P3 – 191

Hormone-secreting pituitary adenomas in children and adolescents

[1]Akulevich N., [2]Zhurauliou V., [1]Kuzmenkova E., [3]Solntsava A.
[1]State Center of Medical Rehabilitation, [2]Republican Research and Clinical Center of Neurology and Neurosurgery, [3]Belarusian State Medical University, Minsk, Belarus

ESPE 2019 VIENNA
58TH MEETING 19-21 SEPTEMBER 2019

Background and Objective

Hormone-secreting pituitary adenomas in children and adolescents are rare.

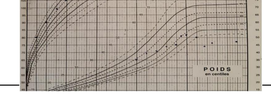
Methods

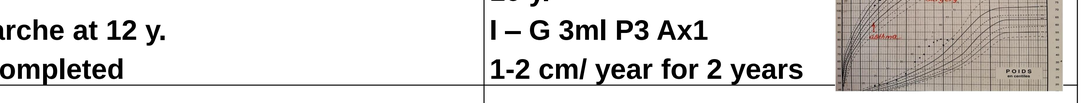
We report clinical course, management and outcome of 6 cases diagnosed in 2013-2019: 3 ACTH-secreting adenomas (ACTHA) and 3 prolactinomas (PROLA). The family history for endocrine tumors was negative in all patients. All ACTHA cases and 2 PROLA girls have been operated with endoscopic endonasal transsphenoidal surgery (EETS).

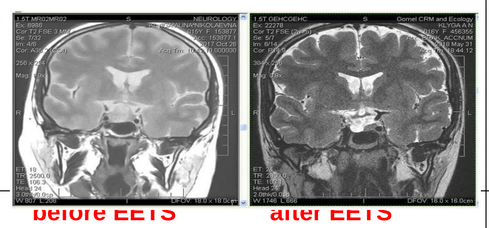
Results

Table 1 Characteristics of the patients with ACTHA

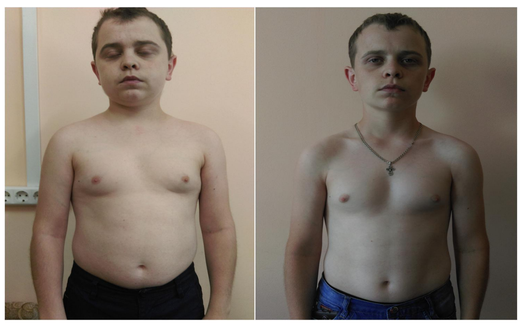
Age at the 1st examination/ / gender	Pt. 1 14 0/12, M	Pt. 2 15 4/12, F	Pt. 3 10 10/12, M
Complaints	weight gain, growth/puberty arrest, high BP, back pain	weight gain, skin rush, striae, irregular menstrual cycle	weight gain, growth arrest, mood changes
Height, cm/ SDS Weight, kg/ SDS MPH, cm/ SDS BA, y. Tanner stage GV	140 cm / -2,5 SDS 44 kg / -0,5 SDS 181 cm/ +1 SDS 11 y. II – G4ml P3 Ax3 1-2 cm/ year for 3 years	161 cm/ -0,5 SDS 68 kg / more + 3 SDS 163,5 cm (median) NA V – menarche at 12 y. growth completed	131 cm / -1,5 SDS 50 kg / more + 3 SDS 186cm (+2 SDS) 10 y. I – G 3ml P3 Ax1 1-2 cm/ year for 2 years
Lab exam and visualisation: serum cortisol and ACTH 2 x FUC 1 mg overnight Dexamethasone test Pituitary MRI Adrenal CT ACTH at separating blood sampling from sin. cavernosus, petrosus inferior Dexa	 impaired circadian secretion 850 and 900 (ref. 50-190 mcg/24 h) no cortisol suppression Negative - no tumor seen Negative Right/ left gradient = 43 Zs = -5,4; no fractures	 moderately elevated NA no cortisol suppression Macroadenoma (right) 13*17*26mm Negative NA Normal	 high, impaired circadian secretion 798 and 1649 (ref. < 485 nmol/ 24h) no cortisol suppression Microadenoma 6*6*7mm Negative NA Normal
Age at EETS, y. Follow-up	18 0/12 21 9/12 - H 149cm, W 47kg Tanner st V, no hormonal replacement	15 5/12 17 3/12 - H 162 cm, W 53 kg Levothyroxin 37,5 mcg Regular menstrual cycle	10 11/12 12 6/12 - H 147 cm, W 49 kg, GV 12 cm/year Hydrocortisone 12 mg/m2/day Tanner st II – G8ml; BA 11 9/12







ACTHA patients before and after pituitary surgery



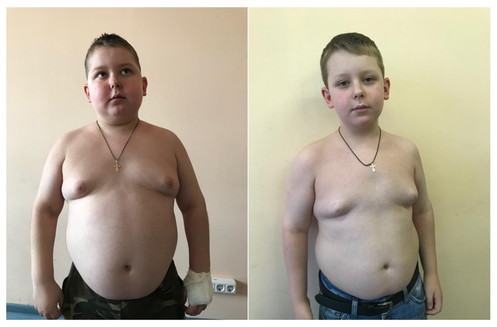


Table 2 Characteristics of the patients with PROLA

Age at the 1st examination/ gender	Pt. 4 11 6/12, F	Pt. 5 14 0/12, M	Pt. 6 16 4/12, F
Complaints	haedache, visual impairment, puberty delay	monolateral pthosis (OD), haedache, puberty delay, tachicardia, tremor	Haedache, primary amenorrhoea
Height, cm/ SDS; Weight, kg/ SDS MPH, cm/ SDS BA, y. Tanner stage	153 cm / > +3 SDS; 40 kg / > +3 SDS 174 cm/ +2 SDS 10 y. Tanner I – B1 P1Ax1 (prepubertal)	165 cm / +0,9 SDS; 51 kg / >+ 0,9 SDS 178 cm /+1 SDS 13 y. Tanner II – G4ml P1Ax1	162,5 cm / median; 51 kg / median 162cm / median NA; Tanner III – primary amenorrhoea
Lab exam and visualisation: serum PRL TSH, FT4, IGF-1 Pituitary MRI Initial treatment time of treatment PRL under DA Additional conservative treatment	 2500 (ref. 102-496 uIU/ml) Nl Macroadenoma 19*22*28mm, parasellar growth, invasion into right sinus cavernosus, chiasma compression dopamine agonists (DA) 6 mg/wk x 2 years 1800 to 2800 uIU/ml	 > 10000 (ref. 102-496 uIU/ml) TSH 6,78 (0,27-4,2 uIU/ml), FT4 40,15 (12-22 pmol/l), TSH-R-AB- negative Giant adenoma 38*61*39mm with endo-, ante-, infrasellar growth and compression of the chiasma DA 1,5 mg – to 3,5 mg/wk x 4 years 4000 to 1000 uIU/ml Sandostatin 30 mg/mo	 138000 (ref. 79-347 mIU/l) TFTs – Nl, IGF-1 initially elevated Macroadenoma 59*32*51mm, invasive growth, chiasma compression EETS – partial resection of the tumor 10000 to 389 uIU/ml DA 2 mg/wk x 6 mo
Follow-up	14 5/12 - H 170,3cm, W 58kg Levothyroxine 50 mcg Tanner st. III	18 6/12 - H 174 cm, W 60 kg, Tanner st IV – G20ml DA 3,5 mg/wk Sandostatin 30 mg/mo	17 4/12 Tanner st DA 2 mg/wk No osteoporosis by DEXA no HRT

Conclusion

Pituitary adenomas represent a challenge for diagnosis and follow-up in children and adolescents. A good collaboration between pediatric endocrinologists, neurosurgeons and other specialists of the team can improve clinical outcomes of such patients.

The authors have nothing to disclose. Correspondence for Dr. Natallia Akulevich: natamedical@mail.ru

References: 1. *Locatelli D. et al. 2019 Child's Nervous System.*
1. Storr H., Savage M. 2015 Eur. J Endocrinology.
2. Kim Y. et al. 2018 Endocrine J.