Wolfram Syndrome: Presentation of Two Cases with Type One and Type Two Diabetes Mellitus

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Introduction: Wolfram syndrome (WS) is a rare (1/770.000), autosomal recessive genetic disorder with *WFS1* gene mutation. It generally presents with autoantibody (-) type 1 diabetes mellitus (T1DM), optic atrophy, diabetes insipidus (DI), deafness, and renal anomalies. In 2002, the syndrome was related with T2DM for the first time. Consequent publications supported the relationship between *WFS1* mutation and T2DM requiring high insulin doses. As the components of the syndrome are similar with the common complications of DM, the diagnosis can occasionally be missed. We hereby present two cases with Wolfram syndrome who were admitted to our clinic with T1DM and T2DM.

Case 1: A 10-year-old girl presented with polyuria, polydipsia, weight loss, and hyperglycemia [blood glucose (BG): 450 mg/dL]. She had enuresis nocturne since the age of six. WS was suspected with autoantibody (-) T1DM and DI. Four years later, bilateral optic atrophy and sensorineural deafness were detected. BG was regulated with 0.3 U/kg/day basal-bolus insulin. Neurogenic bladder with bilateral hydronephrosis developed when she was 31 years old. Screening of family members revealed WS with DM, DI, and optic atrophy in a first-degree cousin at the age of seven.

Case 2: A 46-year old woman who had T2DM for 16 years was admitted to our clinic for BG regulation before thyroidectomy. Medical history revealed sudden vision loss and deafness which started seven years and one year ago, respectively. Bilateral optic atrophy and sensorineural hearing loss were detected in further evaluation. The patient was diagnosed with WS, and BG was regulated with 2.1 U/kg/day basal-bolus insulin and metformin treatment.

Conclusion: Diagnosis of Wolfram syndrome should be considered when early-onset or unexpected DM complications such as retinopathy or neuropathy are detected in the surveillance of patients with T1DM and with T2DM as well.

Key words: Wolfram syndrome, diabetes mellitus, autosomal recessive

Abdominal Obesity May Be Caused by Increasing Cortisol Levels with Age

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Cortisol is a stress hormone and chronic stress can increase visceral fat accumulation. Elevated levels of blood cortisol intensify central fat deposition importantly. This type of obesity is predicting all cause and cardiovascular disease mortality.

Aim: We investigated the relationship between abdominal obesity and cortisol level and their risk factors in our obese patients group (totally 118). Some characteristics of the study group are as follows [mean± standard deviation (SD)]: age(years) 49±15, body mass index (BMI) (kg/m^2) 32.2±7.5, waist circumference (cm) 98.9±16.3, waist/hip ratio (WHR) 0.87±0.07, systolic blood pressure (BP) (mmHg) 128.2±19.2, diastolic BP (mmHg) 78.3±9.6, basal insulin level (μU/mL) 12.21±11.41, C-peptide (ng/dL) 3.04±1.71, hemoglobin A1c (HbA1c) (%) 5.74±1.35, low-density lipoprotein cholesterol (ng/dL) 145.2±64, high-density lipoprotein (HDL)-cholesterol (ng/dL) 43.31±12.2, triglyceride (ng/dL) 153.5±92.2, basal cholesterol level (μg/dL) 14.3±7.7.

Results: When we divided the patients into two groups according to their basal cortisol levels (≤10μg and >10μg), we found elevated fasting blood glucoses, basal insulin (n.s.), C-peptide, prolactin, free thyroxine, and HbA1c levels, diminished HDL, increased waist circumference and WHR with relatively aged patients in the second group.

Conclusion: Cortisol levels even at near upper limits may show an insulin resistance, impaired fasting glucose concentrations, and abdominal obesity.

Key words: Abdominal obesity, cortisol