A PROSPECTIVE STUDY OF ETIOLOGIES OF HIRSUTISM AND SCREENING FOR NON CLASSIC CONGENITAL ADRENAL HYPERPLASIA DUE TO 21 HYDROXYLASE DEFICIENCY IN 93 FEMALE ADOLESCENTS

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Abstract - In this study 93 hirsute females (Ferryman and Gallway score > 8) refered to institute of endocrinology & metabolism and office were assesed. Baseline testostrane (Tes), Dehydroepiandrosterane sulfate (DHEA-SO4). Follicular stimulating hormone (FSH), leutcinizing hormone (LH), prolactin (PRL), thyroid stimulating hormone (TSH), baseline 17 hydroxyporgestrone (17 OHP) and in some cases ACTH stimulation test for screening of non classic congenital adrenal hyperplasia (NC-CAH) due to 21 hydroxylase deficiency was done. The ovaries were visualized by ultrasonography in 56 patients, of the 93 hirsute female, 5 had NC-CAH (5.4%), 46 patients diagnosed as having polycystic ovary syndrome (49.5%) and 42 cases diagnosed as having idiopathic hirsutism (45.1%) Acta Medica Iranica 38 (4): 238-239; 2000

Key Words: Idiopathic hirsutism, polycystic ovary syndrome (PCOS), Non classic hyperplasia (NC-CAH)

INTRODUCTION

Excessive adrenal or ovarian secretion of androgen or excessive conversion of androgens in peripheral tissues leads to histutism (3). PCO syndrome and idiopathic hirsutism are the most common causes of hirsutism (4) Hirsutism due to 21-Hydroxylase deficient NC-CAH appears between 1% and 2% and in some population up to 10% (5).

This study is done in order to determine the etiologies of hirsutism and screening for 21 OH deficient NC-CAH in 93 hirsute females.

MATERIALS AND METHODS

In a prospective study from July 1997 to July 1999, 93 female patients with hirsutism (Ferryman and Gallway score > 8) have been seen in endocrine clinic and office.

The age of the patients was from 14 to 19 years. The degree of hirsutism was assessed scored by the method of Ferriman and Gallway (F.G score) by an Endocrinologist. A total score 8 or more accepted as hirsutism. The height, weight and BMI were measured and answered about mense irregularity and history of hirsutism and drug consumption. Baseline testostrone, DHEA, SO4, FSH, LH, prolactin, TSH, cortisol and 170 H progestrone were measured in all patients. 170 H progestrone measurement was done in the morning during follicular phase and in 14 patiens that 17-OHP level was greater than or equal to 200 ng/dl, ACTH stimulation test was done and baseline 17-OHP and cortisol and stimulated 17-OHP and cortisol after 30 minutes 250 μg short act ACTH (Cosyntropin) iv bolus injection were measured for screening of 21-Hydroxylase deficient NC-CAH during follicular phase. The ovaries were visualized by ultrasonography in 56 patients.

RESULTS

Of the 93 female patients who were studied, 21 cases (22.5%) had mild hirsutism, 56 cases (60.2%) had moderate hirsutism and 16 cases (17.3%) had severe hirsutism (Table 1).

They were from 14 to 19 years. Of the 93 patients 46 cases (49.5%) were diagnosed as having pcos, 42 cases (45.1%) had idiopathic hirsutism and 5 cases (45.1%) had 21-Hydroxylase deficient NC-CAH diagnosed with rapid ACTH stimulation test and comfirmed with clinical response to low dose dexamethazone (25-0.5% mg at bed time) that

diminshed significantly the clinical manifestation of hyperandrogenism in all-5 cases (Table 2).

Presenting complaints other than hirsutism in our patients were in decreasing of frequency: Obesity 78 (83.8%), irregular mense: 51 (54.8%), Acnea 26 (28%) and Alopecia 2 (2%), (Table 3). Four patients out of the 5 cases with NC-CAH had short stature (SDS > -3).

Table 1. Degree of hirsutism in 93 female adolescent

Degree	FG Score	No.	Percent
Mild	8-12	21	22.5%
Moderate	12-18	56	60.2%
Severe	> 19	16	17.3%
Total		93	100

Table 2. Etiologies of hirsutism in 93 female adolescent

Etiology	No.	Percent
PCOS	46	49.5%
idiopathic	42	45.1%
NC-CAH (21-Hydroxylase)	5	5.4%
Total	93	100%

Table 3. Clinical manifestation of hyperandrogenism in 93 female adolescent

Clinical manifestation	No.	Percent
Hirsutism	93	100%
Obesity	78	# 83.8%
Mense	51	54.8%
irregularity Acnea	26	#28%
Alopecia	2	2%

DISCUSSION

Hirsutism is defined as the prescence of excessive growth of hair in location where hair growth in women is normally minimal or absent (1). Hypertrichosis must be differentiated from hirsutism that is an androgen independent growth of hair that is vellus porminent in non sexual area (2). Androgen production in women has three separate sources, the ovaries, adrenal glands and periferal tissues (the largest of which is skin) (4).

The effect of androgen on female skin include: acnea, hirsutism and alopecia (4). The degree of hirsutism scored by the method of Ferriman and Gallwey (the nine body area that posses androgen sensitive pilosebaceous unit are graded from 0 (no terminal hair) to 4 (frankly virile) and then summed. Normal hirsutism score is less than 8. A total score 8 or more by definition is hirsute. A total score of 8-12 is compatible with mild hirsutism, 12-18 = moderate hirsutism and FG score > 19 = severe hirsutism. PCOS and idiopathic hirsutism are the most common causes of hirsutism. Idiopathic hirsutism is a familial condition and the patients had no menstrual irregularity and normal serum Testostrone and DHEAS.

In our study, and other study done in shirazs University by Dr. omrani and coworkers, the most common causeswere PCOS (49.5%) and idiopathic hirsutism (45.1%) that is compatible with international references. The incidence of 21-Hydroxylase deficient NC-CAH in our study was 5.4% which shows that the incidence of this type of hirsutism may be higher in Iran than other parts of the world, perhaps because of the high occurence of family marriage in Iran suggest that we should think.

The results suggest that we should think about the non classic form of congenital adrenal hyperplasia (Late onset) especially 21-Hydroxylase deficient type in female adolescents with hyperandrogenism camplaints (irregular mense, hirsutism, acnea and alopecia) that can be confirmed with ACTH test. We do not recommend the routine use of the ACTH test as a screening method for NC-CAH in all hyperandrogenic patients. This test should only be performed when the morning follicular unsuppressed 17-OHP level is greater than or equal to 200 ng/dl (6 nmol/L).

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