ADENOMAS PITUITARY IN CHILDREN

Introduction

Pituitary adenomas are rare in children. Most of them are found in adolescents. Macroadenomas and secreting adenomas are the most common. They can be sporadic, familial, belong to tumor syndromes and be associated with distinct genetic defects.

OBJECTIVE

Report phenotypic and genotypic characteristics of pituitary adenomas in children

Population, methodology

8 children with pituitary adenoma were identified in 20 years. All underwent clinical examination, paraclinical Assessment (hypophysiogramme, MRI ± CT, Ophthalmic bilan ) and a genetic study (Menine, AIP, FIPA)

RESULTS

Average age was 7 ± 0.4 (6-11) years. Sex ratio F / G: 1.5. Tumor syndrome is constant with ophthalmological disorders in 70% and gigantism in one case. The causes are: adenoma prolactin : 6; Somatotropic adenoma: 2. The tumor lesion is large in all cases: mean tumor height: 26 mm ± 0.8 (18- 45) Endocrine evaluation revealed dissociated anterior pituitary insufficiency n: 100% (GH: 7, TSH: 8, ACTH: 5) The genetic investigation revealed NEM 1 n: 4. The search for the AIP gene and FIPA was negative for the others cases.

Discussion

Pituitary adenomas are rare in children and adolescent; They represent 3% of intracranial tumors. The secreting adenomas are most common; In order of frequency, we have prolactinomas representing about 50à70% of pituitary adenomas, corticotroph adenomas (Cushing's disease): 20% and somatotropic adenomas: 10%. In children and adolescents, tumor syndrome is the main reason for consultation, followed by hypogonadism. The hypersecretion of GH in children, can cause gigantism resulting in adulthood by an exceptionally large stature. If hypersecretion starts at the end of growth, it can cause acromegaly dysmorphic syndrome. The ACTH hypersecretion resulting hypercortisolism manifested by obesity with hypertension, diabetes and delay stature
The phenotype of pituitary adenomas is typical; tumors are larger and more invasive. Therefore, the therapeutic management can be difficult and laborious.
Genetically, we can find a mutation in some cases, particularly the AIP gene or menin gene cause of these adenomas

conclusion

Pituitary adenomas are exceptional in pre pubertal period. They are in large majority, characterized by clinical and genetic diversity; They must be diagnosed and treated precociously because of their locoregional agressivity