HEREDITARY SENSORY AND AUTONOMIC NEUROPATHY TYPE II

To the Editor,

Hereditary sensory and autonomic neuropathies (HSAN) are rare disorders and 5 types can be distinguished.[1] HSAN type II is congenital with recessive inheritance and patients have pansensory neuropathy affecting limbs more than trunk or face. Painless paronychia, whitlows, ulcers of hands and feet leading to mutilating acropathy associated occasionally with reduction in sweating is characteristic of HSAN type II. Sfural and cutaneous nerves are abnormally small and contain few or no myelinated fibres and reduced number of unmyelinated fibres.[1] In congenital indifference to pain, neuropathies are absent and there is loss of perception of pain, other sensory perceptions being unaffected.[2] Hereditary plantar ulcers are seen in HSAN type I and dysautonomia is prominent in HSAN type III.[3]

A 10-year-old boy had multiple bullae and scars on both upper and lower limbs associated with prominent mutilating deformity of hands and feet since early childhood. He was product of a non-consanguinous marriage, full term normal pregnancy and vaginal delivery. Family history of similar disease was negative. He had low intelligent quotient, all milestones and dentition were delayed. He had no feeding difficulty although he suffered from frequent episodes of diarrhoea during infancy and early childhood. Bullae on hands, feet, lower legs and forearms appeared without accompanying pain or pruritus since early childhood. Bullae ruptured with resultant painless ulcers, superimposed secondary infections, rarely osteomyelitis and healed with pigmented scars, contractures, loss of digits and gross mutilating deformity of hands and feet. A trophic ulcer was seen on the plantar surface of left big toe. Loss of sensation of touch, temperature and pain were upto upper part of knees and elbow joints. cranial nerves, peripheral cutaneous nerves, corneal reflexes were normal and pupils reacted to light and accommodation normally. Bilateral cataracts and nystagmus were seen. He was anaemic, poorly nourished and had short stature, pigeon chest deformity, rickety rosary, slight flexion deformity of elbows, painless swelling of ankles, knees and some small joints of hands and feet. Pharynx, larynx, smelling and hearing powers were normal. Parasternal heave, accentuation of first heart sound in mitral area and mid-diastolic murmur were present. Haemoglobin was 8 gm%. X-ray of feet revealed absence of distal phalanges of both fifth toes, absence of proximal and distal phalanges of right great toe, resorption of the proximal phalynx left great toe with bony fragments of other left phalanges. Skin smears were negative and biopsy was normal. Bullae and ulcers healed rapidly with antibiotics.

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PROBABLE MECHANISM OF VARICELLA-INDUCED KOEBNERIZATION IN PSORIASIS

To the Editor,

Koebner's or isomorphic phenomenon is commonly observed in psoriasis due to several