

Degenerator (bottom 16 percentiles,  $n = 67$ ), or Intermediate ( $n = 290$ ), with dramatically increased, decreased, or steady DMFD%, respectively. Fisher's exact test, ANOVA, and multifactorial logistic regression analyses were performed to identify significant risk factors. A preliminary microarray experiment was also performed on a subset of samples to identify regeneration-related genes and pathways.

**Results:** DMFD% were  $35.6 \pm 17.4$  (Regenerator),  $-4.8 \pm 12.1$  (Intermediate), and  $-39.8 \pm 11.0$  (Degenerator). HbA1c at baseline was the only risk factor significantly different between Regenerator ( $8.3 \pm 1.6\%$ ) and Degenerator ( $9.2 \pm 1.8\%$ ). Support Vector Machine classifier using HbA1c alone demonstrated 62.4% accuracy of classifying subjects into Regenerator or Degenerator. Gene expression studies revealed that up-regulated genes in Regenerator are enriched in cell cycle and myelin sheath functions, while down-regulated genes are enriched in immune/inflammatory responses.

**Conclusions:** Our data suggest that HbA1c level predicts myelinated nerve fiber regeneration and degeneration in patients with DN, and that maintaining optimal blood glucose control is essential to prevent continued nerve injury in patients with DN.

doi:10.1016/j.jns.2013.07.2410

#### Abstract – WCN 2013

No: 2720

Topic: 36 – Other Topic

#### Epigenetic miRNA dysregulation as a mechanism for sporadic amyotrophic lateral sclerosis

C. Figueroa-Romero<sup>a</sup>, J. Hur<sup>a</sup>, D.E. Bender<sup>a</sup>, M.D. Cataldo<sup>a</sup>, A.L. Smith<sup>a</sup>, R. Yung<sup>b</sup>, B.C. Callaghan<sup>a</sup>, E.L. Feldman<sup>a</sup>. <sup>a</sup>Neurology, University of Michigan, Ann Arbor, MI, USA; <sup>b</sup>Internal Medicine, University of Michigan, Ann Arbor, MI, USA

**Background:** Amyotrophic lateral sclerosis (ALS) is a progressive and fatal neurodegenerative disease. The majority of cases are sporadic (sALS) with unknown causes, and the late onset of sALS suggests that long-term exposures to environmental factors may play a role in disease pathogenesis. Epigenetic mechanisms, such as miRNAs, may be driven by adverse environmental factors proposed to contribute to sALS etiology to promote an accumulation of altered gene expression, culminating in sALS.

**Objective:** Our goal is to address the role of miRNAs in sALS pathogenesis. We hypothesize that differential expression of miRNAs in human sALS spinal cord promotes dysregulation of key genes and biological pathways leading to sALS.

**Materials and methods:** We combined two high-throughput profiling assays to identify differentially expressed miRNAs (TaqMan OpenArray) and differentially expressed genes (DEGs; Affymetrix GeneChip Human Genome U133 Plus 2.0 Array) in postmortem human spinal cord tissue from sALS patients and controls. Since miRNAs negatively regulate gene expression, inverse correlations between miRNA and DEGs were identified and examined with multiple miRNA prediction target databases to determine potentially relevant miRNA/DEG pairs. Gene and miRNA expression was confirmed by qPCR.

**Results:** We identified 90 differentially expressed miRNAs and 1182 DEGs. Of these differentially expressed miRNAs and DEGs, prediction databases identified miR-577, let-7 miRNA family members, miR-133b, and miR-140b-5p as potential miRNA regulators of the gene targets FAS, CD4, EIF2C4, and CCL2.

**Conclusion:** Changes in miRNAs and their corresponding gene targets may represent, in part, a pathogenic mechanism leading to sALS.

doi:10.1016/j.jns.2013.07.2411

#### Abstract – WCN 2013

No: 2715

Topic: 36 – Other Topic

#### Spontaneous remission and relapse of primary central nervous system lymphoma: Case report and review

J. Vilacura, F. Montoya, P. Silva, M. Rivera. Universidad de los Andes, Clinica Davila, Santiago, Chile

There are numerous reports of lesions affecting the central nervous system which disappear without treatment. They are called vanishing tumors. The most common of these is the primary central nervous system lymphoma, which accounts for 4% of all intracranial neoplasms. There are other factors that induce remission such as corticosteroid therapy, chemotherapy and radiotherapy. However, many of these vanishing tumors relapse. We present the case of a 58 year old, male patient who presented with partial third cranial nerve palsy. The brain MRI showed a periventricular mass and the biopsy was informed as diffuse large B-cell non-Hodgkin lymphoma. A control MRI was requested displaying complete disappearance of the mass. Patient was followed during 3 months with MRI, full body CT and lumbar puncture showing no evidence of the disease. One year after discharge patient presented with subtle memory alterations and progressive temporal-spatial disorientation. MRI showed a heterogeneous, hypointense, expansive process with left temporal periventricular epicenter. Spectroscopy was suggestive of lymphoma. Thoracic, abdominal, and pelvic CT scans evidenced an L1 lumbar fracture; MRI was suggestive of lymphoma metastasis. Daily methylprednisolone pulses were initiated and treatment with Methotrexate, Cytarabine and Leucovorin was begun, with clinical improvement. Many reports present relapsing vanishing tumors, suggesting that full therapy should be initiated in spite of spontaneous remission. Clinical trials should be performed in order to prove the effectiveness of this proceeding.

doi:10.1016/j.jns.2013.07.2412

#### Abstract – WCN 2013

No: 2768

Topic: 36 – Other Topic

#### Syndroma Brown-Séguard as a rare manifestation of von Hippel-Lindau disease

I. Marjanovic<sup>a</sup>, M. Jecmenica-Lukic<sup>a</sup>, M. Milicev<sup>a</sup>, S. Peric<sup>a</sup>, I. Basta<sup>a</sup>, Z. Stevic<sup>a</sup>, V. Rakocevic-Stojanovic<sup>a</sup>, S. Lavrnic<sup>b</sup>, D. Lavrnic<sup>a</sup>. <sup>a</sup>Clinic of Neurology, Clinical Centre of Serbia, Serbia; <sup>b</sup>MRI Center, Clinical Center of Serbia, University of Belgrade, Belgrade, Serbia

**Case report:** We report a 45-year-old female patient with Brown-Séguard Syndrome (BSS). The onset of disease was 14 years before admission to our Department, during the first pregnancy, when she felt thoracic pain. Five years later, during the next pregnancy, she felt numbness at umbilical region and drop of right foot, but all symptoms disappeared spontaneously 10 days after delivery. MRI of thoracic spine showed syrinx from Th5 to L1. Now, at the admission neurological examination revealed spasticity, increased muscle reflexes and positive Babinski's sign on the right leg, with ipsilateral loss of tactile discrimination, vibratory, and position sensation below the level of right rib arch. Contralaterally there was reduction in pain and temperature sensation below the level of Th8. MRI of thoracic spine showed multiple intradural nodular tumors at the levels of Th3–5 and Th11 with hydrosyringomyelia in whole spinal cord. MRI of the brain showed multiple hemangioblastomas in the craniocervical region with spreading at the level of C1 and in both cerebellar hemispheres. Multi-slice computed tomography (MSCT) of abdomen showed multiple pancreatic cysts and suspected liver hemangioma. Ophthalmological, otological and gynecological findings were normal. Genetic analysis