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INDETERMINATE HANSEN'S DISEASE IN A PATIENT WITH HEREDITARY SENSORY AND AUTONOMIC NEUROPATHY

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Hereditary sensory and autonomic neuropathy (HSAN) is a group of rare hereditary diseases. A young female with Type I HSAN with indeterminate Hansen's disease (HD) is reported which is a rare association.

Key Words : Hereditary sensory and autonomic neuropathy, Hansen's disease

Introduction

HSAN is a group of inherited diseases characterised by neuronal atrophy and degeneration predominantly affecting the peripheral sensory neurons. In 1994 Dyck classified HSAN into 5 types.[1] The World Federation of Neurology classified HSAN into 7 types.[2] Type I is autosomal dominantly inherited disease affecting second or third decade of life. The pathological changes are degeneration of the dorsal root ganglia, which is most severe in first and second sacral and last two lumbar segments. Degeneration of posterior column of spinal cord and axonal degeneration of peripheral nerves are also noted. The sensations of pain and temperature are lost bilaterally on the lower extremities. The tactile sensation also is impaired. Most of the patients present with callositis and ulceration of foot. The upper limbs are usually not affected. There will be no tenderness or thickening of nerves, which comes in the differential diagnosis of neuritic Hansen's disease.[3]

The association of Hansen's disease and HSAN is extremely rare. We are reporting a case of histologically proved indeterminate Hansen's disease (HD-I) in a patient with HSAN type-I with a family background.

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Case Report

An 18-year-old girl born of a full term normal delivery to second degree consanguineous parentage, presented with recurrent painless ulcers of both feet of 4 years duration (Fig.1). The ulcers healed with

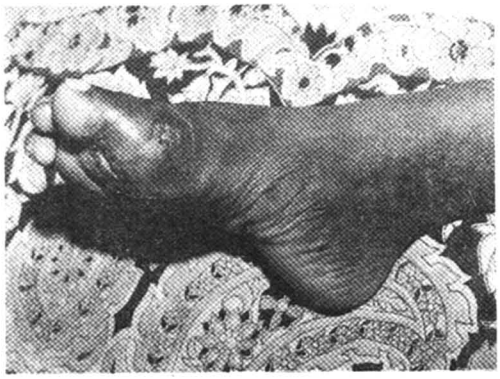

Fig. 1. Right foot of patient showing deep ulcers on base of big toe on plantar aspect and medical border.

scarring. No other skin lesions were noted during this period. She noticed a hypopigmented ill-defined patch on her upper back over the inter-scapular area extending to right scapular area with mild sensory blunting of about 4 months duration (Fig.2).

There was complete loss of all modalities of sensation below knees bilaterally. All peripheral nerves were not enlarged and nontender. All deep tendon

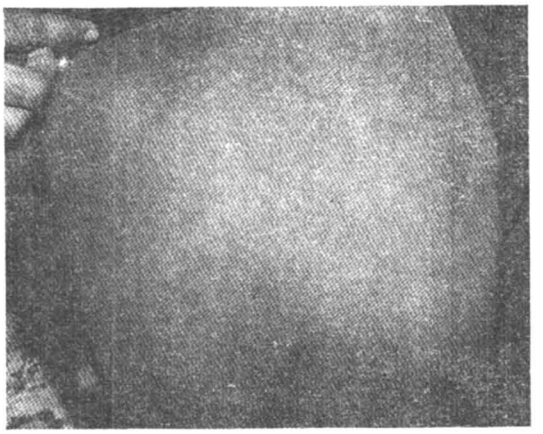

Fig. 2. Ill-defined hypopigmented patch on interscapular area extending to right scapular area.

reflexes were present except ankle jerk, which was absent bilaterally. The hypopigmented patch had ill-defined borders and a size of 16x14 cm, on the interscapular area with mild impairment of sensation, extending to the right scapular area.

All baseline haematological and urine examinations were within normal limits including a blood VDRL. Ear lobe smear and slit and scrape smear from hypopigmented patch were negative for acid fast bacilli (AFB). Biopsy of the skin lesion showed periappendageal lymphocytic infiltrate (Fig. 3)

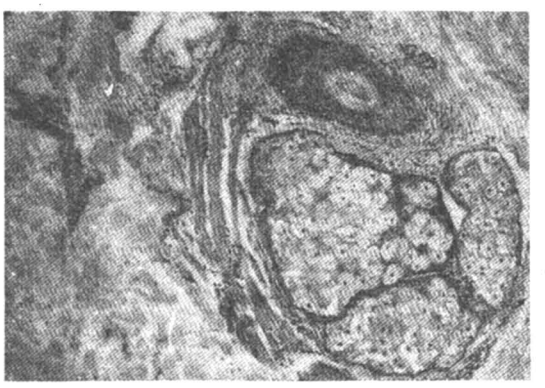

Fig. 3. Histopathology of the patch showing periappendageal lymphocytic infiltrate (H&E x 40).

with demonstrable AFB consistent with indeterminate HD. Nerve conduction study showed severe axonopathy of common peroneal nerves with decreased sensory conduction. Left sural nerve biopsy showed no granuloma or AFB, but the unmyelinated fibres were almost absent. Roentgenogram of right foot showed a neuropathic foot.

Her 48-year-old father also gave history of similar painless ulcers of feet of long duration. Multiple scars were seen on his feet with deformity of right big toe. There were no thickened and tender nerves. All other family members were normal.

Discussion

Symmetrical involvement of both legs and feet of long duration, absence of nerve thickening and absence of granuloma or AFB on nerve biopsy, family history of similar illness and radiological and nerve conduction study results made us to consider the diagnosis of HSAN type I. The associated hypopigmented patch which is of recent onset is proved to be Hansen's disease (indeterminate type) histopathologically. Such an association is reported very rarely. HSAN type I itself can be misdiagnosed as neuritic Hansen's disease.

References

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