Case report of autoimmune polyglandular syndrome IIIa

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Autoimmune polyglandular syndrome type III (subtype IIIa) has polygenic inheritance and is most common among other polyendocrine autoimmune syndromes. As the main feature polyglandular syndrome-IIIa includes one of the autoimmune thyroid diseases combined with other autoimmune endocrine diseases that are not included in the polyendocrine (polyglandular) autoimmune syndromes types I and II. Because of the rarity of the reported cases of autoimmune polyglandular syndromes, we present the case of patient, diagnosed with type IIIa autoimmune polyglandular syndrome. In the described case the first recorded autoimmune disease was autoimmune diabetes mellitus type I, followed by autoimmune thyroiditis. Based on the occurrence of polyglandular syndrome-IIIa type, it is necessary to emphasize the need for registration and dynamic monitoring of the patients with identified autoimmune diabetes mellitus type I or autoimmune thyroid diseases in order to early diagnosis the combination of these organ-specific autoimmune diseases.

Keywords: autoimmune polyglandular syndrome type III, APS-III, APS-3.

Aim

Because of the rarity of the reported cases of autoimmune polyglandular syndromes (APS), we have studied the case of patient Zh.V. diagnosed with type IIIa autoimmune polyglandular syndrome.

Material and methods

A 35-year-old woman consulted a doctor in August, 2015. The follow-up period was 3 years. An anamnesis was taken and clinical examination performed.

Blood levels of autoantibodies towards: islet cell, glutamic acid decarboxylase (GAD) (Biomerica), insulin, thyroperoxidase (TPO), thyroglobulin (TG) (ORGENTEC Diagnostika), thyroid stimulating hormone (TSH) receptors (Medipan GmbH), 21-hydroxylase (Biovendor) were measured. Clinical dynamic observation was established for the patient. Blood concentrations of TSH, free thyroxine (fT4), cortisol, glucose and glycosylated hemoglobin were also determined in dynamics.

Results

Three months before admission against the background of type I diabetes, the patient showed a deterioration after a respiratory infection complicated with bronchitis. Moreover, the patient noted attacks of "unexplained" weakness, impairments of efficiency and

memory, periodic chills. These complaints continued despite the insulin doses correction and the presence of satisfactory glucose values ranging from 108 to 126 mg/dl. Type I diabetes was diagnosed in the patient at the age of 25 when she appealed to the outpatient department complaining of dry mouth, polydipsia and weight loss of 3 kg for 2 weeks. Actropide therapy was started after elevated glucose levels (198 mg/dl and 216 mg/dl) detection. The patient's condition was compensated and remained satisfactory for a long time with glycosylated hemoglobin level ranging from 6.5 to 7 %.

Allergic reactions were not noted. On physical examination upon admission the patient's characteristics were the following: body mass — 84 kg; height — 185 cm. The skin was clean. Tongue with teeth imprints, oedematose and pale. The feeling of an enlarged tongue was also subjectively noted by the patient. The patient had vesicular breathing with no râles/wheezing. Heart sounds were muffled. There was respiratory arrhythmia. The pulse rate was 58 beats per minute. Body temperature in the right and left axilla was 36.1 and 36.3 °C. The abdomen was soft and painless on palpation. Liver border was at the edge of the costal arch. The spleen was not palpable. An ultrasounography of the thyroid gland revealed a volume of 13.5 ml, a heterogeneous structure due to alternating fields of different densities (including hypoechogenic ones), and single fibrous cords. During ultrasound monitoring of the adrenal glands, they were not revealed. The level of TSH was increased and the level of fT4 — reduced (Table 1). The cortisol serum concentration was 780 nmol/l. The glucose level was 113.4 mg/dl. The concentration of glycosylated hemoglobin was 7.2 %. The level of thyroperoxidase autoantibodies was increased (Table 2). Elevated concentrations of autoantibodies to insulin and to GAD were also identified.

Table 1. **Indicators of the endocrine function in patient Zh.V**

Parameters	Hormone concentration, glucose and glycosylated hemoglobin levels				
	Free thyroxine (pmol/l)	TSH (mcIU/ml)	cortisol (nmol/l)	glucose (mg/dl)	glycosylated hemoglobin (%)
Patient Zh.V.	9	5	780	113.4	7.2
Reference intervals	10.0–23.2	0.23–3.4	150.0–660	< 110	4.2–6.2

Table 2. **Autoantibody levels to organ-specific autoantigens in patient Zh.V**

Parameters	Autoantibody levels to autoantigens						
	Pancreas			thyroid gland			adrenal gland
	islet cell (ICA ratio)	insulin (U/ml)	GAD (U/ml)	TPO (IU/m l)	TG (IU/ml)	TSH receptors (IU/l)	21-hydroxyllase (U/ml)
Patient Zh.V.	0.66	14.2	4.4	270.4	50.0	0.8	0.35
Reference interval	< 0.95	< 10	< 1	< 50	< 100	≤ 1	< 0.45

Diagnosis

Autoimmune polyglandular syndrome type IIIa. Autoimmune diabetes mellitus type I (moderate course). Autoimmune thyroiditis. Moderate hypothyroidism.

The patient's condition improved after the appointment of Euthyrox in a dose of 50 mcg per day. Efficiency increased, chills stopped. The feeling of an enlarged tongue and teeth imprints disappeared. The concentration of TSH returned to normal (1.91 mcIU/ml). Glucose during dynamic examination did not exceed 108-124.2 mg/dl. The concentration of cortisol during 2 weeks of treatment decreased and amounted to 721.3 nmol/ml followed by a further decrease to the upper limits of normal values. During the differential diagnosis we excluded other types of autoimmune polyendocrine syndromes [1–3]. In order to exclude the presence of autoimmune disorders of the adrenal glands we tested the levels of 21-hydroxylase autoantibodies which were within normal values.

Discussion

Thus, we have diagnosed autoimmune polyglandular syndrome type III (subtype IIIa) in the patient. It should be noted that it has polygenic inheritance and is most common among other polyendocrine autoimmune syndromes. As the main feature PAS-IIIa includes one of the autoimmune thyroid diseases combined with other autoimmune endocrine diseases that are not included in the polyendocrine (polyglandular) autoimmune syndromes types I and II [3]. It is worth to mention that this syndrome is more common among women. In the described case the first recorded autoimmune disease was autoimmune diabetes mellitus type I, although such an order not always occurs.

Conclusion

Based on the occurrence of PAS-IIIa type, it is necessary to emphasize the need for registration and dynamic monitoring of the patients with identified autoimmune diabetes mellitus type I or autoimmune thyroid diseases in order to early diagnosis the combination of these organ-specific autoimmune diseases.

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