

Search for DHEASism and DHEASoma

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Introduction

Recently we described the clinical case¹:

- 36-year-old woman was referred because of the secondary amenorrhea, hirsutism, weight gain, decreased sexuality and epileptic seizures.
- On CT tumor in the right adrenal region was discovered.
- High increase of DHEAS and testosterone was observed.
- The symptoms of the disease disappeared immediately after surgery and last until now (24 months).
- The goal of the study was to find more adrenal DHEAS secreting tumors cases.



Methods

DHEAS-centered investigation for DHEASism / DHEASoma.

Search of increased DHEAS according to the lists of laboratory investigations at Kaunas and Vilnius university hospitals during 1 year – 2014.

Women were referred for investigation of DHEAS because of hirsutism, menstrual cycle disturbances and infertility.

Search for medical records with high DHEAS.

Search of clinical, laboratory and imaginative data from medical records.

Results

Increased DHEAS was considered, when the patient's DHEAS concentration was higher than the maximal value of DHEAS for age, indicated in the assay kit.

Ratio $\frac{\text{DHEAS patient}}{\text{DHEAS maximal value}} > 1$ was found in 11.76% of women referred for the investigation.

Mild increase was in 87.4% ($\frac{R}{\text{DHEAS}} < 1,3$), moderate increase was in 9.8% ($1,3 < \frac{R}{\text{DHEAS}} < 2$),

high increased was in 2.8% ($\frac{R}{\text{DHEAS}} > 2$).

Search of 18-50-year-old woman of DHEAS determinations in 2 university hospitals during 1 year – 2014

1215 Patients (885 from Kaunas and 330 Vilnius)

146 patients with increased DHEAS

100 medical records

56 medical records with the right data

4 patients with increase of DHEAS

3 patients with adrenal adenoma

1 patient with no adrenal adenoma

We managed to analyze outpatient files of 56 patients, that have R/ DHEAS >1.

Results of age, height, weight, BMI and DHEAS in the whole group are shown in the table 1.

	R/DHEAS >1				
	Age	Height	Weight	BMI	R/DHEAS
Mean	26,32	168,55	80,38	27,87	1,22
Standard Deviation (SD)	6,12	5,76	21,08	7,18	0,24
Count (n)	56	56	56	56	56

Table 1. Phenotypic data in 56 patients with R/DHEAS>1

Conclusion

DHEAS-centered investigation of 1215 women, referred for DHEAS investigation because of the hirsutism, menstrual cycle disorders or infertility, revealed that the dehydroepiandrosteronism (DHEASism) is found frequently - in 11, 8 %. On contrast to that, the dehydroepiandrosteroma (DHEASoma) was found rarely - only in 3 patients. More prospective investigations using not only the determination of DHEAS, but also the clinical findings would be performed for detection of DHEASoma.

Compared the results of patients with R/DHEAS <1,3 and R/DHEAS > 1,3

	Age		Height		Weight		BMI		R/DHEAS	
Mean	27,1	24,37	169,16	167,03	81,70	77,09	28,29	26,82	1,10	1,52
Standard derivation (SD)	6,59	4,30	5,98	5,02	21,71	19,70	7,45	6,57	0,11	0,21
Count (n)	40	16	40	16	40	16	40	16	40	16

Table 2. Phenotypic data of patients with R/DHEAS<1,3 and R/DHEAS>1,3

■ R/DHEAS <1,3

■ R/DHEAS >1,3

3 patients with increased DHEAS and adrenal adenoma.

Patient No.	Symptoms	Age	Weight	High	BMI	R/DHEAS	CT
1. (I.G.)	Hirsutism, weight gain	33	98	173	32	1,435	Tumor in the left adrenal region
2.(J.P.)	Hirsutism, secondary amenorrhea	27	65	172	21,5	1,3	Left adrenal hyperplasia
3.(J.R.)	Hirsutism, acnea	31	68	165	24,97	1,46	Tumor in the left adrenal region

Table 3. Phenotypic data of patients with adrenal adenoma

Case report about one patient with increased DHEAS, but without adrenal tumor:

- 37 year-old woman referred because of inability to get pregnant, primary amenorrhea;
- Obj.: High-152 cm, Weight -78 kg, BMI- 34,2 kg/m², short neck, small breasts, multiple pigmented moles, large ears.

Hormones

- 17-OH-P - 2,82nmol/l
- DHEA-S - 26,8 μmol/l
- Testosterone- 6,05nmol/l
- SHBG - 23 nmol/l (LAI 26,3)
- LH- 11,3 U/l
- FSH- 29,4 U/l
- Progesterone- 6,2 nmol/l
- Prolactin - 130 mU/l
- FT4 - 10,52 pmol/l;
- TSH - 4,63 mU/l;
- Anti-TPO - 948,08 kU/l
- Cortisol - 386 nmol/l
- ACTH - 3,2 pmol/l
- E2 - 327 pmol/l

Karyotype blood test showed the typical Turner syndrome with 70% of 45 X0 and the rest is the mosaicism with Y chromosome.

CT: Both adrenal glands without additional derivative.

Gynecologist consultation:

Labia minora are visualized, but the labia majora are absent. Clitoris is enlarged and the uterus is small.

US: uterus 2,89 x 1,40 x 4,70 cm; Thread-like endometrium. Right ovary - 0,8 x 0,5 cm. Left ovary – not visualized.

Diagnosis: Turner syndrome; Ovary and labia majora dysgenesis; Primary amenorrhea; Hypoplastic uterus; idiopathic DHEASism; Autoimmune thyroiditis; Eutirois.



Discussion

The DHEASism is diagnosed often, especially when polycystic ovary is diagnosed^{2,3,4}. When search for DHEASism/DHEASoma is centered on DHEAS investigation, during one year we were able to suspect 3 patients with DHEASoma, which is a rare disease^{5,6,7}. Retrospective analyses are not so informative, due to the fact that the medical records are frequently very bad documented. Prospective analyses should be done in multinational and multicentral studies.

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