

CASE 1996 [5]

Submitted by: Hans H. Goebel, Division of Neuropathology, Mainz University Medical Center,
Langenbeckstrasse 1, D-55131 Mainz, Germany

Clinical History:

This 63-year-old woman had impaired sensitivity to touch and absent sensitivity to pain on the left leg, the first fingers of her right hand and her right jaw as long as she could remember. She also had impaired hearing on the left which progressed over the years. When 8 years old she experienced diphtheria which subsided without residual symptoms. When 55 years old, she had surgery of her stomach owing to recurrent ulcers and an amputation of her right second toe. A paternal aunt and grandmother also had hypacusis on the left, but no relative had any sensory deficits. Both her daughters are healthy at the ages of 37 and 38 years, respectively. Since the age of 52 years she had recurrent corneal ulcers resulting in decreased vision and bilateral corneal transplants at the age of 63 years.

Neurological examination at the age of 63 years revealed corneal ulcers bilaterally with respective impaired vision, hypesthesia and hypalgesia of her oral cavity on the right and of her right nasal cavity. She had left-sided hypacusis and generalized lack of deep tendon reflexes, no pyramidal signs. She had hypesthesia and hypalgesia of her first and second fingers on the right, of her entire left leg, the circular border at the groin level. Vibration sense was normal over her wrists and right ankle, but reduced to 2/8 over her left ankle. Position sense of her left leg was impaired, and she had an ulcer of her left foot sole. Her left peroneal muscles were reduced to $\frac{3}{4}$ with a circumference of 29 cm compared to the equivalent of the right lower limb. She showed mild ataxia when standing or walking with closed eyes. Blink reflex was impaired bilaterally. Somato-sensory potentials over the right median nerve and of the left tibial nerve were normal. Motor nerve conduction velocities were normal, but amplitudes from the left leg and the right arm were reduced. No sensory potentials could be elicited from her right ulnar and median nerves as well as from her left sural nerve.

A full size biopsy of her right sural nerve at ankle level was performed at the age of 63 years.

Material submitted:

1 plastic-embedded section of biopsied sural nerve, stained with toluidine-blue.