Peripheral neuropathy in Whipple's disease: A case report

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SUMMARY
Whipple's disease is a chronic multisystem inflammatory disease with predominantly gastrointestinal manifestations due to *Tropheryma whipplei* infection. Typical neurological abnormalities include dementia, eye movement abnormalities, hypothalamic dysfunction and oculomasticatory myorhythmias. The literature on peripheral neuropathy in Whipple's disease is sparse and the involvement of peripheral nerves in Whipple's disease has not been documented convincingly so far.
We present a case of Whipple's disease presenting by axonal peripheral neuropathy without gastrointestinal involvement. The diagnosis was confirmed by a sural nerve biopsy and consequent PCR of the sample. All clinical signs disappeared progressively during the antibiotic therapy. Two years after the *T. whipplei* infection, the patient developed dopa-sensitive Parkinson's disease, although these two events seem to be unrelated.
This case illustrates the value of peripheral nerve biopsy in cases of axonal neuropathy of unexplained origin and extends the clinical spectrum of Whipple's disease to a new modality.

Keywords: Whipple's disease – peripheral nerve – axonal neuropathy – polymerase chain reaction – electron microscopy

Periferní neuropatie u Whippleovy choroby: Popis případu

SOUHRN
Whippleova choroba je mutlisystémové onemocnění s převážně gastrointestinální manifestací, způsobné infekcí *Tropheryma whipplei*. Mezi typické neurologické poruchy patří u této choroby demence, okohybné poruchy, hypothalamická dysfunkce a okulomastikatorní myorytmie. Postižení periferního nervu zatím nebylo u této choroby přesvědčivě zdokumentováno.
Popisujeme případ Whippleovy choroby, která se projevila periferní neuropatií axonálního typu bez postižení zažívacího traktu. Diagnóza byla potvrzena biopsií periferního nervu s následnou konfirmací pomocí PCR. Příznaky neuropatie vymizely během terapie antibiotiky. Dva roky poté se u pacienta rozvinula dopa-senzitivní Parkinsonova choroba; tyto dvě události se ale zdají být bez souvislosti.
Náš případ dokumentuje přínos biopsie periferního nervu v případech axonálních neuropatií neznámé etiologie a rozšiřuje klinické spektrum symptomatologie Whippleovy choroby.

Klíčová slova: Whippleova choroba – periferní nerv – axonální neuropatie – PCR – elektronová mikroskopie

Cesk Patol 2012; 48(2): 97–99

Additional video file is available at www.CSpatologie.cz

Whipple's disease (WD) is a rare chronic multisystem infection caused by *Tropheryma whipplei* (1). Most common presentations include fatigue, profound weight loss and various gastrointestinal signs from constipation to diarrhea. Fever, arthralgias and peripheral lymphadenopathy are also frequent. The clinical picture is often insidious and non-specific, which prolongs the time between the first signs and the diagnosis. Neurological manifestations of WD (Neuro-Whipple) are rare and usually central with cognitive impairment, supranuclear ophthalmoplegia, psychiatric features, oculomasticatory myorhythmias, and hypothalamic dysfunction (2–4). The involvement of peripheral nerves in Whipple's disease is extremely rare and it has not been documented convincingly in the literature so far.

Here we present the first case of WD presenting by peripheral neuropathy without GIT involvement, confirmed by sural nerve biopsy and consequent PCR of the sample.

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CASE REPORT

A 64-year old man with a history of arterial hypertension and mild depression developed over 6 months progressive, involuntary buccolingual and palpebral movements associated with increasing fatigability and nocturnal leg paresthesias. The patient did not complain about any gastrointestinal disturbances and he had no weight loss for the given period.