

Fainting matters: a case of an autoimmune polyglandular syndrome (APS) with an atypical involvement of the parathyroid gland

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Introduction: The autoimmune polyglandular syndrome (APS) is characterized by the coexistence of at least two glandular autoimmune mediated diseases.

We report a combination of an autoimmune thyroid disease, Addison's disease and an atypical involvement of the parathyroid gland.

First visit:

A 73-years old white woman was admitted to the clinic for evaluation due to recurrent syncope during the previous 3 years that presented along with:

- vertigo
- cold sweats
- general fatigue

and was associated with prolonged standing.

- Loss of 15 kg weight within the past year
- No skin changes observed

3 months ago she was diagnosed with:

- Chronic autoimmune thyroiditis and primary hypothyroidism
- Parathyroid gland adenoma (SPECT-CT)
- Orthostatic hypotension

Investigations:

- Head CT scan – basal ganglia calcinosis
- Head MRI, EEG, brachiocephalic vessel duplex ultrasonography, Holter monitoring revealed no substantial changes
- DXA scan – lumbar T-score -3,1 SD
- Antibody screen (ANA, thyroid peroxidase, transglutaminase) was negative.
- Low cortisol in 24h urine
- No data on diabetes mellitus or malignancy

Conclusions:

- PTH level was normal, but the coexistence of hypocalcaemia and parathyroid gland adenoma must be taken into consideration
- We would like to stress the importance of investigation for autoimmune glandular diseases and electrolyte level in case of unexplained syncope.

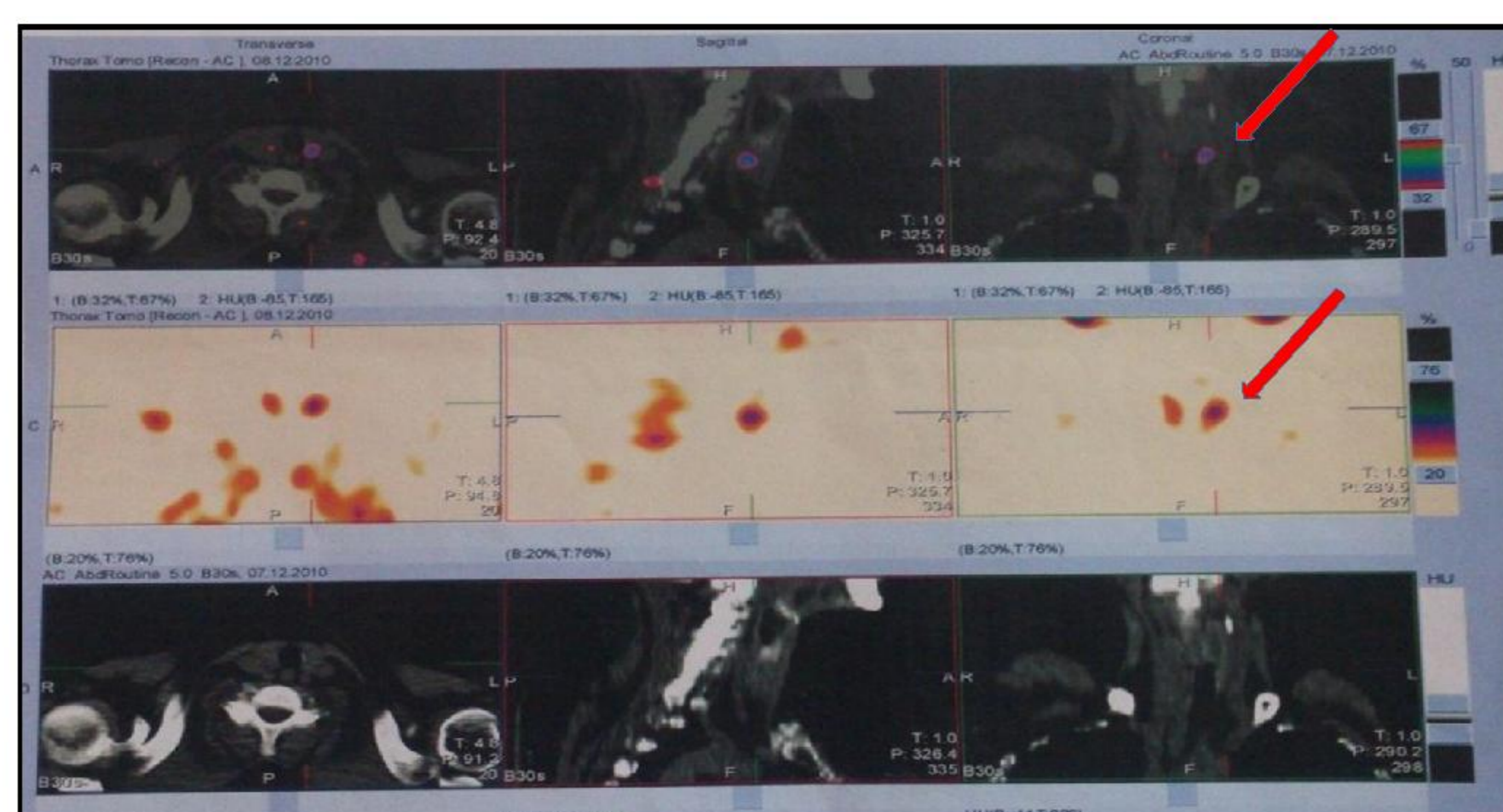
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Aldosterone (40 – 310 pg/ml)	3.7
Renin (4.4-46.1 mU/l)	2.1
ACTH (7.2 – 63.3 pg/ml)	44.5
Cortisol 8:00 (3.7-19.4 mkg/dl)	3.7
TSH (0.35 – 4.94 mU/L)	12
FT4 (0.71 – 1.85 ng/dl)	0.95
Calcium (2.1 – 2.55 mmol/l)	2.03
PTH (15 – 68 pg/ml)	62.4
Creatinine (62 – 106 mkmol/L)	95

Insulin stress test – insufficient rise

Plasma cortisol at 8:00 (3.7-19.4 mkg/dl)	3.7
30 min after infusion	4.1
60 min after infusion	4.4
15 min after hypoglycemia	10.0
60 min after hypoglycemia	12.9

SPECT-CT – left upper parathyroid gland adenoma



Diagnosis:

- Primary adrenal insufficiency with glucocorticoid and mineralocorticoid deficiency
- Chronic autoimmune thyroiditis with primary hypothyroidism
- Hypocalcaemia
- Calcinosis of basal ganglia (possible Fahr's syndrome)
- Parathyroid gland adenoma
- Secondary osteoporosis due to malabsorption
- Chronic kidney disease

Based on these findings, type 2 autoimmune polyglandular syndrome was diagnosed.

Treatment:

- L-Thyroxine
- Oral calcium
- Alphacalcidol
- Hydrocortisone
- Fludrocortisone

A substantial improvement of symptoms was seen in a control visit after 3 months

