CLINICAL CASE OF MULTIPLE ENDOCRINE NEOPLASIA (MEN) TYPE 1

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Introduction

Thyrotropin (TSH)-secreting pituitary adenoma (thyrotropinoma) is a rare (0.5% of all pituitary adenomas) and usually benign pituitary lesion, arising from the monoclonal expansion of neoplastic thyrotrophs. Thyrotropinoma usually presents with symptoms thyrotoxicosis (milder compared to those originating from the primary thyroid disorders) and mass effects of the pituitary tumor.

The aim

To describe the clinical course of thyrotropinoma presented with paroxysmal atrial fibrillation with high sensitivity to somatostatin analogs.

Case report

In a 53 y.o. man the disease manifested in 2001 at the age of 41 years with paroxysmal atrial fibrillation and hypertension effectively medically treated with beta-blockers. The paroxysms were well controlled but became a lot frequent since December 2013. The hormonal profile showed elevated levels of TSH - 4.3 μU/ml (0.25-3.5), FT4 – 23.56 pmol/l (9.0-20.0), FT3 – 7.63 pg/ml (2.5-5.5), and normal levels of IGF-1, ACTH, cortisol, LH, FSH. Thyroid autoantibodies were negative. Ultrasound revealed diffuse changes without increasing the thyroid gland size, MRI demonstrated a pituitary macroadenoma 18*12*11 mm with para(D)-sellar extension. Test with short acting octreotide showed normalization of TSH, FT4 and FT3 levels during the first week of treatment. Subsequent therapy with octreotide LAR was started with dose 10 mg once in 28 days with further induction of euthyroidism, decrease of vertical size of pituitary adenoma on MRI at 5 months of medical treatment. Subjectively, the patient noted an improved health with disappearance of atrial fibrillation paroxysms.

In 2014 the patient underwent a transnasal adenectomy. The immunohistochemical analysis of the removed tumor showed positive staining for TSH, GH, SSTR2 and SSTR5 (pic. 1). One week after surgery, TSH was suppressed. Postoperative laboratory tests one month after surgery confirmed euthyroidism: TSH – 2.21 μU/ml (0.4-4.0), FT4 -11.1 pmol/l (9.0-20.0), FT3 -5.3 pg/ml (2.3-6.3).

Conclusion

The TSH-secreting adenoma is a rare cause of hyperthyroidism. Diagnosis is usually delayed due to milder and nonspecific clinical picture, because of that patients can be managed by cardiologists for long periods of time. Surgery is still the mainstay of treatment, although somatostatin analogs may be effectively used as medical therapy which is reflected by expression somatostatin receptors in the adenoma.