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.

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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]

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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

liver and severe impairment of hepatic functions is considered a contraindication to PUVA therapy.[2] We report a case of acute reversible hepatic toxicity by trimethoxy psoralen (trioxsalen).

A 45-year-old Hindu female presented with vitiligo extending to chest and legs. After complete evaluation and baseline investigations like haemogram, blood sugar and LFT, she was given trimethoxy psoralen as 10 mg orally daily and was advised exposure to sunlight after 2 hours. After 7 days she came back with loss of appetite, nausea, vomiting and icterus. Psoralen was stopped and LFT done. Bilirubin was 3 mg%, SGOT 58 IU and SGPT 110 IU. Serum alkaline phosphatase was 27.5 KA units. She was managed for acute liver dysfunction with diet and drugs. Three weeks follow-up investigations revealed normal serum bilirubin, SGOT/SGPT and serum alkaline phosphatase. Patient was asked to discontinue treatment for 2 months. This resulted in increase in vitiligenous lesions. On her insistence she was again put on trioxsalen therapy, but within 3 days she developed nausea, and vomiting followed by icterus. Her investigation revealed increased serum bilirubin levels, SGOT and SGPT were 62 and 132 IU, respectively. Serum alkaline phosphatase was 31 KA units. Trioxsalen was stopped. Presently she is under observation and requires alternate therapy for vitiligo.

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References

1. Pariser DM, Wyles RJ. Toxic hepatitis from oral methoxalen photochemotherapy. J Am Acad Dermatol 1980; 3: 248.
2. Wolf K, Honigsmann H. Clinical aspects of photochemotherapy. Pharmacol Ther 1981; 12: 381.

PRIMARY LOCALISED CUTANEOUS AMYLOIDOSIS

To the Editor,

It was interesting to learn about the effectiveness of colchicine in the treatment of primary cutaneous amyloidosis.[1] In the materials and methods the authors state, "Clinical features were so characteristic that other investigations for confirmation of diagnosis were not considered necessary".

Macular amyloidosis can, at best be suspected on clinical examination. It is not easy to differentiate it from other conditions such as lichen planus pigmentosus, frictional melanosis, ashy dermatosis and numerous other conditions which have an interface dermatitis as histopathological finding. Although I do not doubt the clinical ability of the authors, in my opinion amyloid should have atleast been detected by H & E (a difficult task) if not be special stains. I hope the authors publish another paper confirming the efficacy of the drug after establishing the diagnosis of primary cutaneous amyloidosis.

CR Srinivas
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Reference

1. Chakravarthy K, Chanda M. Role of colchicine in primary localised cutaneous amyloidosis Ind J Dermatol Venereol Leprol 1995 ; 61 : 268-9.

REPLY

To the Editor,

We thank Dr Srinivas for taking keen interest in our article. I am putting our clarification as follows :

1) We agree with Dr Srinivas that it is a difficult task to detect amyloid by H & E stain. Amyloid can often be recognised in H & E section provided that it is present in sufficiently large amount.[1] As it in not a confirmatory test