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Author(s)	Lee, PCH; Chow, WS; Yuen, MAM; Tso, AWK; Xu, A; Law, LSC; Cheung, SCW; Chau, MT; Cheung, BMY; Lam, KSL
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Brainstem encephalitis in neuromyelitis optica spectrum disorders

<u>JCY Lee</u>¹, SYY Pang¹, GKK Lau¹, KC Teo¹, SL Ho², RTF Cheung², JSC Kwan¹, KH Chan² University Department of Medicine, Queen Mary Hospital,, The University of Hong Kong

²Research Centre of Heart, Brain, Hormone and Healthy Aging, LKS Faculty of Medicine, The University of Hong Kong, Hong Kong

Introduction: Neuromyelitis optica spectrum disorders (NMOSD) are severe CNS inflammatory demyelinating disorders characterised by extensive acute myelitis (AM) and optic neuritis (ON). NMOSD are probably due to aquaporin-4 (AQP4) autoimmunity as ~60-80% NMO patients are seropositive for AQP4 autoantibodies (AQP4 Ab). We recently reported that brain involvement is common in NMOSD in Hong Kong Chinese, and the brainstem is the most common site of brain involvement, which clinically presents as brainstem encephalitis (BE).

Aim: To study the clinical and radiological features of BE in local NMOSD patients.

Methods: NMOSD patients diagnosed (according to Wingerchuk's criteria) and followed up in QMH from January 1988 to September 2011 were reviewed. The clinical and neuroradiological characteristics of those with brainstem encephalitis were studied in details. AQP4 Ab was assayed by cell-based immunofluorescence using transfected HEK293 cells expressing full length human AQP4 gene.

Results: Forty NMOSD patients (21 NMO, 11 relapsing AM, 6 relapsing ON, 1 relapsing BE, 1 AM with BE) were studied. Their mean onset age was 39.5 years (range, 17-80), 36 (90%) were female; mean follow-up duration 5.0 years (range, 1-16). AQP4 Ab were detected in 25 (62.5%). Twelve (30%) of the 40 patients had clinical attacks of BE presenting with diplopia, dizziness, ataxia, vertigo, nausea, vomiting, third nerve palsy, internuclear ophthalmoplegia, facial weakness (both LMN and UMN type), tinnitis, hearing loss, aphonia, dysphagia, aspiration pneumonia, autonomic dysfunction, facial sensory loss, long-tract sign. MRI brain revealed lesions in midbrain (10%), pons (10%) and medulla (20%), with brainstem lesion continuous with extensive cervical myelitis as the most common brainstem lesion in patients with BE. Two patients succumbed during severe BE involving medulla in continuity with extensive cervical myelitis.

Conclusion: Brainstem encephalitis is common in NMOSD among Hong Kong Chinese, which present with diverse bulbar symptoms and signs. The medulla is the most frequently affected site.

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PCH Lee¹, WS Chow¹, MAM Yuen¹, AWK Tso¹, A Xu¹, LSC Law¹, SCW Cheung², MT Chau², BMY Cheung¹, KSL Lam¹ Department of Medicine, The University of Hong Kong, Hong Kong ²Department of Radiology, Queen Mary Hospital, Hong Kong

Introduction: Hypoadiponectinaemia has been shown to predict the development of type 2 diabetes, hypertension, and myocardial infarction in prospective studies. We have previously reported that hypoadiponectinaemia is associated with impaired endothelium-dependent vasodilation both in healthy controls and diabetic subjects. In this community-based prospective cohort study, we examined the predictive value of serum adiponectin levels on carotid intima-medial thickness, a marker of atherosclerotic disease, in Hong Kong Chinese.

Methods: Subjects were recruited from the Hong Kong Cardiovascular Risk Factors Prevalence Study 2 (CRISPS 2) cohort. Those with known cardiovascular disease(s) were excluded. Fasting plasma glucose, insulin, and lipid profile were measured. Baseline serum levels of adiponectin and carotid intima-media thickness (IMT) were determined. Subjects were followed prospectively for 5 years.

Results: 265 subjects (129 male, 54.6 ± 12.3 years) were included in the final analysis. At baseline, 86 and 34 subjects have hypertension and type 2 diabetes respectively. Their body mass index (BMI) was 24.9 ± 3.7 kg/m², systolic blood pressure 125 ± 20 mm Hg, diastolic blood pressure 76 ± 11 mm Hg, LDL-cholesterol 3.4 ± 0.8 mmol/L, HDL-cholesterol 1.3 ± 0.4 mmol/L, fasting glucose 5.3 ± 1.2 mmol/L, HOMA-IR median (interquartile range) 1.80 (1.28-2.76), and adiponectin 5.28 mg/L [3.29-7.93]. Over 5 years, carotid IMT increased from 0.62 mm (0.52-0.73) to 0.67 (0.57-0.78) [P<0.001, paired t-test]. Linear regression analysis showed that age (β =0.22, P=0.02), smoking status (β =0.16, P=0.014), baseline IMT (β =0.59, P<0.001) and sex-specific median adiponectin (β =0.14, P=0.031) were independent predictors of IMT at 5 years in men, after controlling for the conventional cardiovascular risk factors. Adiponectin (sex-specific median) was not significantly predictive of 5-year IMT in women.

Conclusion: In this 5-year prospective study, hypoadiponectinaemia was an independent predictor of IMT thickening in Hong Kong Chinese men. Our data suggest that the measurement of serum adiponectin would allow for early risk stratification even in a community-based cohort, and enable timely implementation of medical interventions to reduce future cardiovascular events among subjects with hypoadiponectinaemia.