in cisplatin treated mice than in untreated controls, and significantly ameliorated by PEA treatment ($p<0.001$). Mice treated with cisplatin progressively developed mechanical hyperalgesia that was partially but significantly reversed by PEA treatment ($p<0.05$ from −23.4% to −10.7%). Similarly, cisplatin caused a significant decrease (−15%) of caudal and digital nerve amplitude and NCVs compared to controls that was non-significantly prevented by PEA treatment. Our findings suggest that PEA can exert some beneficial effects on thermal sensitivity and mechanical allodynia in cisplatin-induced CIPN but did not lead to a significant improvement of nerve amplitude and NCV, indicating that treatment could not protect from nerve damage.

L- SERINE SUPPLEMENTATION SUPPRESSES THE FORMATION OF NEUROTOXIC DEOXYSPHINGOLIPIDS AND IMPROVES NEUROPATHY IN A TYPE 1 DIABETIC RAT MODEL

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1-Deoxysphingolipds (dSL) are atypical sphingolipids which are formed by the enzyme serine-palmitoyltransferase (SPT) due to a promiscuous use of L-alanine over its canonical substrate L-serine. 1-deoxy-sphingolipids lack the C1 hydroxyl group of normal sphingolipids and are therefore not converted into complex sphingolipids nor degraded by the normal catabolic pathway. They are neurotoxic and induce neurite retraction in cultured primary neurons. Pathologically increased dSL levels are the underlying cause for the inherited axonal neuropathy HSAN1 which is associated with several missense mutations in SPT. Plasma dSL levels are significantly elevated in patients with the metabolic syndrome and diabetes which suggests that increased dSL formation could also be involved in the pathology of the diabetic sensory neuropathy (DSN). The formation of dSLs is markedly suppressed at increased L-serine levels. An oral L-serine supplementation resulted in significantly decreased dSL formation as it was demonstrated in mouse models for HSAN1 and HSAN1 patients. Under serine supplementation HSAN1 mice showed normal dSL levels and did not develop neuropathic symptoms. We studied the effect of serine supplementation on the prevention and treatment of diabetic neuropathy in a streptozotocin (STZ)-induced diabetic rat model. Diabetic and non-diabetic rats were supplemented for 16 weeks with a serine enriched diet. STZ injected rats developed substantial hyperglycaemia (400–700 mg/dl) and no gain in body weight whereas control animals had normal blood glucose and doubled their body weight within the test period. Serine supplementation had no impact on blood glucose and body weight. Plasma dSL levels were highly increased in diabetic rats but not different to controls in the L-serine supplemented rats. Regular sphingolipid levels were the same in all groups. After 16 weeks of supplementation mechanical sensitivity, NCV and neuronal Na^+,K^+-ATPase activity was significantly better in serine treated vs. non treated diabetic rats. Thermal sensitivity was not different between diabetic and control animals. All animals showed a highly significant negative correlation between plasma dSL levels and NCV ($p = 5.2E-12$). Besides improvements in the neuropathy, we also observed significantly less cataracts in the serine treated animals. In summary, our data support the hypothesis that increased dSL formation is involved in DSN and that suppression of these lipids by simple and inexpensive serine supplementation could be a novel therapeutic option to treat DSN.

MRI EVIDENCE OF TRIGEMINAL SENSORY NEUROPATHY IN SJÖGREN'S SYNDROME

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Sensory ganglionopathy is a rare form of neuropathy that may complicate Sjögren's syndrome. It results from lesions of the dorsal root ganglia, and spinal MRI may show posterior column T2-weighted hyperintensity, expression of the retrograde degeneration of the large afferent fibers in the posterior column. Clinically it presents with sensory ataxia, and sometimes upper limb choreoathetotic movements. Cranial nerves may occasionally be involved, with trigeminal nerve being the most commonly affected. We describe a 66-year-old woman with Sjögren's syndrome with sensory ganglionopathy and severe trigeminal nerve involvement whose brain MRI revealed trigeminal nerve abnormalities consistent with the clinical picture. A 66-year-old woman with a 5-year history of untreated Sjögren's syndrome (the patient refused any therapy at the time of diagnosis) came to our attention for the subacute onset of paraesthesia, numbness and decreased sensation in the second and third cutaneous branch of left trigeminal nerve, associated with hand paraesthesias and clumsiness. At neurological evaluation she presented with instable gait; vibratory sensation was absent at upper limbs (up to the elbow) with wrist and finger pseudoathetosis, and reduced at lower limbs. Muscle strength was preserved. Deep tendon reflexes were absent. Cranial nerve examination revealed decreased sensation in the second and third cutaneous branch of the left trigeminal nerve. Corneal reflexes were present. Neurophysiologic evaluation showed widespread loss in sensory nerve action potential amplitude, worse at upper limbs. Needle EMG and blink reflex were normal. CSF analysis was normal. Spinal MRI was unremarkable. Brain MRI (1.5 T with a 3D high resolution CISS sequence) revealed atrophy of left trigeminal nerve, likely secondary to Gasser ganglion sensory neuron damage. The axial, coronal and sagittal reformatted sections showed a selective involvement of the sensory root of the left trigeminal nerve, which appeared smaller than the contralateral. No signs of neuro-vascular conflict were detected. The diameter and cross-sectional area of the cisternal segment of the trigeminal nerves, measured on