

Fluctuating primary ovarian insufficiency in a case of type 2 autoimmune polyendocrinopathy

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Context: Type 2 autoimmune polyendocrinopathy is a rare disease, occurring with a prevalence of 1.4-2 per 100 000 people. It groups autoimmune adrenal insufficiency, autoimmune thyroid disease, type 1 diabetes mellitus, premature ovarian insufficiency and several other endocrine and non-endocrine autoimmune diseases. It is most often diagnosed in female patients, in the third and fourth decades. Its early diagnosis is crucial as unrecognized adrenal insufficiency can have life-threatening consequences.

Objective: We report a case of autoimmune type 2 polyendocrinopathy diagnosed due to premature ovarian insufficiency.

Case report: The 37-year-old female patient presented to her endocrinologist for exploration of secondary amenorrhea after cessation of her contraceptive 3 months earlier. The patient complained of hot flushes, fatigue and weight loss of 9kg over the past 6 months. She had a negative medical history except for autoimmune hypothyroidism diagnosed several years previously. The hormonal work-up revealed primary hypogonadism (low oestradiol, FSH >200UI/l). Anti-ovarian and anti-adrenal antibodies were found to be positive. Further on, adrenal insufficiency was diagnosed on a Synacthen stimulation test and hydrocortisone treatment was started, as was mineralocorticoid substitution. Under treatment, the symptoms that the patient previously described disappeared and her general well-being improved significantly. 2 weeks later, biology revealed a normal FSH. The patient was addressed to the fertility center for rapid ovocyte preservation.

Conclusions: We illustrate a case of type 2 autoimmune polyendocrinopathy that reunites thyroid, adrenal and ovarian targeting by auto-antibodies. An autoimmune etiology is found in less than 10% of all cases of premature ovarian insufficiency. The ovarian disease in this case was found to be fluctuating with a regain of ovarian activity. Our case supports the idea that autoimmune disease is associated with a preservation of the follicular pool. However, the follicular decline is most likely accelerated and therefore, autoimmune ovarian insufficiency imposes a close follow-up in order to allow early ovarian preservation.

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