CHARACTERISATION OF POLYGLANDULAR AUTOIMMUNE SYNDROME IN ENDOCRINOLOGY PAEDIATRIC CONSULT: CASES REPORTS

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INTRODUCTION

Polyglandular Autoimmune Syndrome is diagnosed when two or more endocrine systems are dysfunctional, along with other non-endocrine immune disorders, specially autoimmune skin diseases.

To describe 1.4-2 casos por 100.000 people for years.

Due to an autoimmune mechanism: the activity of autoantibodies or T activated lymphocytes against organs or Endocrine glands.

These include tipe 1 diabetes, vitiligo, Addison´s disease, pernicious anemia, candidiasis, hypothyroidism, alopecia hyperthyroidism, hyperparathyroidism, and other diseases whose etiology is most often autoinmune.

The polyglandular autoimmune syndromes (PAS) (table 1) comprise a wide spectrum of autoimmune disorders

and are divided into a very rare juvenile (PAS type I) and a relatively common adult type with (PAS II) or without adrenal failure (PAS III).

First clinical manifestation of PAS I usually occurs in childhood, whereas PAS II mostly occurs during the third and fourth decades. PAS I is caused by mutations in the autoimmune

regulatory (AIRE) gene on chromosome 21 and is inherited in an autosomal recessive manner.

Mutations in the AIRE gene result in defect proteins which cause autoimmune destruction of target organs by disturbing the immunological tolerance of the patients.

OBJECTIVES

To describe the patients include simultaneous deficiencies in the function of several endocrine glands and other non-endocrine problems in two institutions of Cali Colombia

MATERIAL AND METHODS

A study case series with 73 patients with include simultaneous deficiencies in the function of several endocrine glands and other non-endocrine problems in the Pediatric Endocrinology service Clinic foundation Club Noel and Valle del Lili foundation in the city of Cali Colombia.

CLINICAL CHARACTERISITCS

Total patients were 73. Female patients 40 (54.7 percent) and male patients 33 (45.3 percent).
Table 2.

PATOLOGIES	NUMBER PATIENTS	%
DIABETES	53	72
HYPOTHYROIDISM	27	37
VITILIGO	12	16
ALOPECIA	9	12
IDIOPATHIC ARTHRITIS	5	7
BOCIO	3	4
POLIMYOSITIS	1	1,3

RESULTS

The asociation more frequents were: Table 3.

POLYENDOCRINOPATHY	NUMBER PATIENTS	%
Diabetes + Hypothyroidism	11	15
Hypothyroidism + Vitiligo	11	15
Hypothyroidism + Alopecia	4	5.4
idiopathic arthritis + Diabetes	2	2.7
Hypothyroidism, Bocio + vitiligo	2	2.7
Hypothyroidism + juvenil idiopathic arthritis	2	2.7
hypothyiroidism, Alopecia, Vitiligo, dermatomyositis and juvenil idiopathic arthritis	1	1.3
Hypothyroidism ,Alopecia + Vitíligo	1	1.3
Candidiasis, Alopecia +Hypothyiroidism	1	1.3
Vitiligo + alopecia	1	1.3
Dermatomyosistis +Vitiligo	1	1.3

Characteristics of the polyglandular autoimmune syndromes (PAS). Table 1

	PAS TYPE I	PAS TYPE II
Prevalence	Very rare	Relatively common
Incidence	1:100 000/year	1–2:10000/year
M/F	3:4	1:3
Onset	Childhood	Childhood through adulthood
Inheritance	Monogenic (AIRE gene)	Poligenic
Autoimmune endocrine diseases	Hypoparathyroidism (80–85%)	Thyroid disease (70–75%)
	Addison's disease (60–70%)	Type 1 diabetes (50–60%)
		Addison's disease (40%
	Hypogonadism (12%) H thyroid disease (10%))	Hypoparathyroidism (3%) Hypopituitarism (0–2%)
Concomitant disease	Mucocutaneous candidiasis (70–80%)	No candidiasis
Non-endocrine diseases Immune	gastritis, pernicious anemia, celiac disease,immune hepatitis, vitiligo, alopecia areata, Sjo ¨ gren's syndrome, systemic lupus erythematodus, rheumatoid arthritis, myasthenia gravis	

DISCUSSION

Several anomalities endocrine dysfunction may be associated and simultaneously causes various organs in the same patient, the most frequent in this study were diabetes with hypothyroidism, vitiligo and alopecia hypothyroidism.

The importance of this study is sensitize the physician in the search of patologhy, because patients are children and can pass 20 years from diagnostics of an endocrinopathy and the appearance of another disease autoinmune.

After that is necessary an active searching of associations to get the diagnosis of a possible syndrome.
A correct diagnosis give as more information, to improve the treatment and start a correct familiar following, specially when it is proven a genetic heredity as in this case.

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