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 adenocortical failure. PAS-II comprises of Addison’s disease, autoimmune thyroid disease and/or type 1 diabetes (Carpenter’s syndrome). PAS-II is defined by the presence of autoimmune disorders other than Addison's disease and hypoparathyroidism, while PAS IV involves non-endocrine autoimmune disorders and Addison’s disease, but not hypothyroidism.

Case reports

Case 1: 55-year-old woman
- **Primary ovarian failure** (FSH=16.26 mIU/ml) at age of 33.
- **Addison’s disease and chronic autoimmune thyroiditis** at age of 42. At 9 a.m. plasma cortisol = 1.22 μg/dl, ACTH = 70.46 pg/ml, TSH = 3.7 μU/ml, T4 = 6.54 μg/dl, T3 = 117 ng/dl, ATPO = 1026 IU/L; 36 years old: TSH=7.9 μU/ml
- **Thyroid ultrasound**: diffuse, heterogeneous, hypoechoegenicity, with increased blood flow.
- **Abdominal CT scan**: hypoplasic adrenal glands.

**Diabetes mellitus**

46 years: OGTT: FPG 111 mg/dl + 2 h: 395 mg/dl
47 years: Hyperglycemia (>180 mg/dl)
48 years: Hba1c=12.2% + C-peptide 0.821 mg/ml
51 years: Hba1c=12%

GADA >2000 IE/ml + OAD

Her father – type 1 diabetes mellitus

Latent Autoimmune Diabetes of the Adults (LADA)

Currently on preixed 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 III:
- Basedow’s disease
- Type 1 diabetes mellitus
- Vitiligo

Case 2: 42 old man
- **Vitiligo** since 14 years old.
- **At 36 years Basedow’s disease**: muscle weakness, intense fatigue, sweating, tremor of the extremities, palpitations, weight loss. Labs: TSH <0.03 μU/ml (0.5-4.5 μU/ml), T4 >20 μg/dl (4.5-13 μg/dl), T3 >500 ng/dl (80-200 ng/dl). Thyroid ultrasound: diffuse hypoechoegenicity, heterogeneous with increased vascularity.

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

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 IU/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 μU/ml, FT4=18.2 pmol/l, IGF-1=392 ng/ml (228-803 ng/ml).
  - Thyroid ultrasound: small goiter, diffuse hypofunctionicity.
- 18 years old screening for other autoimmune diseases: estradiol = 110.91 pg/ml, LH=67.3 mlU/ml, FSH=8.46 mlU/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 have siblings with autoimmune diseases.

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Clinical case reports - thyroid/others

References