AUTOIMMUNE POLYENDOCRINOPATHIES in children and Adolescents

INTRODUCTION

Autoimmune polyendocrinopathies (PEA) is a rare disease characterized by the coexistence of two endocrine autoimmune deficiencies, sometimes with a non-endocrine autoimmune disease associated concomitently of occurrence or metachronous way. Depending on the age of onset of the disease and the characteristics observed, we can distinguish several subgroups.

OBJECTIVE: Search frequency of PEA in children and adolescents and study their phenotype.

POPULATION, METHODOLOGY

This is a retrospective study of children and adolescents with PEA. All patients underwent a clinical examination, paraclinical exploration looking autoimmune diseases. Annual reassessments were performed.

RESULTS

16 cases have been reported: 87.5% (14/16) PEA II, 12.5% PEA I. The age at diagnosis was 4 and 8 years in type I (both male) and 12 years (8-18) in type II (No sex predominance). The diabetes mellitus 1 was revealing PEA II with concomitant discovery (n: 4) or metachronous (n:6) of autoimmune thyroiditis and other autoimmune diseases (adrenal insufficiency n: 2, pernicious anemia n: 1, autoimmune oophoritis n: 3, hypoparathyroidism n: 1) after a mean of 12 years.

Adrenal insufficiency was revealing in type I.

Mucocutaneous candidiasis was present in 2 children with the hypoparathyroidism at diagnosis in the eldest but only appeared three years later in the younger.

No other disease was observed (mean of 6 years). The PEA family was found in 25% Therapeutically, monitoring was difficult with glycemic control poor in all cases of diabetic (Mean H A1C 8.6%), several ketotic decompensation and acute adrenal in case of adrenal insufficiency.

CONCLUSION

PEA is rare in children and are represented by PEA I. Adrenal insufficiency is indicative in the majority of cases. PEA II can appear during adolescence. It combines Addison’s disease and autoimmune thyroid disease and/or diabetes type 1. Illnesses that caused increased morbidity.