

Premature Adrenarche in a 4 year Old Nigerian Girl: A Case Report with Review of Literature

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Abstract: *Premature adrenarche refers to an early increase in adrenal androgen production that usually results in the development of pubic hair or pubarche before the age of 8 years in girls and 9 years in boys with or without axillary hair and pubertal odour, and with no other signs of sexual development. We report a case of 4 year old girl who presented with complaint of increasing pubic hair noticed from birth with subsequent development of axillary hair and body odour from 2 years of age. Physical examination revealed a body mass index of 16.3kg/m² and pubic hair at Tanner stage 2. The biochemical results were within the reference limits for age and sex except for low FSH and LH. Based on the clinical and biochemical features a diagnosis of premature adrenarche was made. The parents were counseled and the child is currently on follow - up in the clinic. Premature adrenarche though a benign condition, in girls with LBW, requires follow - up due to higher risk of obesity - related metabolic disturbance.*

Keywords: Premature adrenarche, four year old girl, Paediatrics, Case report, Nigeria

1. Introduction

Adrenarche refers to a maturational increase in the secretion of adrenal androgen precursors (AAPs) in mid - childhood, occurring typically at around 5 - 8 years of age. The main AAPs are dehydroepiandrosterone (DHEA) and its sulfate (DHEAS). The clinical signs of adrenal androgen action are normally seen after the age of 8 and 9 years in girls and boys, respectively. Premature adrenarche (PA) refers to the presentation of androgenic signs - appearance of pubic and axillary hair, adult type body odour, oily hair, acne or comedones before the age of 8 years in girls or 9 years in boys in the absence of central puberty, steroidogenic enzyme defects, androgen - producing tumours, or exogenous source of androgens, concurrently with circulating AAP concentrations above the usual low prepubertal level^{1, 2, 3, 4, 5}. Premature adrenarche occurs with increasing frequency between the ages of 3 and 8 years although it may present as early as 6 months of age. It is usually of great concern to the child, the parents, or the primary care physician, who may mistake it for the onset of true precocious puberty. It is not clear if premature adrenarche is as a result of an exaggerated production of Dehydroepiandrosterone (DHEA) due to early zonarecticularis development or an early finding of steroidogenic abnormalities. Several studies have shown an association between this premature androgen excess with low birth weight, metabolic syndrome in children and polycystic ovary syndrome (PCOS), suggesting that a pre - natal condition such as low birth weight could be followed by excessive weight gain during childhood, which could lead to a pattern of events culminating with metabolic complications and consequently to premature adrenarche, metabolic syndrome and PCOS^{6, 7}.

Premature adrenarche is a diagnosis of exclusion and conditions such as true centrally mediated precocious puberty, congenital adrenal hyperplasia, exogenous androgens exposure and androgen secreting tumours must be

ruled out. This case is to illustrate a clinical presentation of premature adrenarche in a 4 year old Nigerian girl.

2. Case Report

T. A is a 4 year old girl who presented with complaint of increasing pubic hair noticed from birth with subsequent development of axillary hair and body odour from 2 years of age. There was no history of associated breast enlargement or use of androgen containing creams on her. She was a product of term gestation with birth weight of 2.5kg and no family history of similar condition. Physical examination findings revealed mild hypertrichosis on the back and extremities with normal anthropometric indices for her age (height of 105cm and weight of 18kg with a BMI of 16.3kg/m²). Her breast development is at prepubertal stage while the pubic hair is at Tanner stage 2 with no associated clitoral enlargement. A diagnosis of premature adrenarche was entertained and she was investigated in that line. She had normal serum electrolytes but hormonal assay revealed a low level of FSH & LH (Table 1). Abdomino - pelvic scan done showed normal sized uterus for age.

Table 1: Showing the hormonal assay done at presentation

Parameters	Patient values	Reference values
DHEAS	62.6 ug/dl	15 - 345 ug/dl
Testosterone	< 9.0 ng/dl	14 - 76 ng/dl
Cortisol (a. m)	8.7 ug/dl	3.7 - 19.4 ug/dl
17 - Hydroxyprogesterone	69 ng/dl	<100 ng/dl
Oestradiol	0.01	
FSH	36.7mIU/ml	
LH	0.6mIU/ml	
Progesterone	0.29ng/ml	

FSH= Follicle Stimulating Hormone

LH= Luteinizing hormone

DHEAS= Dehydroepiandrosterone sulphate

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Figure 1: Showing the axillary hair



Figure 2: Showing the pubic hair at tanner stage 2

3. Discussion

The clinical signs of premature adrenarche include oily hair and skin, adult - type body odour and the appearance of pubic and axillary hair^{1, 2, 3, 4, 5}. The androgenic signs in PA often manifest in a typical order but with varying circulating androgen levels between patients.^{3, 8} Serum DHEAS is considered the best marker of adrenal androgen secretions and its concentration are higher in patients with premature adrenarche.³ Androgens are a prerequisite for the growth and development of the pilosebaceous unit (PSU) in

“sexual” areas of skin.^{9, 10} As DHEAS rises >40 to 50 µg/dL adrenal androgens successively initiate sebaceous gland development, apocrine gland development, and pubarche in young children. This is above that of children <6 years of age and similar to that of early pubertal girls of a similar pubic hair stage.¹ The PSU consists of a prepubertal vellus follicle in which the hair and sebaceous components are virtually invisible to the naked eye. In acne - prone skin areas, androgen causes the prepubertal vellus follicle to develop into a sebaceous gland. The index patient had a normal prepubertal DHEAS value of 62.6µg/dl which is not uncommon in a child with signs typical of premature adrenarche.^{3, 5} The PSUs of sexual hair areas respond to increasing androgen levels by increasingly producing a thicker (terminal) hair follicle. Estrogens also seem to directly promote the growth of sexual hair to a small extent.^{9, 10}

The serum levels of progesterone, 17 - OH progesterone and cortisol ruled out any form of congenital adrenal hyperplasia. The serum concentrations of FSH, LH, oestradiol, progesterone and the absence of thelarche do not support any likelihood of precocious puberty.

Premature adrenarche is a benign condition that does not require treatment. However, because it is a forerunner of syndrome X in girls with low birth weight,¹¹ there is need for follow up. In some instances it may predict functional ovarian hyperandrogenism in adolescence. Her parents were counseled on the benign nature of the condition and the need for regular long - term longitudinal follow - up, and the use of deodorants to help control the body odour.

4. Conclusion

Premature adrenarche though a benign condition in girls with low birth weight usually seems to be an extreme variation of normal, requires follow up due to increased risks for obesity, insulin resistance, and possibly mood disorder and polycystic ovary syndrome.

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