

What is the profil of gigantism?:7 observations

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Introduction

Gigantism is a condition characterized by excessive growth and height significantly above average; it is usually caused by a tumor on the pituitary gland(1) . In some cases the condition can be passed on genetically through a mutated gene.

Aim:

Describe clinical, hormonal and morphological profile of 7 children with gigantism.

Patients et méthodes

Patients

7 patients (2F/5 H) were retrospectively reviewed.

- Mean age = 14 years (09-16).
- Age of onset of symptoms= 12 years .
- Tall stature (50%),
- brain tumor syndrome (50%).
- Discrete dysmorphic syndrome (n=1).
- Delayed puberty (n = 4).
- A McCune Albright syndrome (n = 1)
- no signs of MEN type 1

Methods :

Laboratory studies used in the diagnosis :

of gigantism :OGTT/growth hormone (GH) and IGF-I

of hypopituitarism: cortisol ,ACTH,FT4,TSH,FSH,LH,E2,TESTO,PRL

❖Imaging studies include Magnetic resonance imaging (MRI): To image pituitary adenomas

Résultats

The diagnosis is late : In our series 50% were diagnosed after a brain tumor syndrome(2) .

Hormone - diagnosis: OGTT / GH and IGF1 which must be greater than 2DS because this rate can be high in case of large family size(3)

the Treatment is difficult and requires a combination of several treatment arms. In addition there is no indication of AS before age= 15years .

The genetic study is required.

Discussion

- Average of GH =110µui/ml and high IGF1 .
- Hyperprolactinemia (n = 2),
- gonadal failure (n = 3) ,
- hypothyroidism (n = 1) ,
- normal cortisol level (n = 7).
- Hypothalamic-pituitary - MRI: locally invasive pituitary adenoma (n = 4) and aggressive (n = 2).
- Treatment: surgery (n = 6) ,second surgery (n = 3) , surgery+ radiotherapy (n = 4) , surgery + radiotherapy + somatostatin analogs (AS) (n = 3) , surgery + AS (n = 1 case) , AS (n = 1) .

CONCLUSION

Gigantism is a rare disease of the child. A monitoring of growth allows early diagnostic and early treatment.