FIXED DRUG ERUPTIONS IN A 9-MONTH-OLD CHILD

Ramji Gupta

A 9-month-old child developed fixed drug eruptions due to co-trimoxazole. Causative agent was confirmed by repeated provocation tests.

Key words : Fixed drug eruption, Co-trimoxazole, Nine-month-old child.

The incidence and causes of fixed drug eruption (FDE) have been reported from all over the world.[1-6] In majority of these reports, the patients have been adults. Recently, Kanwar et al[7] reported a series of 23 cases of FDE in children. The ages of the children in this series were 3-12 years. The present case is probably the first case of FDE occurring in a child at the age of 9 months.

Case Report

On December 24, 1986, a 21-month-old child was seen having generalised pigmented macules of various sizes. History dated back to the age of 9 months, when he had diarrhoea and vomiting, and was given two different types of syrups. After 4 hours, he developed three, erythematous, itchy macules over his abdomen which turned black during the next 10-12 hours. The diarrhoea and vomiting stopped along with the disappearance of itching during the next 2-3 days. Six months later, he again had a bout of diarrhoea and vomiting and was given similar types of syrups. There was reactivation of the old lesions with erythema and itching, along with development of a few new erythematous itchy macules in other parts of the abdomen within 15 minutes. During the next 6 months, he was repeatedly given the same syrups for similar symptoms with reactivation of the old lesions and development of a few new ones at each occasion. The lesions progressively increased in size and number and involved almost the entire body. In October 1986, he again developed cough with diarrhoea and vomiting, he was given some syrup, and was seen to develop erythematous macules which turned into vesicles, on his eyelids, lips, face and fingers. There were a few ulcers in the oral mucosa. The lesions were controlled in 7 days with systemic administration of syrup ampicillin 500 mg, syrup pheniramine maleate 22.5 mg and prednisolone 15 mg daily and local applications of calamine lotion on the skin and boro-glycerine in the mouth.

Provocation tests were started on January 1, 1987, with salbutamol syrup, cough syrup (diphenhydramine hydrochloride 14.08 mg, ammonium chloride 0.138 gm, sodium citrate 57.08 mg, chloroform 0.01 ml and menthol 1.14 mg in each 5 ml of syrup), co-trimoxazole (trimethoprim 40 mg, sulphamethoxazole 200 mg in each 5 ml), B complex, paracetamol and furazolidone as described by Gupta and Pasricha.[8] There was no reactivation of the lesions with salbutamol syrup, cough syrup, B complex, paracetamol and furazolidone tablets. However, all the lesions became red and started itching within ½ hour after 5 ml co-trimoxazole syrup which subsided spontaneously by the next morning. Provocation test was repeated on January 7, 1987 with 5 ml syrup of co-trimoxazole, Within ½ hour, itching started in all the lesions, with erythema in a few lesions. All the lesions became erythematous during the next 1 hour. Erythema and itching subsided in 6 hours with 1 mg betamethasone given orally.

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Comments

Repeated reactivation of all the lesions with co-trimoxazole syrup confirm that the child was having FDE due to co-trimoxazole. Kanwar et al[7] were the first to draw attention about the occurrence of FDE in children from Libya. They have recorded FDE in 2 children between the ages of 3 and 6 years. Majority of their patients were between the age of 6-12 years. Sehgal and Gangwani[9] in a review of FDE have mentioned 4 cases between the ages of 1 and 10 years. Shah et al[10] have recorded the occurrence of FDE at the age of 1 year in their series of 63 patients of all age groups. Olumide[3] has reported 12 cases of FDE between the ages of 6 and 50 years. In a series of 57 cases recorded by Chaudhary,[2] the youngest patient was a 9-year-old child.

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NEUROTIC EXCORIATIONS

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A 35-year-old female presented with a pruritic plaque on the dorsum of the left forearm. During follow-up it came to light that the lesion was self-inflicted and served as a means to seek help for the underlying psycho-social problem in the patient's life.

Key words : Neurotic excoriations.

Psychosomatic skin disorders are occasionally seen in the dermatology clinics. Amongst the psychosomatic skin disorders, neurotic excoriations and dermatitis artefacta are self-inflicted lesions.[1] In neurotic excoriations the repetitive self excoriations are usually initiated by an itch, or because of an urge to excoriate a benign irregularity on the skin.[2-4] One study reported an incidence of 2% of this disorder among the dermatology patients.[4] It predominantly affects the female sex with a peak age of onset between 30-45 years.[3] The lesions are found on the forearm and other accessible areas and are generally crusted or scarred with post-inflammatory hypo or eg hyper-pigmentation. Depression and suicide are the most common psychiatric complications.[3]

In this report we present one such case of neurotic excoriations.

Case Report

A 35-year-old female, developed a pruritic plaque on the dorsum of the left forearm. The lesion had been slowly increasing in size during 6 months. She had been to many doctors and prescribed various ointments but found no relief. The lesion was raised and circumscribed. It was about 4 cm in diameter with a—few papules in the central area of the lesion. A provisional diagnosis of lupus vulgaris was made and a biopsy was taken from the lesion. Histopathology study revealed it to be a keloid with no evidence to suggest lupus vulgaris. Intralesional corticosteroid on one occasion, had no discernible effect on the lesion. A few days later, the patient developed rectilinear deep excoriated lesions on the dorsum of the left forearm in close proximity to the pruritic plaque. These were similar to the marks made by a sharp instrument or a finger nail. At this juncture, the patient was referred to psychiatry for an evaluation.

The patient was married 15 years ago, but had separated from her husband two years later on grounds of mutual incompatibility. There was opposition from her parents and siblings to her separation. This prompted her to move to an Ashram against her parents' wishes. Both these had been a source of chronic stress to her.

Detailed enquiry revealed that the patient had been leading a reasonably well adjusted life at the Ashram, until about 6 months ago, when she had had a serious disagreement with one of her seniors. Following this, the patient felt distressed and sad. Around the same time, she developed small vesicles on the dorsum of the left forearm. In spite of various ointments and medications prescribed by doctors, these gradually increased in size in 6 months.

Repeated efforts to get a member from the Ashram or household proved futile as the patient steadfastedly refused to let us contact them. Hence, it was decided to keep the therapy focussed on her present problem and bring about symptomatic relief. After repeated interviews, the patient admitted to self inflicted skin lesions

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and attributed it to an irresistible urge to scratch the afflicted area. It was then pointed out to the patient, in a non-confrontational manner, that such self-injurious behaviour was probably related to the various stresses she had to cope with. After these issues were discussed she stopped inflicting fresh lesions. A month later, the patient was doing well and the skin lesion was healing. Subsequently, the patient did not report for follow up.

Comments

Self-injurious behaviour is often initiated by an itch[2] and the skin lesions are generally found in accessible areas like the forearm. Depression is often the most common psychiatric complication.[3] Our subject did complain of a depressed mood, but other concomitants of a depressive syndrome (sleep, appetite disturbance, crying spells and suicidal ideas) were absent and hence no medication was prescribed. Psychotherapy, which initially needs to be focussed on alleviating the symptoms, is generally effective. However, if the nature of these lesions go unrecognised, some cases may even progress to plastic surgery or, very rarely, amputations[5] because of severe disfigurement.

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ACQUIRED CUTIS LAXA

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A 40-year-old male started developing increased folds of skin on the face, 15 days after an attack of measles. He had entropion of the upper eye lids and ectropion of the lower eye lids, grade 2 piles and moderate anemia. There were no other systemic abnormalities.

Key words : Acquired, Cutis laxa.

Cutis laxa is characterised by increased laxity of skin. It is also termed as dermatolysis, dermatochalasia, cutis hyperelastica and cutis pendulata.[1] The condition can be congenital or acquired.[2] The congenital type manifests at an early age, it is extensive and often associated with severe involvement of the internal organs.[2-4] The acquired type is generally seen after some illness and it is associated with only mild to moderate involvement of the internal organs. A case of acquired cutis laxa following enteric fever has been reported from India.[1] We observed a case of cutis laxa occurring after an attack of measles.

Case Report

A 40-year male was in good health upto 30 years of age, when he developed moderate fever which was followed by an erythematous rash all over the body. There was mild pruritus. During the erythematous stage, the patient also had malaise, fever, myalgia, anorexia, conjunctival congestion and dry cough. The illness was diagnosed as measles. The patient was given oral tetracycline 500 mg three times daily and analgin 500 mg two times daily for seven days. With this the recovery was uneventful. Fifteen days later, he noted deepening of the naso-labial folds, laxity of ear lobules and burning pain in both the eyes which went on increasing gradually for the next five years. There was also involvement of the skin of the face and neck. Suspecting it to be leprosy, the patient was given dapsone 100 mg daily for 6 months, but it was omitted later due to lack of response. Entropion of the upper eye lids and ectropion of the lower eye lids resulted in burning in the eyes and spill-over of the tears. He also had bleeding piles for the last 6 months. The family history was negative. The patient had no addiction. There was no history of any major illness in the past. Patient's photograph at the age of 12 years (Fig. 1) revealed no abnormality of skin. Currently, there was increase in the skin folds on the face, neck and ear lobules (Fig. 2). There was minimal redundancy of the skin on the scalp, chest, abdomen and perineum. There was mild conjunctival congestion and excessive lacrimation. Proctoscopic examination revealed grade 2 piles. Hemogram showed moderate anemia and urine analysis was normal. X-ray chest, skull, intravenous pyelography and barium studies of gastro-intestinal tract revealed no abnormality. Histopathological examination of the skin from neck and face showed normal epidermis and collagen fibres in the dermis. The elastic fibres were scanty, fragmented and clumped. There were plenty of sebaceous glands. The findings were consistent with the diagnosis of cutis laxa.

Comments

Acquired cutis laxa generally starts at the age of 5 to 50 years and it closely resembles the congenital type clinically, morphologically and pathologically.[5,6] It has been reported in asso-

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Fig. 1. Clinical appearance at the age of 12 years.

ciation with vesicular eruptions,[2,3] persistent urticaria,[4,6] erythema multiforme,[2,6] eczematous dermatitis[7] and multiple myeloma.[8] It has occasionally been reported following typhoid fever[1] and operative trauma.[6] The exact etiopathogenesis of cutis laxa is not known though Zenker's degeneration in typhoid fever has been postulated.[1] Several systemic abnormalities are known in association with acquired cutis laxa. The common ones being emphysema,[2,3,5] tracheobronchomegaly and pneumothorax[4] and hernia.[2,5] The course of acquired cutis laxa is variable and the commonest cause of death has been congestive cardiac failure.[2,5] There is no drug to halt the progress of the disease.

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Fig. 2. Increased folds of skin on the face giving an appearance of senility.

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ATROPHODERMA VERMICULARIS

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Three siblings of a family developed atrophoderma vermicularis, presenting as itchy, keratotic follicular papules all over the body mainly on the extensor aspect of the extremities and multiple atrophic pits involving the face and atrophic scars on other areas of the body.

Key words : Folliculitis ulerythematosus reticulata, Keratosis pilaris atrophicans.

There is a group of rare inflammatory follicular atrophic conditions, closely related to keratosis pilaris.[1] Minor differences in the regional distribution and morphological changes have been used to differentiate various syndromes viz keratosis pilaris decalvans, keratosis pilaris atrophicans faciei and atrophoderma vermicularis.[2] We saw a child and his younger brother and sister affected by such a disorder.

Case Report

Twelve-year old male child had itchy, keratotic, follicular papules all over the body, multiple atrophic pits involving the face and atrophic scars on other areas of the body for the last 9 years. The lesions were worse during winter and would heal spontaneously in a few weeks time leaving behind atrophic pits. His sister aged 8 years and brother aged 5 years had similar lesions beginning at the age of about 3 years but the course of the disease till date was milder. There was no past history of exanthematous fever. None of the other family members had night blindness, atopy or similar lesions.

Examination revealed a moderately built and well-nourished boy with multiple, superficial, irregular, atrophic pits, 5 to 10 mm in length, 1 to 2 mm in width and about 1 mm in depth predominantly on the central part of the face involving the forehead, nose and cheeks (Fig. 1), the distribution being almost symmetrical. The affected areas felt slightly rougher and firmer than the uninvolved skin. There were multiple keratotic follicular papules distributed all over the body mainly on the extensor aspects of the extremities. Some of the lesions revealed tendency to grouping and coalescence while most of these were isolated. Some papules were linearly arranged particularly on the anterior aspect of the right leg. A few 0.5 to 1.5 cm size, atrophic scars were present on the nape of the neck and dorsum of the left hand. There was hyperkeratosis of the palms and soles. The scalp, eyebrows, nails and mucous membranes were normal. Examination of the eyes as well as other systems was unremarkable.

Fig. 1. Multiple, superficial, irregular, atrophic pits involving the face.

There was no evidence of keratosis pilaris in the parents. Routine investigations like blood

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Fig. 2. Thinned out epidermis with fibrosis and degeneration of the connective tissue (H and E ×100).

cell count, hemoglobin, sedimentation rate, liver function tests, urinalysis and stools examination were normal. Serum vitamin A and carotene levels were not done. Histopathological examination revealed thin to normal epidermis with atrophic or absent sebaceous glands (Fig. 2). There was fibrosis and degeneration of the connective tissue, with a chronic inflammatory cellular infiltrate. At places, a few hyperkeratotic, acanthotic and tortuous hair follicles were also seen.

Comments

Atrophoderma vermiculatum (syn folliculitis ulerythematosa reticulata) is a rare genodermatosis with autosomal transmission in most cases.[1] Since Unna in 1896 described for the first time this condition under the name ulerythema acneiforme, it has been reported under various names viz acne vermoulante, atrophoderma reticulata symmetrica faciei, folliculitis ulerythematosa reticulata, folliculitis atrophicans reticulata, atrophoderma vermicularis, atrophoderma vermiculatum, atrophoderma reticulata[3] and honeycomb atrophy.[4] This condition is characterized by the following clinical features : (1) appearance of the eruption in childhood with slow evolution, (2) symmetrical distribution of the lesions involving both cheeks, (3) erythema that may disappear later, (4) comedones or horny plugs that may disappear later, (5) reticulated atrophy that becomes less marked in adult life, and (6) absence of papules, pustules, crusts or hyperpigmentation.[5]

The condition could be confused with scarring due to acne vulgaris if the patients are first examined in adolescence, but its earlier onset is a distinguishing feature whereas in acne the scars are far deeper.

Familial occurrence with apparently dominant transmission, classical morphology of the lesions, almost symmetrical reticulate atrophy, milia and presence of keratosis pilaris along with compatible histopathological features as seen in our patient go in favour of the diagnosis of atrophoderma vermiculatum. The early onset of the disease, tendency of keratosis pilaris for Koebner phenomenon, bigger atrophic scars on areas other than the face were the unusual features in our cases.

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MENINGOCOCCAL SEPTICEMIA WITH CUTANEOUS VASCULITIS

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Two cases had meningococcal septicemia with vasculitis of skin. One 9-year-old female developed generalised, asymptomatic, purpuric, atrophic plaques on the limbs, trunk and face on the fourth day of high fever. The second patient was 20-year-old male who developed purplish plaques, nodules and bullae mainly on the extensor surfaces of the hands and feet on the tenth day of high fever. Bullae had clear fluid and bluish-black peripheral rim.

Key words : Meningococcemia, Skin lesions.

In fulminating infections with *Neisseria meningitidis*, fever is brief and the infection is divided into septicemic and meningitic stages. In the septicemic stage, purpuric plaques of variable sizes and number are characteristically seen on the trunk and limbs. Rarely, another type of a lesion in the form of solitary or multiple nodules and bullae may appear on the limbs, five to nine days after the onset of illness even if adequate antibiotic treatment has been given.[1]

Histopathology of petechiae demonstrates thrombi composed of neutrophils, platelets and fibrin in dermal vessels. Many meningococci are seen in luminal thrombi, within the vessel walls and around the vessels as Gram negative diplococci.[2] In addition, acute leukocytoclastic vasculitis with damage to the vessel wall and haemorrhages can occur.[3] Intra-epidermal and sub-epidermal pustules filled with neutrophils may also be seen.[4] Cutaneous lesions in the form of purpura, maculo-papular and faint-pink macules were reported in acute meningococcemia.[5]

Case Reports

Case 1

One 9-year-old female had high grade fever, headache, diarrhoea, vomiting and was diagnosed as a case of meningococcemia. On the fourth day of fever, the patient developed numerous, discrete, asymptomatic, purpuric plaques on the limbs, trunk and face (Fig. 1). Purpuric plaques had central atrophy and some had superficial gangrenous changes. Plaques appeared all over the body within one day and remained static for 18 days. Mucous membranes, hair and nails were normal.

Her haemoglobin was 11.5 gm %, TLC 8800 and DLC P74, L20, M3, E3%. Urine and stools were normal. Fasting blood sugar was 120 mg %. Serum creatinine was 1.2 mg %. Blood urea was 64 mg but returned to 40 mg after four days. ECG was normal. Serum sodium was 130 mEq and serum potassium was 3.5 mEq. CSF

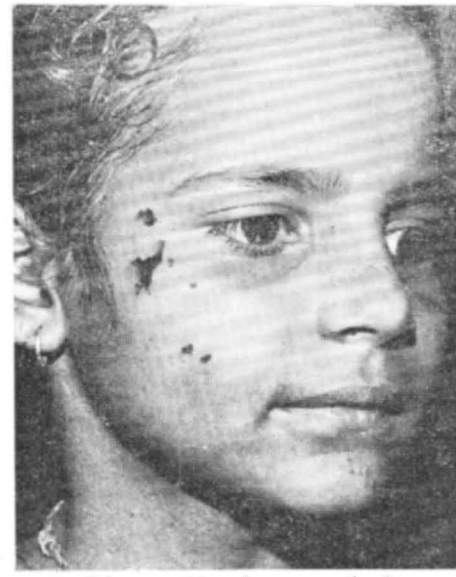

Fig. 1. Purplish atrophic plaques on the face.

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had proteins 600, globulins 4+, sugar 30 mg %, chloride 40 mEq and 30-40 pus cells/HPF. Polymorphs and a few lymphocytes were seen. No organisms were detected on Gram staining. CSF culture showed no growth of organisms. Biopsy of skin revealed meningococci and vasculitis consistent with the diagnosis of meningococcal septicemia.

Case 2

A 20-year-old male had high grade fever and vomiting and became unconscious on the third day of fever. He was diagnosed as a case of pyogenic meningitis and was treated with penicillin and gentamicin leading to marked improvement. On the tenth day of fever, the patient developed purpuric plaques, nodules and bullae mainly on the dorsum of hands and feet. Purpuric plaques varied from 1 to 1.25 cm in diameter and had central atrophy. Bullae had clear fluid with a peripheral bluish-black rim (Fig.2). A few bigger bullae in addition had central bluish-black discoloration and an angular outline. Black crusts covered a few plaques. Mucous membranes, nails and hair were normal.

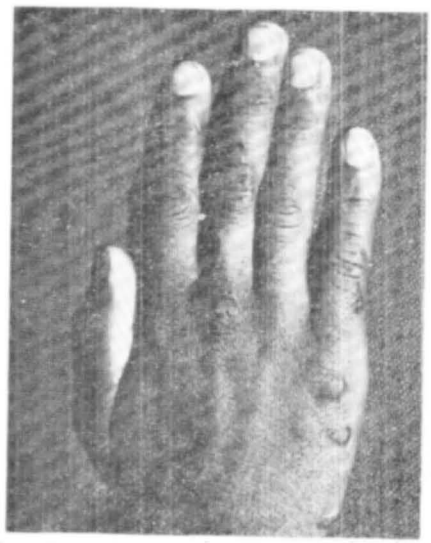

Fig. 2. Bullae with clear fluid in the centre and peripheral bluish-black rim.

His haemoglobin was 10.5 gm %, TLC 10,650/cmm and DLC P70, L24, M2 and E4%. CSF had proteins 450, globulins 4+, sugar 20 mg %, chloride 112 mEq and pus cells 4+. Polymorphs and a few lymphocytes were seen. No organism was seen on Gram stain. Biopsy report was vasculitis, intra-epidermal and sub-epidermal pustules filled with neutrophils. Diplococci were seen. Gram staining confirmed the presence of Gram negative diplococci. Culture of CSF and blood showed no growth of organisms.

Comments

Both patients had purpuric plaques and bullae typical for meningococcal septicemia. Whittle et al in 1973[6] also reported similar lesions. Typical histopathological changes were seen in the form of haemorrhage, leukocytoclastic vasculitis and thrombi in the dermal vessels. Gram negative diplococci were present in and around the blood vessels. In addition, intra-epidermal and sub-epidermal pustules filled with neutrophils were also seen. Greenwood et al[7] could not locate meningococci but detected meningococcal antigen on immunofluorescent staining in cases of meningococcal septicaemia. Presence of circulating antibodies and marked fall of C3 levels led them to conclude that arthritis and cutaneous lesions in their cases could be due to immune complex formation or Arthus phenomenon. Meningococcal toxins and endotoxins were responsible for micro-thrombi and vasculitis in meningococcal septicemia.[2]

The cutaneous lesions in acute meningococcemia probably result from direct damage to capillaries and post-capillary venules. Organisms are found in swollen endothelial cells, in polymorphonuclear leukocytes in the inflammatory reaction, and trapped in fibrin thrombi in the vessels. These features are also consistent with the dermal Schwartzman reaction, but the finding of immunoglobulins and complement in

the walls of involved vessels argues for an immunological active process as well.[8]

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COEXISTENT BULLOUS DARIER'S DISEASE WITH XERODERMA PIGMENTOSUM

Radha Mittal and Samarjit Sandhu

A case of bullous Darier's disease had typical brown follicular papules, greasy crusted papules, discrete keratotic papules and vesiculo-bullous lesions on the dorsum of the hands and feet. In addition, characteristic lesions of xeroderma pigmentosum as extensive freckles intermingled with atrophic white spots were present on the face, trunk and limbs. This combination of bullous Darier's disease with xeroderma pigmentosum has not been reported earlier.

Key words : Darier's disease, Bullous, Xeroderma pigmentosum.

Darier's disease and xeroderma pigmentosum are two rare distinct diseases. Darier's disease is determined by autosomal dominant gene or it may manifest due to gene mutation.[1] Vesiculo-bullous lesions as a bullous variant were described by Jablonska and Chorzelski in 1958.[2] In some cases of Darier's disease, there are keratotic papules on the dorsa of hands and feet that resemble clinically those seen in acrokeratosis verruciformis of Hopf.[3] Histopathologically, these papules may show dyskeratosis and lacunae as seen in Darier's disease or changes characteristic of acrokeratosis verruciformis of Hopf.[4] Acrokeratosis verruciformis of Hopf is an independent entity as it is distinct both genetically and histopathologically from Darier's disease, suprabasal clefting being found only in the Darier type papules.[3,5]

Case Report

A 20-year male had photosensitivity, giddiness and weakness on exposure to sunlight for the last 6 years. Five years back, the patient developed greasy, crusted, follicular papules and scales on the scalp and a few crusted, yellow-brown papules on the seborrhoeic sites such as axillae, groins, naso-labial folds, ears, retro-auricular regions, buttocks, natal cleft and lumbo-sacral region. Simultaneously, he also developed discrete, scaly, 1 to 1.5 cm diameter papules on the dorsum of hands and feet and 1 to 3 mm freckles on the face which spread to the upper limbs, lower limbs and trunk. Four-and-a-half years back, atrophic white spots also appeared. The disease spread more rapidly during summers when vesicles and bullae with secondary infection also appeared mainly on the dorsa of hands and feet. Skin of the palms and soles was thick and waxy. One 2.5 cm diameter cafe au lait spot was present near the umbilicus. The face and neck had diffuse pigmentation. Both eyes had pterygium and left eye had one bitot spot. Hair and nails were normal.

Biopsy from a freckle on the face revealed hyperkeratosis, melanin accumulation in the basal cell layer and upper dermis and mononuclear infiltrate in the dermis. Second biopsy from a follicular papule in the axilla revealed hyperkeratosis, papillomatosis, numerous lacunae with acantholytic cells, corps ronds and grains. Dermis showed oedema and mononuclear cell infiltrate (Fig. 1). Third biopsy from a papule on the hand showed marked hyperkeratosis, grains, irregular hypergranulosis, acanthosis, lacunae, dyskeratosis, corps ronds, mid-epidermal and suprabasilar cleft. Suprabasal cleft, lacunae, corps ronds and dyskeratosis were seen under high power in the section

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