AUTOIMMUNE ENCEPHALOPATHY IN A BOY WITH GRAVES' DISEASE.

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INTRODUCTION

Autoimmune encephalopathy is reported in some patient with Hashimoto's thyroiditis and in Graves' disease is extremely rare, especially in children. Sensitiveness to steroid therapy is very characteristic and therefore is known also as steroid-responsive encephalopathy. The diagnosis is difficult because of lack of specific markers and symptoms. The clinical, laboratory and radiologic findings associated with Hashimoto's encephalopathy are varied. In about 50% of patients focal or diffuse nonenhancing MR imaging abnormalities as well as the elevation of antithyroid antibodies and CSF protein increase are observed.

We report a boy of 15 years with Graves' disease treated ineffectively with thyrostatics. After 2 years of the therapy he had recurrence of hyperthyroidism and underwent radical treatment with ablative dose of ^{131}I. Two weeks after he started to suffer from tachycardia and weakness, increasing sleepiness and progressive qualitative and quantitative disorders of consciousness: agitation, psychosis, transient stupor. Body temperature was normal. Laboratory examinations revealed severe thyrotoxicosis and increase of antithyroid antibodies in the blood (Anex 1, The first episode).

Thyroid storm was suspected (Tab. 1) and typical treatment was administered, including steroids (Anex 2)

Symptoms of thyrotoxicosis and consciousness disorders gradually decrease until complete withdrawal. The dose of Metimazol was systematically decreased and after 6 weeks he did not need the thyrostatic therapy. The patient developed hypothyroidism and substitution with left- thyroxine was introduced. For next three months he maintained euthyroid in a good condition. After this time disorders of consciousness appeared again similarly to the previous episode. The patient was admitted to the hospital. Electroencephalography, CT and MR of the CNS were normal. The cerebro-spinal fluid laboratory evaluation was within normal reference values. In the serum high concentrations of antithyroid antibodies were found without hormonal dysfunction (Anex 1, The second episode).

Taking into account the encephalopathy associated with autoimmune thyroid disease, steroid therapy was administered: first intravenous pulses and than oral doses (Anex 3). The fast improvement was observed with complete withdrawal of neurological symptoms for 8 days.

Lack of thyroid dysfunction during the second episode, high levels of antithyroid antibodies, exclusion of CNS infectious diseases and very good response to steroid therapy confirmed the diagnosis of autoimmune encephalopathy probably triggered by acute injury of thyroid tissue after ^{131}I treatment.

Patient was treated with steroids for 8 months in decreasing doses without the recurrence of symptoms. During three years follow-up he remained in complete remission.

Anex 1

Laboratory evaluation

Before radioiodine therapy:

TSH 1,7 mIU/L fT4 - 0,92ng/dl (N: 0,8-1,8) fT3 - 2,39pg/ml (N: 1,45-3,48)
Anti TPO 705,3 IU/ml (N<12) Anti Tg 1059,5 IU/ml (N<34) TBII 2,66 IU/L (N:<1,75)
Thyroid volume 27 ml.

The first episode:

CRP 1,8mg/dl [n<1,0] WBC 3,600, lymph 30,5% gran 50,4%
Glucose 136mg/dl GOT 36 U/L[N: 15-40]; GPT 56U/L [N: 10-45]
TSH 0,015 fT4: 5,58ng/dl (N: 0,8-1,8) fT3: 24,57ng/dl (N: 1,45-3,48)
Anti TPO: 368,1 IU/ml (<12) Anti Tg: 664,1 IU/ml (<34) TBII 12,3 IU/L
EEG- normal CT of CNS and CSF punction- not done: not compliant patient, anaestesia contraindicated in thyrotoxicosis.
MR of CNS in the 5th day - normal

The second episode.

CRP<0,05mg/dL WBC 5,200/ uL lymph 35% gran 52,8%
TSH 4,3 mIU/L fT4 0,9 ng/dL fT3 1,99 pg/mL
Anti TPO 1500,1 IU/mL Anti Tg 2657,0 IU/ml

Toxicological urine examination- negative.
CSF examination: within normal ranges, pleocytosis 0
EEG- normal, CT of the brain- without focal changes and intracranial hypertention.
MRI of CNS: normal

Anex 2.
Treatment during the first episode.

Favistan (MMI) i.v. 80 mg, and next 40 mg every 6 hour
Hydrocortison i.v. 4 x 100mg
Propranolol 3x 20mg
Mannitol i.v.
Acyclovir i.v.
General treatment: liquid balance, HR, BP monitoring.

Anex 3.
Treatment during the second episode.

1. Steroid pulses: intravenous infusion methylprednisolonum 1,0g for 5 days
2. Prednisonum 60mg for 30 days, and next 30 days the dose reduction to 50%.
3. Slow gradual dose reduction to complete withdrawal.
4. General treatment: liquid balance,HR, BP monitoring.

Table 1.
Diagnostic criteria for the risk of thyroid storm in the patient.

Thermoregulatory dysfunction		Cardiovascular dysfunction	
Temperature 36,7°C	0	Tachycardia 120 to 129	15
Central nervous system effects	20	Atrial fibrillation	0
Psychosis		Heart failure	0
Extreme lethargy			
		Total score	**35**
Gastrointestinal-hepatic dysfunction	0	Conclusion: thyroid storm possible	

Based on: Burch HB, Wartofsky L. Metab Clin North Am 1993

CONCLUSION: The autoimmune encephalopathy associated with autoimmune thyroid diseases is effectively treatable syndrome and should be considered in patients with AITD and neurological symptoms. It can be diagnosed even the serum sensitive TSH level and inflammatory markers are normal, the cerebrospinal fluid profile does not suggest an inflammatory process, and neuroimagiing resuts are normal.

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