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

triggering factors such as physical injury, sunburn, x-radiation, chemical and cytotoxic agents, and infections (bacterial, fungal and viral).[1,2].

A 25-year-old man presented initially with fever and vesiculo-pustular exanthem which was diagnosed as varicella by Tzanck smear. Two weeks later, resolving of lesions was accompanied by numerous small, guttate to coin-shaped erythematosquamous lesions involving face, chest abdomen and extremities. Auspitz sign was positive. Nail changes included oil pits. Biopsy confirmed the diagnosis of psoriasis. Patient had psoriasis at the age of 10 years, which was successfully treated and remained dormant for several years before this episode. The present episode responded favourably to a combination of UVB therapy, oral antibiotics (Erythromycin 1 gm/day) and local steroid ointment (mometasone furoate ointment 0.1% once daily).

The Koebner's or isomorphic phenomenon is a reactive state in many diseases; the exact mechanism of which remains elusive. A special feature of psoriasis is the capacity to reproduce skin lesions at the site of endogenous or exogenous insult. Infrequently, measles, chicken pox and herpes zoster have been described as trigger factores.[2,3]

Several theories have been put forward to explain Koebnerization in psoriasis. In rubeola and varicella, isomorphic response may be a result of fever and endogenous pyrogens.[3] Neurogenic inflammatory mediators such as Substance P has been suggested as the triggering mechanism. Moreover, potentiation of Substance P by viral infection, further supporting this hypothesis, has been put forward.[3] However, more recent studies show that Substance P does not stimulated epidermopoisis of psoriatic keratinocytes in vitro.[4]

Recently, bacterial and fungal superantigens have been implicated in the guttate lesions of psoriasis.[5,6]

The Staphylococcus aureus superantigens activates T-cells without the help of antigen presenting cells. This is through T-cell receptor variable (VB) beta gene expression.[5-7] There is direct binding of these activated T-cells with major histocompatibility HLA class II molecules culminating in T-cell response. It is likely, that superantigens may also be the underlying mechanism of reactivation of psoriasis after viral infections with measles and varicella.

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