British Journal of Dermatology (1993) **129**, 447–448.

Hidradenitis suppurativa as a presenting feature of premature adrenarche

F. LEWIS, A.G.MESSENGER AND J.K.H.WALES*

*University Departments of Dermatology and *Paediatrics, Sheffield Children's Hospital, Western Bank, Sheffield S10 2TH, U.K.*

Accepted for publication 19 April 1993

Summary We report a girl with premature adrenarche, in whom the presenting feature was hidradenitis suppurativa. This association lends support to the view that hidradenitis suppurativa is an androgen-dependent disorder.

Case report

A girl, aged 7·8 years, presented to the dermatology clinic with a 10-month history of episodic painful swelling and redness in the groins. She had previously been well, and was not taking any medication. Her parents had noticed that her perspiration had an 'adult odour'. Her father had a history of recurrent pilonidal sinuses.

On examination, she was obese (weight on the 97th centile), and her height was on the 50th centile. There was induration of both groins, with several comedone-like lesions. Pubic hair stage 3 and axillary hair stage 2 were present, and she had mild clitoromegaly and increased labial pigmentation. There was no breast development.

A diagnosis of hidradenitis suppurativa was made, and she was referred for endocrinological assessment. An ultrasound scan of the abdomen and pelvis was normal. TW2 bone age was calculated at 9·6 years, at a chronological age of 7·8. The results of hormonal investigations showed a normal serum 17-hydroxyprogesterone (17-OHP) of 3 nmol/l, and a mildly elevated serum testosterone of 0·9 nmol/l. A 24-h urinary steroid profile showed an excess of androgen and 17-OHP metabolites. In order to exclude late-onset congenital adrenal hyperplasia secondary to atypical 21-hydroxylase deficiency, stimulation tests with ultra-low and standard-dose adrenocorticotrophin (ACTH) were performed. If this condition is present, there is a markedly elevated response of 17-OHP to stimulation by ACTH. In our patient, both tests gave normal results (Table 1).

Her sister, aged 5·1 years, has a height and weight on the 97th centile, a TW2 bone age of 6·3 years, and stage 2 pubic hair growth, with no other signs of sexual precocity, and no evidence of hidradenitis suppurativa. She also has a generalized over-excretion of adrenal steroid metabolites in her urine.

The conclusion was that these children have benign premature adrenarche. The older child was treated with topical clindamycin on the groins twice daily, and the lesions improved within a few weeks. Occasional courses of treatment are required to deal with recurrent lesions.

Correspondence: Dr F.Lewis, Department of Dermatology, Royal Hallamshire Hospital, Glossop Road, Sheffield S10 2JF, U.K.

Discussion

Premature adrenarche is defined clinically as the development of sexual hair before the age of 8 years in girls, and 9 years in boys.[1] It is thought to be due to early maturation of the zona reticularis of the adrenal cortex, which leads to an increased production of androgens.[2] It occurs predominantly in girls between the ages of 3 and 8 years, and the presence of adult axillary odour appears to be a characteristic finding.[3]

The signs can be mistaken for the onset of true precocious puberty, but may also represent the first

Table 1. Results of Synacthen stimulation tests

Time (min)	Cortisol (nmol/l)	Testosterone (nmol/l)	17-OHP (nmol/l)
Low-dose (500 ng)			
0	702	<0·7	3·7
20	953		7·4
60	711		2·5
Standard-dose (250 μg)			
0	273	<0·7	1·8
20	779		6·2
30	852		6·8
60	973		8·0

features of androgen-secreting tumours or non-classical congenital adrenal hyperplasia, especially if other signs of virilization are present. The non-classical types of congenital adrenal hyperplasia due to 21-hydroxylase deficiency can be variable in their presentation. Patients with mild forms of 21-hydroxylase deficiency present with hirsutism, menstrual irregularity or infertility. There is an elevated response of 17-OHP to ACTH stimulation in this condition.[4]

There has been debate about whether all patients with premature adrenarche may have evidence of late onset congenital adrenal hyperplasia.[5] Adrenal androgen levels have been found to be higher in children with premature adrenarche than in normal prepubertal children, but similar to those found at puberty.[6] Exaggerated 17-OHP responses to ACTH have also been found in some children with premature adrenarche,[5,7] but the significance of this finding is unclear, and long-term follow-up is required to see if there is an increased incidence of androgen excess after puberty in this group of patients. Generally, patients with premature adrenarche do not have other features of androgen excess, although in one series of 18 girls with premature adrenarche, five had severe acne requiring treatment.[8]

Hidradenitis suppurativa is a chronic suppurative disorder of the regions bearing apocrine glands. The areas can become indurated and scarred, and there is sinus formation in severe cases. The underlying mechanisms of hidradenitis suppurativa are unknown, but there is evidence that the condition is androgen-dependent in adults.[9] Some patients have been shown to have increased total testosterone concentrations and free androgen indices (testosterone:sex hormone binding globulin) compared with normal controls,[10] but many women with hidradenitis suppurativa have normal androgen indices. It is unclear, therefore, if the disorder results from excess circulating free androgens, or whether there is an abnormal response of the target organ to androgens.

It is of interest that our patient's father has recurrent pilonidal sinuses. These may be associated with hidradenitis suppurativa,[10] and it has been suggested that pilonidal sinuses are an example of perianal hidradenitis suppurativa.[11] There is evidence that the transmission of hidradenitis suppurativa in some families is consistent with single gene dominant inheritance, with 34% of first-degree relatives affected in 11 families in one study.[12] However, in the same study, there was a negative family history in nine families and familial occurrence only in a further three families.

Hidradenitis suppurativa as a presenting feature of premature adrenarche is very unusual, and lends support to the view that hidradenitis suppurativa is an androgen-dependent disorder.

References

1 Silverman SH, Migeon C, Rosemberg E, Wilkins L. Precocious growth of sexual hair without other secondary sexual development: 'premature pubarche', a constitutional variation of adolescence. *Pediatrics* 1952; **10**: 426–32.

2 Korth-Schutz S, Levine LS, New MI. Evidence for the adrenal source of androgens in precocious adrenarche. *Acta Endocrinol* 1976; **82**: 342–52.

3 Kaplowitz PB, Cockrell JL, Young RB. Premature adrenarche. *Clin Pediatr* 1986; **25**: 28–34.

4 Miller WL. Congenital adrenal hyperplasias. *Endocrinol Metab Clin North Am* 1991; **20**: 721–49.

5 Morris AH, Reiter EO, Geffner ME *et al.* Absence of non-classical congenital adrenal hyperplasia in patients with precocious adrenarche. *J Clin Endocrinol Metab* 1989; **69**: 709–15.

6 Korth-Schutz S, Levine LS, New MI. Serum androgens in normal prepubertal and pubertal children and in children with premature adrenarche. *J Clin Endocrinol Metab* 1976; **42**: 117–24.

7 Granoff AB, Chasalow FI, Blethen SL. 17-Hydroxyprogesterone responses to adrenocorticotrophin in children with premature adrenarche. *J Clin Endocrinol Metab* 1985; **60**: 409–15.

8 Voutilainen R, Perheentupa J, Apter D. Benign premature adrenarche: clinical features and serum steroid levels. *Acta Paediatr Scand* 1983; **72**: 707–11.

9 Ebling FJG. Hidradenitis suppurativa: an androgen-dependent disorder. *Br J Dermatol* 1986; **115**: 259–62.

10 Mortimer PS, Dawber RPR, Gales MA, Moore RA. Mediation of hidradenitis suppurativa by androgens. *Br Med J* 1986; **292**: 245–8.

11 Jackman RJ, McQuarrie HB. Hidradenitis suppurativa: its confusion with pilonidal disease and anal fistula. *Am J Surg* 1949; **77**: 349–51.

12 Fitzsimmons JS, Guilbert PR, Fitzsimmons EM. Evidence of genetic factors in hidradenitis suppurativa. *Br J Dermatol* 1985; **113**: 1–8.