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IgG4-related hypophysitis: A case report
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Introduction

IgG4-dependent disease is a newly defined fibroinflammatory disease. This disease has been shown to affect almost all organs in the body, especially the pancreas, salivary gland, orbital tissue, lymph node, lung and kidney. IgG4 hypophysitis is a rare and inflammatory process that mimics pituitary tumors. The diagnosis of IgG4-related hypophysitis can be done in many ways. The definitive diagnosis is biopsy; however, it is not necessary in most cases. If the other organs are diagnosed with IgG4-related disease with biopsy, the appearance of pituitary mass is also diagnostic.

Case

A 28-year-old woman was admitted to our clinic with blurred vision and double vision during her pregnancy. Our patient was receiving replacement therapy due to total hypophyseal insufficiency. In June 2018, pituitary biopsy was performed and the pathology report was revealed as IgG4 pituitary. In November 2018, outward shifting started in the right eye. The patient presented with a Gamma-knife recommendation. TSH: 0.026 (N: 0.27–4.2 IU/ml), free T4: 0.836 (N: 0.93–1.7 ng/dl), FSH: 1.24 (N: 3.5–12.5 mIU/ml), LH: 0.4 (2.5–11.2 IU/ml), E2 (3 pg/ml), Prolactin: 0.466 (N: 4.79–23.3 ng/ml), IgG: 340 (N: 80–140 mg/dl), Pituitary MRI revealed bilateral cavernous sinus, optic chiasm and infundibulum infiltration, and T2 hypointense hypophysitis. The patient was diagnosed with IgG4-releated hypophysitis and pulse steroid treatment was started, but because there was no clinical and radiological improvement, rituximab 1000 mg was given twice daily for 15 days.

Conclusion

There is still no consensus about the treatment of IgG4-releated hypophysitis, and in patients with steroid resistance, rituximab therapy may be required.

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Trunk fat increase is prevented both in patients undergoing long-lasting continuous r-hGH therapy and in those who discontinued r-hGH compared to untreated patients: results from baseline data of the MAGHD study
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Background

Adult growth hormone (GH) deficiency (AGHD) is related with alterations in body composition, increased abdominal and visceral adiposity, decrease in lipid and carbohydrate metabolism, and reduction of bone mineral density (BMD). Aim

To compare baseline outcomes concerning body composition and biochemical/hormonal data among adult patients with AGHD referring to a single endocrinological center and grouped according to their history of r-hGH therapy.

Methods

The Management of Adult Growth Hormone Deficiency Study (MAGHD) is a prospective, single-center trial aiming to improve AGHD management through a smartphone app (MAGHD App: Manage AGHD) and a wearable device (MAGHD study) compared to untreated patients: results from baseline data of the MAGHD study. Up-to-now, a total of 74 patients during first generation SSA treatment before pasireotide LAR start. Results

Mean duration of AGHD disease was 121.2 ± 105.8 months; it was lower in Group 3 compared to both Group 1 and 2 (P < 0.05). Waist circumference (Group 1 95.1 cm; Group 2 96.5 cm; Group 3 109.0 cm), total fat mass (Group 1 25013g; Group 2 26491g; Group 3 33887g) and trunk fat mass (Group 1 11743g; Group 2 12598g; Group 3 16761g) were significantly higher in Group 3 compared to both Group 2 and 1 (P < 0.05). While total fat mass and trunk fat mass did not differ significantly among Group 2 and 3, serum insulin (%) were significantly higher in Group 3 than Group 1 (P < 0.05). IGF-1 and IGFBP3 were significantly higher in Group 1 compared to both Group 2 and 3 (P < 0.0001).

BMD, circulating lipids, and fasting glucose did not differ among the 3 groups.

Conclusions

r-hGH therapy seems to confer a long-lasting beneficial effect on body fat, especially trunk fat even after its discontinuation in AGHD patients, but not on metabolic parameters.

Acknowledgment

This clinical study is conducted thanks to the competitive assignment of an Independent Grant for Learning & Change (‘IGCL’): Dissemination & Implementation (‘D&I’) by Pfizer Inc.

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Hyperglycemia and pasireotide lar in acromegaly: a study with continuous glucose monitoring
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Background

Pasireotide LAR is a multireceptor targeted somatostatin analogue that has been shown to obtain a better biochemical control of acromegaly. However, pasireotide LAR could induce hyperglycemia in acromegalic patients with higher baseline glucose values. The devices that can track interstitial glucose levels such as continuous glucose monitoring (CGM) could be a useful for studying the impact of SSA on patients’ glucose status.

Aim

We aimed to study the glucose metabolism with CGM in a group of acromegalic patients during first generation SSA treatment before pasireotide LAR start.

Methods

We studied 10 patients with uncontrolled acromegaly (Male 5; median age 58y) in therapy with first generation SSA eligible for the treatment with pasireotide. At pasireotide start (T0) we performed a CGM of 9 days to investigate the glucose variability (J whole, GRADE, MAGE, CONGA). We also collected endocrinological and metabolic data (GH, IGF 1, fasting plasma glucose -FPG-, HbA1c) at T0 and after at least 3 months of therapy (T1).

Results

Analysis of the data, revealed a significant decrease in GH (T0 2.06 vs T1 1.02 ug/L, P < 0.01) and in IGF1 (T0 275.5 vs T1 193.5 ug/L, P = NS) after treatment with pasireotide, with a median treatment duration of 9 months. There was also a significant increase in FPG and HbA1c (FPG T0 97 vs T1 124 mg/dl, P 0.01; HbA1c T0 41.5 vs T1 44.5 mmol/ml, P 0.01) At T0, 5 patients (50%) had glycemic alterations: 2 patients had diabetes mellitus (DM) in therapy with metformin and 3 patients had an impaired fasting glucose (IFG). At T1, 5 patients (50%) received antidiabetic medications and among this 60% started antidiabetic treatment after T0. Patients with FPG >100 mg/dl at T0 showed higher glucose variability for most important CGM-based variability indexes (FPG <100 vs FPG >100: J whole 17.3 vs 25.2 P < 0.03, GRADE 2 vs 4 P < 0.03, MAGE 54.7 vs 96.1 P < 0.01, CONGA1 12.8 vs 28.3 P < 0.03).One patient discontinued the drug due to severe hyperglycemia (>500 mg/dl). At T1 there were no significant correlations between HbA1c/FPG and glycemic indexes.

Conclusions

We confirm the efficacy of pasireotide and the effect on glucose metabolism. For the first time in literature we found higher glycemic variability indexes in acromegalic patients with known alterations of glucose metabolism. Further studies are needed to determine the role of CGM in acromegalic patients on pasireotide treatment.

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