemphigus needs further exploration.

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REFERENCES

Hereditary leukonychia totalis

Sir,
Leukonychia or white nails have been known from a long time. They are commonly known as gift or fortune spots. Unna classified leukonychia based on distribution of whiteness into the following - leukonychia punctata, leukonychia striata, leukonychia totalis and leukonychia partialis. Others classified it into true leukonychia, where the pathology involves the nail plate and apparent leukonychia, where the pathology is in the subungual tissue. Hereditary total leukonychia is a rare clinical entity.

A 66-year-old male patient presented with itchy and scaly lesions over the body of 2 months’ duration. Initial skin lesions were noted on the trunk that gradually extended over other parts of the body. There was no history of photosensitivity, oral ulceration or loss of hair. He was a known diabetic and was on metformin therapy. His eruptions were not attributable to metformin. For the previous 4 months, he was also taking ayurvedic medicines of unknown nature for presbyacusis. On cutaneous examination, he had multiple violaceous scaly papules as well as plaques distributed all over the body. Postinflammatory hyperpigmentation was observed at a few places. There was no abnormality in the oral or genital mucosa. All of his 20 nails showed white discoloration that involved the entire nail plate [Figure 1].

On further enquiry, the patient informed that the nails had
been white ever since he could remember. His maternal grandmother, mother, two of the four siblings and two of his three offsprings have had a similar condition of the nails. However, no one in his family had diabetes.

He was diagnosed to have a lichenoid drug rash with hereditary total leukonychia. He was advised to discontinue the use of ayurvedic medicines and was treated with oral dapsone 100 mg/day and was reassured regarding his white nails. During the next follow-up, after 4 weeks, he had marked improvement in his lichenoid rash; however, the nails remained white.

Abnormal keratinization of the nail plate is believed to be responsible for development of true leukonychia. Large keratohyaline granules are found in the keratinocytes and the keratohyaline containing cells reflected light, resulting in a white nail that prevents the visualization of the underlying vascular tissue. [2] It is difficult to ascertain whether our patient had true or apparent leukonychia. Our patient refused a nail biopsy.

There are very few reports of hereditary leukonychia totalis in the literature. [2-9] Familial leukonychia totalis has been found to be inherited in both autosomal dominant[3] and recessive patterns. [5]


Leukonychia totalis has also been associated with severe systemic diseases such as hepatic cirrhosis, chronic renal failure, congestive cardiac failure, diabetes mellitus, chronic hypoalbuminemia and Hodgkin's lymphoma. [16] Although our patient had diabetes mellitus, it does not appear to be the cause of leukonychia in him since he had the discoloration since early childhood and many of his family members were having a similar problem.

It has been proposed that leukonychia partialis may be a phase of leukonychia totalis, both being the expressions of the same genetic defect. [17] Both conditions occurring in different members of the same family [18] and on different digits of an individual have been described.

The importance of this report lies not only in the fact that this condition is rare but also that it may have associated abnormalities. If no association is detected, proper counseling of the patient regarding the benign nature of the condition should suffice.

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REFERENCES

Sir,
A one and a half month old boy presented with multiple swellings on the body. He had been born prematurely at 34 weeks, by caesarian section of a mother who had had pregnancy-induced hypertension. There was severe hypoxia at birth and he had a seizure on day one. On day two, he developed unconjugated hyperbilirubinemia, for which he was treated with phototherapy. He also developed thrombocytopenia and was given packed red cells. There was no history of steroid intake. The swellings were noticed 10 days after birth, had gradually increased in size and were bilaterally in the neck, axillae and inguinal area. They ranged from 1.5 to 2 cm and were mobile, firm and non-tender. There was also a 3 cm, firm, tender epigastric swelling. His hemoglobin was 8.5 gm% and a TORCH screening was negative. The clinical diagnosis was generalized lymphadenopathy of unknown cause. A biopsy of the axillary swelling, believed to be a lymph node was performed; this showed adipose tissue containing a lobular panniculitis with foci of fat necrosis. Multiple macrophages, foreign body giant cells and lymphoid cells were present. Many needle-shaped crystals in a radial arrangement were seen in the cytoplasm of the macrophages [Figure 1]. The histologic features were those of subcutaneous fat necrosis. Evaluation of serum calcium was advised but the parents chose to get the child discharged and the patient was lost to followup.

Subcutaneous fat necrosis is characterized by rubbery firm, mobile nodules and erythematous violaceous plaques over the trunk, arms, buttocks, thighs and cheeks. These lesions appear in neonates who have had fetal distress. The child is usually afebrile and appears well. Most cases of subcutaneous fat necrosis are self-limiting.[1,2] Thrombocytopenia is a common association with subcutaneous fat necrosis and anemia, as in our patient, has also been recorded.[1,3] Neonatal stress due to difficult delivery and hypothermia are believed to be aetiologic factors.[1,2] Recognition of the condition is important because a small but significant percentage of cases proceed to a hypercalcemia. When associated with hypercalcemia, seizures, failure to thrive, weight loss, irritability, apathy, hypotonia and even mortality can result.[1,2,4] Treatment consists of analgesics. Hypercalcemia must be treated aggressively with fluid loading, calcium wasting diuretics and low calcium/ vitamin D diet. Serum Calcium must be monitored for several weeks.[1,2,4] The differential diagnosis includes erythema nodosum and sclerema neonatorum. [5] These conditions have different morphologies and are unlikely to be misdiagnosed by a pathologist. Erythema nodosum is a septal panniculitis, with little fat necrosis of the lobules and no crystals. Children with sclerema neonatorum are severely ill and though the skin biopsy shows crystals, there is no inflammatory infiltrate or fat necrosis. Poststeroid panniculitis is a condition which is morphologically similar but is preceded...