Autoimmune Polyglandular Syndrome Case Report Series

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Introduction

The polyglandular autoimmune syndromes (PAS) are rare conditions characterized by the failure of several endocrine glands sometimes associated with other non-endocrine autoimmune diseases.There are four categories of PAS: PAS-I includes at least two out of: mucocutaneous candidiasis, hypoparathyroidism and adrenocortical failure. PAS-II comprises of Addison's disease, autoimmune thyroid disease and/or type 1 diabetes (Carpenter's syndrome). PAS-III is defined by the presence of autoimmune disorders other than Addison's disease and hypoparathyroidism, while PAS IV includes non-endocrine autoimmune disorders and Addison's disease, but not hypothyroidism.

Case reports

Case1 55-year-old woman

•**Primary ovarian failure** (FSH=16.26 mUI/ml) at age of 33.

•**Addison's disease and chronic autoimmune thyroiditis** at 42

9 a.m. plasma cortisol = 1.22 µg/dl, ACTH = 704.46 pg/ml, TSH=3.7 µIU/ml, T4= 6.54 µg/dl, T3=117 ng/dl, ATPO=1026 IU/L; @46 years old : TSH=7.9 µIU/ml

Thyroid ultrasound: diffuse, heterogeneous, hypoechogenicity, with increased blood flow.

Abdominal CT scan: hypoplastic adrenal glands.

•**Diabetes mellitus**

46 years	OGTT: FPG 111 mg/dl + 2 h: 395 mg/dl	Nutritional medical therapy
47 years	Hyperglycemia (>180 mg/dl)	Oral antidiabetic drugs (OAD)
48 years	HbA1c-12,2% + C-peptide 0,821 ng/ml	Basal insulin + OAD
51 years	HbA1c-12%	Intensified insulin therapy + OAD

Her father – type 1 diabetes mellitus

GADA >2000 IE/ml

Latent Autoimmune Diabetes of the Adults (LADA)

Pancreatic antibodies	Abnormal range	Normal range
GADA	>2000 IE/ml	<10 IE/ml
Pancreatic islet cell antibodies	1/1000	<1/10
Anti-insulin antibodies	10,9 U/ml	<0,4 U/ml
Anti-insulin receptor antibody	IgG=positive	

- @38 years old: **Type 1 Diabetes mellitus**: poliuro-polydipsia and weight loss (12 Kg in the last 5 months). HbA1c 9% ; basal bolus insulin therapy.
- @ 39 year-old total thyroidectomy + levothyroxine replacement.
- @ 40 year-old TSH=6.9 µIU/ml, FT4=19.4 pmol/l, ACTH= 10.29 pg/ml (3-66 pg/ml), cortisol= 12.78µ g/dl (6.7-22.6 µg/dl), HbA1c=9.67% (due to noncompliance with diet/ antiinsulin antibodies).

Currently on basal-bolus insulin therapy, levothyroxine 137.5 mcg/day after thyroidectomy.

Diagnoses: **Polyglandular autoimmune syndrome type III:**
- **Basedow's disease**
- **Type 1 diabetes mellitus**
- **Vitiligo**

Currently on premixed insulin plus metformin 2.25 g/day, levothyroxine 75 mcg/zi, hydrocortisone 25 mg/day, fludrocortisone 0.1 mg/day, statin. HbA1c 7.0%.

Diagnoses: **Polyglandular autoimmune syndrome type II:**
- **Addison disease**
- **Chronic autoimmune thyroiditis**
- **Latent autoimmune diabetes of the adults**
- **Primary ovarian failure**

Case 2 42 old man

•**vitiligo** since 14 years old.

•@ 36 years **Basedow's disease**: muscle weakness, intense fatigue, sweating, tremor of the extremities, palpitations, weight loss. **Labs**: TSH <0.03 µIU/ml (0.5-4.5 µUI/ml), T4 >20 µg/dl (4.5-13 µg/dl), T3 >500 ng/dl (80-200 ng/dl). **Thyroid ultrasound**: diffuse hypoechogenicity, heterogeneous with increased vascularization.

Case 3 22-year-old woman

•**type 1 diabetes mellitus** and **primary hypothyroidism** since she was 16 years old. ATPO= 0.18 IU/ml (<5.61 UI/ml).

•@17 years old: height=150 cm, weight=36 Kg, IMC=16 Kg/m². **Labs**: HbA1c=4.81% TRAb=0.45 IU/ml, TSH=4.3 µUI/ml, FT4=18.2 pmol/L, IGF-1=382 ng/ml (226-903 ng/ml).

Thyroid ultrasound: small goiter, diffuse hypoechogenicity

• @ 18 years old screening for other autoimmune diseases: estradiol = 110.91 pg/ml, LH=6.73 mIU/ml, FSH=8.46 mIU/ml– in normal range, PRL=7.48 (2.8-29.2 ng/ml), 9 a.m. plasma cortisol=20.09 (4.30-22.40 µg/dl), ATPO< 0.3 IU/L (ATPO may be absent in 10-15% of Hashimoto's disease, mostly in young).

Currently on basal-bolus insulin therapy and levothyroxine 50 mcg/day.

Diagnoses: **Polyglandular autoimmune syndrome:**
- **Type 1 diabetes mellitus**
- **Primary hypothyroidism**

Conclusion

We emphasize the importance of screening for PAS after the first autoimmune disease is diagnosed. The key to successfully managing patients with PAS is to identify and treat their disorders early before complications occur. This may be achieved by early screening for autoantibodies or subclinical endocrine failure. Patients should be educated to comply with the lifelong medical surveillance and encourage their family members to be screened for autoimmune diseases as about 50% of patients with PAI II have siblings with autoimmune diseases.

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Topic **Clinical case reports - thyroid/others**

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