Hereditary Neuropathy with liability to Pressure Palsies-HNPP

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Abstract

Hereditary neuropathy with liability to pressure palsies (HNPP) or « tomaculous » neuropathy is a peripheral neurologic disorder of the young adult clinicaly characterized by repeated episodes of isolated peripheral palsies, which are easily provoked by minor traction or local pressure on the peripheral nerves (fall-hand, claw-hand or foot-drop).

The first episode is most often observed between 4 and 25 years of age. Spontaneous complete recovery in weeks or months is usual.

The trait is passed on a dominant mode of inheritance with high penetrance ; the genetic anomaly is a large deletion (1,5 megabase) of PMP 22 (peripheral myelin protein) gene.

Electrophysiology shows a diffuse decrease in nervous sensitive conduction and a lengthening in motor distal latency.

Sural nerves shows multiple bubbles (lat : tomaculum) in the myelin sheat and thickening of some internodal segments.

HNPP incidence is higher than expected formerly.

In our personal series of 1465 releases of peripheral nerve entrapments, we observed only one HNPP, recently diagnosed although the first episode (left median nerve) was observed in 1980 when she was eleven years old. Within the next two years, she presented plasy episodes on four different peripheral nerves, electromyography was positive in only one episode.

At that time, ignorant of the HNPP pathology, we released various entrapments with complete clinical and electrophysiologic recovery.

Twenty years later, she presents again with a right ulnar nerve palsy with discrete dicrease in nerve conduction. When this palsy was spontaneously recovering, she developed a homolateral radial palsy.

DNA analysis by *southern blot* and *FISH* confirmed the diagnosis of HNPP which must be kept in mind when young patient presents repeated episodes of isolated peripheral nerve palsies.

Differential diagnosis is made with Charcot-Marie-Tooth type 1A neuropathy or MEN (multiple entrapment neuropathy).