INCONTINENTIA PIGMENTI

The case report by Purohit et al (Ind J Dermatol Venereol Leprol 1995; 61 : 295-6) has an omission of earlier case reports from India itself.[1-4] The references may please be added to update.

Rakesh Bharti
Amritsar

1. Handa F, Aggarwal RR, Sharma SC, et al. Incontinentia pigmenti - a case report with review of literature. Ind J Dermatol Venereol Leprol 1975; 41: 63-5.
2. Pavithran K, Ramachandran P, Zochraih J. Incontinentia pigmenti. Ind J Dermatol Venereol Leprol 1984; 50: 274.
3. Giharupuray MB, Joshi MB. Naik SV, et al. Incontinentia pigmenti stage II. Ind J Dermatol Venereol Leprol 1987 ; 53 : 122-3.
4. Bharti R, Bal MS. Incontinentia pigmenti. Ind J Dermatol Venereol Leprol 1995 ; 61 : 166-7.

21 HYDROXYLASE DEFICIENCY MANIFESTING WITH ACNE

To the Editor,

Delayed adrenal hyperplasia is one among the aetiology of hirsutism. These patients may have enzyme deficiencies like 11 beta hydroxylase, 3 betahydroxysteroid dehydrogenase and 21 hydroxylase (in 95% of cases).[1]

A 35-year-old woman came with acne of recent onset, not responding to conventional therapy. She gave the history of oligomenorrhoea and secondary infertility. There were closed comedones, papules and a few cystic lesions on her cheeks. The hairs on the upper lip were coarse. General dermatological and systemic examinations were normal. Endocrine evaluation revealed high levels of free testosterone (FT) : 4.9 pico gm/ml and leutinizing hormone : 15.84 m IU/ml. The dexamethasone suppression test was positive as FT had fallen to 1.3 pico gm/ml at the end of 48 hours of O.5mg of dexamethasone tab qid. Routine blood, urine, ultrasound and pelvic examinations were normal.

In the synthesis of adrenocortical hormones, 21 hydroxylase is the enzyme that converts 17 hydroxy progestone to cortisone. In the absence of this enzyme, the biochemical precursors are diverted towards androgen pathway. Therefore the level of androgen ie, FT is elevated. There is also a decrease in the level of cortisol. ACTH secretion is increased. The level of 17 ketosteroids, the metabolic end product of cortisol, in the urine may be normal or high. When dexamethasone is given, due to the negative feed back mechanism the cortisol production is reduced, so also the accumulation of its precursors. Hence the level of FT will come down. As the dexamethasone suppression test was positive in this case, the most common aetiology is 21 hydroxylase deficiency. Clinically delayed onset of congenital adrenal hyperplasia may resemble polycystic ovary syndrome. However dexamethasone suppression test is negative in the case of later.[1]

S R Narahari
Kasaragod

Reference

1. Williams GH, Dluhy RG. Diseases of adrenal cortex. In: Wilson JD, Braunwauld, eds. Harrison's principles of internal medicine. New york : McGraw-Hill, 1991: 301-4, 1713-35, 1776-95.

ACUTE REVERSIBLE HEPATIC TOXICITY BY TRIMETHOXY PSORALEN

To the Editor,

Psoralens are known to cause hepatitis[1] which occurs due to chronic cumulative toxicity of these drugs. Psoralens are metabolised in