Ind J Dermatol Venereol Leprol 1994; 60 : 335-6

SHORT COMMUNICATIONS

HEREDITARY SENSORY NEUROPATHY

S Mishra, P C Singh

Three cases of hereditary sensory neuropathy are reported. Two of them belong to same parents with family history of this disease.

Key Words : Sensory neuropathy, Acro-osteolysis, Leprosy

Introduction

Hereditary Sensory Neuropathy - also known as Thevenard Syndrome, Familial acro-osteolysis, is a rare familial disorder characterised by appearance in childhood or early adult life of analgesia of lower limbs with neuropathic sequelae and often associated with nerve deafness. The process is purely osteolytic without any evidence at bone regeneration which gradually extends proximally and eventually involves other acral bones.[1] Shortening of foot is characteristic.

Case Report

Two children (Case - I, II) one sister and one brother, of same family were seen with recurrent ulceration and gradual mutilation of feet. Their parents were normal, but their late paternal grandmother also suffered from same type of disease of both feet. They are the only two children of their parents. Case - III, not related to Case - I, II, presented with recurrent ulcer and shortening of right foot.

Case I

The eldest child, a girl of 15 years of age, presented with non-healing ulcers under great toe and second toe of right foot and shortening of right great toe. She is having such recurrent ulcers on right foot since she was eight years old. Left foot and upper extremities are normal. On examination of right foot there is an ulcer 1.5 cm x 1 cm size on plantar aspect of great toe. Another ulcer 1 cm x 0.5 cm was on plantar aspect of second toe. The ulcers were painless. Sensations of pain, temperature and touch were completely lost upto ankle joint. Motor functions including reflexes were normal. There was no wasting of muscles. There was no nerve deafness. Sensations and motor functions in left lower limb and upper extremities were normal. There was no hypopigmented or erythematous patch on body. Peripheral nerves were not thickened, not tender. Lacrimation and sweating were normal. Systemic examination did not reveal any disease.

Routine blood, stool, urine examination were normal. FBS 80 mg / dl, blood VDRL test - nonreactive. Nerve conduction, nerve biopsy and muscle biopsy could not be done due to objection of the patient. Slit skin smear for AFB from ear lobes, right leg was negative. X'ray right foot showed irregular destruction around metatarso-phalangeal joint area of 1 and 2 toe. X'ray left foot was normal.

Case II

The younger child, 13 years old boy, developed spontaneous ulcers on sole of left foot when he was six years old. Gradually

From the Department of Skin & VD, VSS Medical College, Burla, Sambalpur (Orissa) - 768 017 India.

Address correspondence to : Dr P C Singh

nall ulcers developed on toes of both feet. 'here was no loss of digits but there were ealed scars and shortening of toes with eformity of both feet. Upper extremities were ormal.

General physical examination, higher inctions, cranial nerves were normal. There as no hypopigmented or erythematous patch n body. Peripheral nerves were normal. ensation of touch, pain and temperature ere lost in both feet. There are ulcers on lantar aspect of toes of both feet with ıortening of both the great toes. Systemic xamination did not reveal any disease. outine blood, stool and urine examination ere normal. Slit skin smear for AFB from arlobes, both legs-negative. Nerve condution, erve biopsy could not be done as in first case. 'ray - irregular destruction of phalanges and stal part of metatarsal bones of both feet.

ase III

Married lady of 35 years of age, having vo children, presented with recurrent ulcers ı right foot. Her children were normal. There no family history of such disease. She is aving such ulcers since last ten years. On xamination, left leg, and upper extremities e normal. There is no clinical evidence of prosy as in other two cases. Sensation of ain, touch and temperature were lost in right ot upto ankle joint. There were 3 ulcers on anter aspect of right foot near metatarso-alangeal joints with shortening of the foot.

Routine blood, stool and urine xamination were normal. FBS 80 mg / dl. DRL test non-reactive. Nerve condution, erve biopsy could not be done due to ojection of the patient. X'ray right foot vealed osteolytic changes in phalanges and stal part of metatarsal bones.

Discussion

Familial acro-osteolysis have been described by Smith[2] and Heller et al.[3] Behera et al[4] reported two brothers in a family having recurrent ulcer and mutilation feet. In the elder one there was autoamputation of right foot upto the ankle joint. Gurvinder et al[5] reported 6 cases of familial acro-osteolysis in 15 members of 3 generations of a family. The inheritance pattern was autosomal dominant.

The exact aetiology is unknown. The basis is a hereditary degeneration of certain of the craniospinal ganglion. The sensory neurons that connect centrally with spinothalamic tract are particularly affected. Cheney[6] postulated a process of vascular bone resorption associated with ingrowing of capillaries, the bone being resorbed ahead of advancing vessels producing ischaemic necrosis of bone.

Thevenard syndrome is to be differentiated from leprosy. Many cases are misdiagnosed as leprosy and bear a social stigma. It is also to be differentiated from congenital absence of pain where sensations other than pain are well appreciated.

References

1. Thevenard A L. Acropathic Ulcero-mutilante familiala. Acta Neurol Belg 1953; 52 : 1-24.
2. Smith E M. Familial neuropathic oesseous atrophy. JAMA 1934; 102 : 593.
3. Heller I H, Rebb P. Hereditary sensory neuropathy. Neurology 1955; 5 : 15-29.
4. Behera D K, Prusty P, Mishra S, Rath P C. Hereditary sensory neuropathy. The Journal of Orissa Orthopaedic Association 1989; 11 : 33-6.
5. Gurvinder P Thami, Sharma S C, Bhargava S K, Singal A, Gautam R K, Baruah M C. Familial acro - osteolysis through three generations. Ind J Dermatol Venereol Leprol 1993; 59 : 284-7.
6. Cheney W D. Acro - osteolysis. Am J Roentgen 1965; 94 : 595.