A 57-year-old male with type 2 diabetes and hyperpigmentation of the palms, armpits; weakness in the eyes. Objectively: asthenic body type, BMI 16.5 kg/m², the dose of hydrocortisone 10 mg/day did not change with age. Diagnosis: primary chronic adrenal insufficiency; the increase in ACTH 470 pg/ml (0.0–46 pg/ml), cortisol 0.05 µg/DL. The secretion of mineralocorticoids was evaluated: plasma aldosterone and renin levels were within the reference values. Ophthalmologist: injected conjunctiva, sclera. Schirmer’s test: mild alacrimia. It allowed to make the diagnosis: “Primary chronic adrenal insufficiency. Condition after surgery for achalasia (1997). Alacrimia. Allgrove Syndrome.” The dose of hydrocortisone was increased to 17.5 mg/day. In 2019, the patient complained of a sharp deterioration of health, darkening of the skin. The dose of hydrocortisone was increased to 25 mg/day (15 mg at 8.00, 10 mg in the afternoon). The ophthalmologist noted an increase in the severity of alacrimia, artificial tear drops was recommended. The diagnosis was confirmed by pathogenic mutation c.43C>T of the AAAS gene. Discussion: Despite the full clinical picture, the right diagnosis was made only after 14 years. We shown the difficulty of diagnosis is due to the lack of awareness of clinicians about the disease, the importance of interdisciplinary interaction, as well as the need for follow-up of such patients. Reference: (1) Handschug K, Sperling S, Yoon SJ, et al. Triple A syndrome is caused by mutations in AAAS, a new WD-repeat protein gene. Human Molecular Genetics. 2001;10:283–290.

Adrenal

ADRENAL CASE REPORTS I

Allgrove Syndrome: How to Suspect a Problem

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SAT-219

Background: Allgrove syndrome (triple A syndrome) is a rare autosomal recessive multisystem disease characterized by adrenal insufficiency, alacrimia and achalasia. It is caused by a mutation in the AAAS gene (12q13) encoding the protein ALADIN (1). This syndrome is often associated with neurological dysfunction disorders, amyotrophy, in such cases, it is named 4A and 5A syndrome, but sometimes there is also 2A syndrome. Prevalence:<1/100000.

The first description was in 1978. Clinical case: A 18-year patient A. complained of fatigue, weakness, darkening of the skin. From anamnesis of life: born from the first pregnancy, no complications, weight 3200g. Parents often turned to the pediatrician with complaints: lethargy, frequent regurgitation, ARVI up to 6–7 times a year. Slow weight gain, dyspeptic syndrome (nausea, vomiting) was noted objectively. At the age of 3, the boy entered the surgical department with acute abdomen, fever, vomiting. Achalasia was revealed, reconstructive surgery was carried out. In the diagnostic search for the causes of body weight loss he was directed to the endocrinologist. There were an increase in ACTH 470 pg/ml (0.0–46 pg/ml), cortisol 0.05 µg/DL. Diagnosis: primary chronic adrenal insufficiency; the dose of hydrocortisone 10 mg/day did not change with age. An in-depth examination found: the patient never cried with tears. Objectively: asthenic body type, BMI 16.5 kg/m², hyperpigmentation of the palms, armpits; weakness in the proximal muscles of the limbs. Laboratory studies: ACTH 95 PG/ml, cortisol 0.1 µg/DL (3.7–19.4 µg/DL). The secretion of mineralocorticoids was evaluated: plasma aldosterone and renin levels were within the reference values. Ophthalmologist: injected conjunctiva, sclera. Schirmer’s test: mild alacrimia. It allowed to make the diagnosis: “Primary chronic adrenal insufficiency. Condition after surgery for achalasia (1997). Alacrimia. Allgrove Syndrome.” The dose of hydrocortisone was increased to 17.5 mg/day. In 2019, the patient complained of a sharp deterioration of health, darkening of the skin. The dose of hydrocortisone was increased to 25 mg/day (15 mg at 8.00, 10 mg in the afternoon). The ophthalmologist noted an increase in the severity of alacrimia, artificial tear drops was recommended. The diagnosis was confirmed by pathogenic mutation c.43C>T of the AAAS gene. Discussion: Despite the full clinical picture, the right diagnosis was made only after 14 years. We shown the difficulty of diagnosis is due to the lack of awareness of clinicians about the disease, the importance of interdisciplinary interaction, as well as the need for follow-up of such patients. Reference: (1) Handschug K, Sperling S, Yoon SJ, et al. Triple A syndrome is caused by mutations in AAAS, a new WD-repeat protein gene. Human Molecular Genetics. 2001;10:283–290.

Diabetes Mellitus and Glucose Metabolism

DIABETES COMPLICATIONS II

Emphysematous Gastritis and Diabetes

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MON-688

Background: Emphysematous gastritis (EG) is a rare and severe form of gastritis of infectious origin. Diabetes is an important underlying risk factor as it leads to a systemic predisposition to infections. Other risk factors include long term steroid use, nonsteroidal anti-inflammatory (NSAID) use, alcohol use, corrosive ingestion, and pancreatitis, all of which disrupt gastric mucosa. First described in the 1800s, it is characterized by the presence of air in the stomach wall and differentials for these cases include gastric emphysema and cystic pneumatosis both of which are non-infectious in origin.

Clinical Case: A 57-year-old male with type 2 diabetes presented with a one-day history of abdominal pain, non-bloody diarrhea, and vomiting. One day prior to presentation, he developed diarrhea which was followed by episodes of projectile vomiting reported as orange-tinged with mucus. On the day of admission, he was afebrile, tachycardic in 120s with stable blood pressure. Laboratory evaluation was significant for leukocytosis at 18.8 k/uL (4.3–11.3 k/uL) and lactic acidosis 2.37 mmol/L (0.7–2.1 mmol/L). Abdominal examination was notable for soft abdomen with diffuse tenderness to deep palpation without rebound or guarding. Further workup with Computed Tomography (CT) was concerning for emphysematous gastritis with air in the gastric vein, splenic vein, and portal vein. Given hemodynamic stability and benign abdominal examination, medical management was initiated. He was...
Discussion: EG results from disruption in gastric mucosa which facilitates translocation of gas-producing bacteria commonly Klebsiella pneumonia, Escherichia coli, Pseudomonas aeruginosa, and Enterobacter subspecies. Immunosuppression with diabetes is an important underlying factor and patients are at risk even with controlled diabetes. Additionally, patients with diabetic complications like gastroparesis with frequent retching are at increased risk. Considering variable and non-specific symptoms of presentation, a high index of clinical suspicion is required for recognition as it may have a fulminant course with high mortality risk. CT scan is the imaging of choice for diagnosis. Management primarily consists of bowel rest, antibiotics and monitoring for signs of peritonitis. In the absence of complications including rupture or stricture formation, surgery is not recommended. In our case, possible gastroenteritis with subsequent vomiting and retching in the setting of underlying diabetes predisposed to the development of emphysematous gastritis. Although air in the portal venous system is associated with higher mortality, our patient was successfully managed conservatively. As the diagnosis carries a high mortality risk, early recognition is imperative for a successful outcome.

Cardiovascular Endocrinology
ENDOCRINE HYPERTENSION AND ALDOSTERONE EXCESS

Epigenetic Regulation of 11beta-Hydroxysteroid Dehydrogenase 1 and 2 Gene in Salt-Sensitive Hypertensive Rats

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ENDO 2020
Epigenetic regulation of 11beta-hydroxysteroid dehydrogenase 1 and 2 gene in salt-sensitive hypertensive rats

Objective: 11 Beta-hydroxysteroid dehydrogenase type1 (11-HSD1) is the modulator of glucocorticoid hormone and type2 (11-HSD2) is the modulator of mineralocorticoid hormone. We investigated the effect of high salt diet on the methylation of both enzyme gene in salt-sensitive hypertensive (SSH) rats. Methods: SSH rats were fed a high (7% NaCl) or normal (0.45%) salt chow for 4 weeks. Body weight, blood pressure, plasma and urinary aldosterone (ALDOSTERONE) concentrations and PRA were measured. DNA was extracted from kidneys and visceral fats. Bisulfite sequencing and Pyrosequencing were done for the analysis of methylation status of 11-HSD1 and 2 gene. Results: High salt diet significantly decreased methylation ratio of 11-HSD2 gene in visceral fats of SSH rats compared with controls (p<0.05). The methylation ratio of 11-HSD2 gene in the kidney of SSH rats was not influenced by high salt diet. Discussion and Conclusion: 11-HSD2 gene overexpression in visceral fats in mice was reported to show SSH. We reported decreased 11-HSD2 activity in the artery in SSH rats. In this study confirmed, papillary microcarcinoma with metastasis to 1 regional lymph node was revealed. Diagnosed: papillary thyroid cancer I st (pT1aN1aM0x), 2 clinical group. The patient was prescribed suppressive therapy with L-thyroxine 100 µg/day, against which after 3 months TSH reached the target values (0.2–0.5 Mme/l). Taking into account the histological characteristics of the tumor, the nature and volume of the lesion, age, the patient belongs to the group of intermediate cancer risk of progression of cancer. According to scintigraphy residual functioning thyroid tissue (20x15 mm) was detected. Radioiodine therapy was carried out in a specialized hospital. Suppressive therapy of L-thyroxine 150 µg/day, target values of TSH 0.1 - 0.5 Mme/l was recommended. After 6 months, TSH reached target values, and according to the results of ultrasound of thyroid gland no data for structural relapse was found.

Conclusion: Patients with long-existing, often recurrent Graves’ disease and questionable effect of conservative therapy, in the presence of nodular formation should be assigned to the risk group for the presence of thyroid cancer and carefully examined, because the need for further surgery depends on it.