Osteoarthropathy in hereditary sensory radicular neuropathy

A case report

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A young man had hereditary sensory radicular neuropathy with relapsing ulcer of the foot and, in addition to previously known clinical features, osteoarthropathy with hallux valgus, metatarsus primus varus, exostosis, and pes planus.

Hereditary sensory radicular neuropathy is characterized by progressive, distal–to–proximal loss of sensation in the extremities due to degeneration of dorsal root ganglia. The condition may be hereditary, familial, or sporadic (Denny–Brown 1951, Shahriaree et al. 1979), and typically entails forefoot hyperkeratosis with fissures and recurrent ulcers leading to osteomyelitis.

We report a case of hereditary sensory radicular neuropathy with associated clinical features, notably osteoarthropathy, that has previously not been reported.

Case report

A 17-year-old male was referred to our unit because of relapsing ulcer of the forefoot. At the age of 11 years, the patient sustained lacerations of both feet. Because the wounds caused no pain, the patient failed to take proper care. An ulcer of the left foot recurred repeatedly, and osteomyelitis developed in the distal phalanx of the big toe. After curettage and nail removal, the ulcer healed for a short time, but then relapsed. For the last 2 years, an ulcer on the metatarsal weight-bearing area of the right sole had increased in size without responding to conservative treatment (Figure 1 A). The family history revealed a mild autosomal recessive pattern of hereditary sensory radicular neuropathy.

The patient had bilateral pes planus, a deformed right foot with hallux valgus and metatarsus primus varus (Figure 1 A and B), and a deformed left ankle joint. There was loss of touch, pain, and temperature sensation in both lower legs and feet (Figure 1 E). Thermoanesthesia was more extensive than analgesia, and analgesia was more extensive than anesthesia. Sensation in the hands was also affected in a similar manner (Figure 1 E). The plantar reflex in both feet was absent, and the ankle jerk of both legs was weak. The knee jerk was present, and there was no muscle wasting; muscle power and tone were normal.

All the values of routine laboratory tests were within normal limits. A roentgenogram of the left ankle showed Charcot's lesions (Figure 1 D), and the right foot showed hallux valgus, metatarsus primus varus, abnormal hypertrophy, and exostosis of the first metatarsal bone (Figure 1 C). Myelography revealed no abnormality. Motor-nerve conduction tests were normal. Sensory conduction was not recordable. Histopathologic examination of the right medial plantar nerve showed mild degeneration of fibers, and normal skin from the leg showed a decrease in nerve fibers. The ulcer itself was 2.5 cm in diameter with surrounding hyperkeratotic tissues (Figure 1 A). A biopsy specimen of the ulcer showed no changes.

The exposed flexor-tendon sheath of the right second toe was excised and necrotic tissue was debrid-

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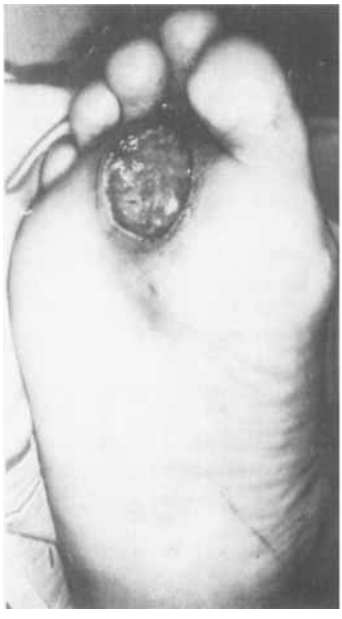
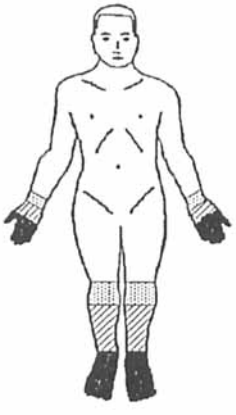


A. Ulcer, deformed great toe, and pes planus.

B. Four months after resurfacing. Deformity of right great toe is more conspicuous.

C. Hallux valgus, metatarsus primus varus and exostosis, hypertrophy and shortening of first metatarsal.

D. Left ankle with Charcot's changes.



E. Sensory chart. Dots represent temperature loss, oblique lines represent pain loss, and black color represent touch loss.

Figure 1. A 17-year-old male with hereditary sensory radicular neuropathy.

ed. The wound was then resurfaced with a plantar flap based on the lateral plantar artery. Postoperatively, the patient was kept in nonweight-bearing positions for 2 months and advised to restrict his activities and to take meticulous care of the foot. After 1.5 years, hyperkeratosis again developed in the same area. The hyperkeratosis was subsequently excised, and the area was covered with a rotational flap. Due to residual hyperkeratosis, the ulcer relapsed after 1 year. A free medial plantar fasciocutaneous flap from the left foot was transferred to cover the defect. The patient has now been free from his ulcer and hyperkeratosis for 10 months.

Discussion

In 1922, Hicks described hereditary ulcers of the foot. In 1951, Denny-Brown examined one of Hicks' original patients clinically and at necropsy. There was degeneration of dorsal root ganglia, and the condition was named hereditary sensory radicular neuropathy. Syringomyelia and syringobulbia were eliminated by myelography and laboratory tests. In congenital sensory neuropathy, absence of pain sensation is present from birth, but sensation improves with age (Winkelmann et al. 1962). In sensory radicular neuropathy, the onset is usually in ad-

olescence and sensory loss is progressive in nature (Denny-Brown 1951, Shahriaree et al. 1979).

Several cases have had peroneal muscular wasting (Campbell and Hoffman 1964), optic disc atrophy (Turkington and Stifel 1966), progressive deafness (Denny–Brown 1951, Munro 1956, Pallis and Schneeweiss 1962), and periosteal elevation and thickening of long bones (Minton et al. 1973).

In sporadic forms, deafness is usually absent (Denny-Brown 1951, Turkington and Stifel 1966). In 1966, Turkington and Stifel reported optic disc atrophy in sensory radicular neuropathy. Campbell and Hoffman in 1964, reported peroneal muscle wasting in hereditary sensory radicular neuropathy. In our patient, muscle wasting, deafness, and optic disc atrophy were absent.

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