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Autoimmune Polyendocrinopathy Type 2 (PEA Type) Associated with Biermer's Disease: Report of a Case and Review of the Literature

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Abstract **Case Report**

Autoimmune polyendocrinopathy type II (AIEP II) is a more common syndrome that appears mainly in adults. It combines several autoimmune pathologies including Addison's disease and autoimmune thyroid disease (Schmidt syndrome) and/or type 1 diabetes (Carpenter syndrome). We report the case of a 16-year-old patient with notable familial pathological ATCD: Father followed by high blood pressure under antihypertensive treatment and type 2 diabetes under oral anti-diabetes medication. Mother followed for hypothyroidism under hormone replacement treatment with levothyroxine. The patient was followed for hypothyroidism for 2 years, hospitalized in our training for suspicion of adrenal insufficiency. Clinically the patient was asthenic, she had arthralgia and myalgia with anorexia contrasting an appetite for salt with notion of fasting hypoglycemia without melanoderma or vitiligo, the rest of the clinical examination was unremarkable. Weight: 65 kg; Height: 1.69 m; BMI: 23Kg/m²; TT: 70 cm; Pulse: 64 bpm. BP: 110/60 mm Hg in the right arm standing and lying down. Biological exploration found low 08 h plasma cortisol with high ACTH, high TSH and anti-TPO antibodies, and macrocytic normochromic anemia at 8 g/dl with anti-parietal cell antibodies and intrinsic factor have positive. whose explorations were in favor of Biermer's disease. The diagnosis of PEA type 2 was made. The treatment recommended: hormonal replacement of adrenal and thyroid insufficiency as well as vitamin B12 injections with good clinical and biological progress.

Keywords: Autoimmune polyendocrinopathy, Biermer anemia, Addison's disease, Schmidt syndrome, Adrenal insufficiency.

INTRODUCTION

Autoimmune polyendocrinopathy type II (AIEP II) is a more common syndrome that appears mainly in adults. It combines several autoimmune pathologies including Addison's disease and autoimmune thyroid disease (Schmidt syndrome) and/or type 1 diabetes (Carpenter syndrome).

CASE REPORT

We report the case of a 16-year-old patient with notable familial pathological ATCD: Father followed by high blood pressure under antihypertensive treatment and type 2 diabetes under oral anti-diabetes medication. Mother followed for hypothyroidism under hormone replacement therapy with levothyroxine.

The patient has been followed for hypothyroidism for 2 years, hospitalized in our unit for suspected adrenal insufficiency. Clinically the patient was asthenic, she had arthralgia and myalgia with anorexia contrasting an appetite for salt with notion of fasting hypoglycemia without melanoderma or vitiligo, the rest of the clinical examination was unremarkable. Weight: 65 kg; Height: 1.69 m; BMI: 23Kg/m²; TT: 70 cm; Pulse: 64 bpm. BP: 110/60 mm Hg in the right arm standing and lying down. Biological exploration found low 08 h plasma cortisol with high ACTH, high TSH and anti-TPO antibodies, and macrocytic normochromic anemia at 8 g/dl with anti-parietal cell antibodies and intrinsic factor have positive. whose explorations were in favor of Biermer's disease. The diagnosis of PEA type 2 was made. The treatment recommended: hormonal replacement of adrenal and thyroid insufficiency as well as vitamin B12 injections with good clinical and biological progress.

DISCUSSION

Autoimmune diseases affect approximately 5% of the population. They are defined by the activation of the patient's immune system against its own antigens [1].

Adrenal insufficiency constitutes the initial manifestation of 50% of PEA-II, it appears at the same time as diabetes or thyroid damage in 20% of cases, and it occurs after the other manifestations in 30% of cases. Type 1 diabetes, when present, usually appears before adrenal insufficiency, whereas autoimmune thyroid disease can appear either before, at the same time, or after adrenal insufficiency. The age at which the first elements of the syndrome appear varies greatly, from childhood to a late period of adult life, most often between 20 and 40 years old.

The other endocrine components of the syndrome may be peripheral hypogonadism, more frequently ovarian than testicular, and autoimmune hypophysitis, responsible for dissociated anterior pituitary insufficiency affecting, variably, the corticotropic, thyrotropic and somatotropic sectors. There is no parathyroid insufficiency. Non-endocrine autoimmune manifestations are alopecia and Biermer's disease, less often than in PEA-I, vitiligo, myasthenia gravis, Gougerot-Sjögren syndrome, rheumatoid disease, primary antiphospholipid syndrome, as well as pericarditis or pleurisy [2].

Biermer AB anemia is an autoimmune atrophic gastritis that can be associated with endocrinopathies, particularly autoimmune AEI. This association is difficult to diagnose: the anemic syndrome is often atypical, wrongly attributed to endocrinopathy [3].

It occurs in 15% of patients with autoimmune polyendocrinopathy (PEA) type I and in less than 4% of patients with PEA types II and III. Biermer's anemia can present a pseudo-Addisonian picture which can be a source of confusion or diagnostic error [4].

CONCLUSION

Type II AEPs represent the most common form of organ-specific autoimmune disease associations. Treatment is essentially based on hormone replacement therapy. A subject suffering from an autoimmune disease must be monitored in order to detect in time the outbreak of new diseases which can occur at any time during the course of the disease.

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