■ REVIEW ARTICLES

Pityriasis Rubra Pilaris: an Update Review

Dr. Y. P. Fung

Social Hygiene Service (Dermatology), Department of Health, Hong Kong

ABSTRACT

Patients with pityriasis rubra pilaris (PRP) can sometimes be difficult to distinguish from those with psoriasis. The lack of pathognomonic markers and specific clinical and histological diagnostic criteria has made studies of PRP difficult to conduct and interpret. At least three different classification systems had been proposed to categorize patients. Those with atypical forms of PRP may represent variants of other ichthyotic disorders. The aetiology of PRP remains unknown. The role of focal acantholytic dyskeratosis as a distinguishing histological feature remains controversial. Systemic retinoid is currently the first line treatment for those with severe disease.

Keywords: *Pityriasis rubra pilaris, review*

INTRODUCTION

Pityriasis rubra pilaris (PRP) is an uncommon erythematous papulosquamous disorder characterized by erythroderma, palmoplantar keratoderma and follicular hyperkeratosis.[1] Although often exhibited at clinical meetings because of its rarity and difficulty in management,[2,3] its etiology remains unknown. This article presents a review of this enigmatic disorder and subject of debate. Recent focus are also discussed.

EPIDEMIOLOGY

Incidence

Using figures from Hong Kong Social Hygiene Service from 1986 to1999, the incidence of PRP was calculated to be 1:25,000.[4] On average, 1.5 patients were seen each year. This is of the same order compared with the Singapore[5] (1.6 case per year), South Africa[6] (1.5 case per year), and Spain[7] (1.6 case per year) series, but in marked contrast with Griffiths' series (4 cases per year).[8]

The variation may be due to racial difference as the incidence of PRP was reported to be closer to 1:50,000 in India.[9] However, these figures are probably incomparable due to differences in methods of patient selection. The lack of uniform clinical and pathological diagnostic criteria for PRP remains one of the major obstacles in studying the disorder.

Sex ratio

PRP was described to occur equally in men and women in a large study.[1] Differences in sex ratio had been reported by others. Classical adult onset PRP was found to be five times commoner in men than women in one 10-year study.[10] Again, differences in patient selection make comparison of study results difficult.

Age at onset

A bimodal pattern was observed with peaks in the first and fifth decades.[5,11] It has been suggested that a bimodal distribution might reflect a protective factor, possibly hormonal, established during puberty.[12]

AETIOLOGY

The cause of PRP is unknown and has long been a subject of debate. A prominent finding is epidermal over-activity, as the thymidine-labeling index is increased from an average normal of 3-27%. These findings may, however, represent epiphenomena to a more basic defect.[13,14]

Correspondence address:

Dr. Y. P. Fung

Yaumatei Dermatological Clinic

12/F Yaumatei Specialist Clinic

143 Battery Street, Kowloon

Hong Kong

Role of genetics

Familial PRP occurs rarely with 0-6.5% of patients having a family history of the disease.[12,15,16] Most of the familial cases belong to PRP Type V (atypical juvenile PRP).[11] Reports of familial cases were initially interpreted as providing evidence of an infectious tuberculous origin[8] but later studies suggested a genetic factor. It is generally inherited as an autosomal dominant trait with variable expression and reduced penetrance.[17] Immunoblot analysis from one family revealed an additional 45-kd acidic keratin (K17) in diseased skin but not in control skin.[17] Further study is required to determine whether this finding is unique to the familial form of PRP. Gelmetti et al.[12] did not encounter any patient with familial PRP over a 20-year period. It had been suggested that the familial cases reported in the literature could represent a different disease. It is possible that they suffered from an atypical ichthyosis in which a follicular element is not uncommon.[11]

Role of vitamin A

Vitamin A deficiency or abnormal vitamin A metabolism has been incriminated as an etiologic entity in PRP for many years. The histology of patients with cutaneous changes secondary to vitamin A deficiency had been reported to be the same as that seen in PRP. This had led to the suggestion that PRP was a manifestation of vitamin A deficiency and prompted early attempts at treatment with vitamin A.[18] However, patients with PRP frequently have normal serum vitamin A levels. This elicited the suggestion that the failure was one of end organ response.[13,19] Finzi et al. found low levels of serum retino-binding protein, a carrier protein for vitamin A, in eleven patients and their relatives but this had not been confirmed by others.[19,20] While abnormal vitamin A metabolism (such as altered intracellular retinol-signaling in the skin) has not been excluded as a possible cause of PRP, it is likely that the effect of vitamin A or synthetic retinoids is secondary to the anti-keratinizing properties of these agents.

Role of Human Immunodeficiency Virus (HIV)

Recently, PRP had been described in association with HIV infection.[21-25] In these reported cases, the onset of PRP occurred shortly after or at the same time when the patient was tested positive for HIV infection. None had developed PRP before HIV infection was confirmed. This led to the suggestion that in these patients, HIV infection was not coincidental but played a pathogenic part. Their responsiveness to treatment with zidovudine[22-24] or triple therapy,[26] and relapse when therapy was stopped, gave further support to this hypothesis.

By 2000, there were at least 14 reported cases. Miralles et al. proposed the designation of a new category of PRP (Type VI) as this type differs from the classical form clinically.[21] The proposed PRP Type VI is characterized by the presence of HIV infection, usually without evidence of immunosuppression, a poor prognosis and poor response to etretinate, and variable associations with lesions of nodulocystic acne, hidradenitis suppurativa and lichen spinulosus. Acne conglobata, hidradenitis suppurativa and lichen spinulosus have also been reported in HIV-infected patients without PRP. As these lesions had not been described in the same patient before the AIDS epidemic, Resnick et al.[27] hypothesized that the constellation of these features represented a genuine HIV-associated follicular syndrome. It was suggested that in genetically predisposed individuals, HIV infection could induce PRP and modify the features of the disease. Misery et al. went one step further and suggested that HIV serology should be included in routine laboratory test in adult patients with PRP, as it had been reported as the first sign of HIV infection.[28] The success of zidovudine in treating PRP Type VI prompted Griffiths to use it for treating three cases of HIV-negative PRP Type I.[29] The result, however, was disappointing. The dose that was used might have been too low or the course too short or perhaps the effect on HIV itself was a more important action of zidovudine in HIV associated PRP.

Documented cases of PRP in patients with HIV have led to renewed interest in a possible underlying immune mechanisms of the disease. It has been hypothesized that the pathogenesis of PRP may be related to abnormal immune response to antigenic triggers. An underlying immunological mechanism for PRP remains an interesting, but still speculative possibility.

ASSOCIATIONS

PRP has been observed in patients with many concomitant non-cutaneous as well as cutaneous disorders.[1] Associated non-cutaneous disorders include:

autoimmune diseases and internal malignancies.[1,18,30] It had also been reported to occur in a patient with multiple cutaneous malignancies.[31] Erythrodermic PRP was reported in a patient with prominent and eruptive seborrhoeic keratoses (sign of Leser-Trelat), but there was no evidence of an underlying internal malignancy. [32]

CLINICAL CHARACTERISTICS

The clinical manifestations of PRP are well described.[11] When presents in its unique florid form with orange-red erythroderma interspersed with islands of sparing, follicular plugging and palmoplantar keratoderma, the diagnosis of PRP can be made readily. However, patients often display less typical features. Insidious onset of red scaly patches on face and torso progressing to erythroderma may mimic other skin diseases, especially psoriasis (Table 1). Repeated consultations and multiple skin biopsies are often necessary for diagnosis.

CLASSIFICATION OF PRP

The taxonomy of PRP has long been a matter of debate. At least three different classification systems had been proposed.[11,12,16]

Griffiths divided PRP into five categories based on clinical characteristics and prognosis.[11] Type I is classical adult onset PRP, with cephalocaudal spread of erythroderma, palmoplantar keratoderma and follicular hyperkeratosis. Approximately 80% of cases resolve within 3 years. Type II is atypical adult onset and differs from Type I based on its longer duration (often greater than 20 years) and atypical morphological features, such as ichthyosiform scale, lamellar scale of the palms and soles, and occasional partial alopecia.

Types III-V are juvenile onset forms of PRP. Type III is classical juvenile onset PRP and appears to differ clinically from Type I only by onset in childhood. Type III was initially thought to have a worse prognosis than Type I, but is now considered to be prognostically favorable.[33] Type IV is circumscribed juvenile onset PRP, the most common type of PRP in children, with well-defined involvement, frequently affecting the knees and elbows. Type V is atypical juvenile PRP, which like Type II, is chronic and has ichthyosiform features. Sclerodermatous changes of the fingers may develop in these patients. Table 2 shows Griffiths' classification with modifications.[33]

It was suggested that the aetiology of classical PRP (Types I and III) was different from the other types of PRP (Types II, IV, and V). The atypical forms of PRP (Types II and V) might represent variants of ichthyotic disorders. The findings that no patient with circumscribed juvenile PRP (Type IV) progressed to classical PRP could suggest a different aetiology.[11] This concept was however put into question with the report of a patient that demonstrated a clear transition from the classical juvenile (Type III) to the circumscribed (Type IV) PRP.[34]

Table 1. Differential diagnosis of pityriasis rubra pilaris

Differential diagnosis of pityriasis rubra pilaris
Psoriasis
Seborrhoeic Dermatitis
Follicular Eczema
Parapsoriasis
Dermatophytosis
Follicular ichthyosis
Secondary syphilis
Lichen planus
Figurate erythema
Mycosis fungoides
Subacute lupus erythematosus

Table 2. Classification of PRP based on Griffiths with modifications (1992)

Griffiths classification of PRP with modifications (1992)				
	Type	% of cases	Distribution	Prognosis
I	Classical adult	55	Generalized	Most clear in 3 years
II	Atypical adult	5	Generalized	Chronic
III	Classical juvenile	10	Generalized	Most clear in 1 year
IV	Circumscribed juvenile	25	Focal	Uncertain
V	Atypical juvenile	5	Generalized	Chronic

Gelmetti et al.[12] did not find a correlation between the extent of disease in juvenile PRP (circumscribed or generalized) and prognosis, with most cases of either type clearing within 1 year. They proposed a classification based on disease duration, in which either localized or diffuse forms could run an acute or chronic course (Table 3).

Piamphongsant and Akaraphant studied 168 Thai patients with PRP.[16] They observed that skin lesions of Griffiths' Type IV were not always confined to children since similar lesions were present in adults. Moreover, in their study, there were no cases belonging to Griffiths' Type V. While acknowledging that many discrepancies could be due to racial difference, they proposed a modified clinical classification. Their proposed classification grouped adults and children together and paid more emphasis on the clinical importance of keratoderma (seen in 92% of cases). In their classification, PRP was divided into four types according to physical findings. This included one type with palmoplantar keratoderma, but without follicular papules or exfoliative dermatitis. It is interesting to note that most of their recruited patients had palmoplantar keratoderma that extended '*beyond the dorsopalmar and plantar junction*'. This is in contrast to the classical PRP 'sandal' described in most studies that typically does not transgress onto the dorsal surfaces.

The issue of PRP Type VI has already been discussed above. It is at present uncertain whether this addition is justified, because this disorder shows many features differing widely form PRP, and may merely represent an HIV-associated follicular syndrome.

The ideal classification of a disorder is based on its cause. In PRP this cannot be achieved until its aetiology is revealed. To date, despite its imperfections, Griffiths' classification remains the most popular system for categorizing PRP.

HISTOPATHOLOGY

The three most common features were alternating orthokeratosis and parakeratosis in both vertical and horizontal directions, focal or confluent hypergranulosis and follicular plugging.[35] These features were however non-diagnostic. The presence of checkerboard arrangement of parakeratosis and orthokeratosis with areas of hypergranulosis had been reported in ichthyosiform erythroderma and erythrokeratodermia progressive symmetrica.[17,36] While a diagnosis of PRP could not be made solely on these findings, they were helpful to rule out other papulosquamous diseases.

Magro and Crowson, in a study of 26 PRP patients, found focal acantholytic dyskeratosis (FAD) in 23 out of 32 skin biopsies.[37] As these were not seen in the skin biopsies of 23 patients with psoriasis, matched for age and site, they argued that the presence of FAD in PRP was more than incidental and could be used as a clue to diagnosis. FAD has been reported in association with a variety of skin lesions. These include benign and malignant epithelial lesions, fibrohistiocytic lesions, lesions secondary to inflammatory conditions, melanocytic lesions, comedones and ruptured follicles.[38] While the definitive origin of incidental FAD is unknown, there has been speculation that sunlight or UV radiation may contribute to its development.[38] No other large published series has yet repeated the findings of Magro and Crowson. Nevertheless, their unique observation, with up to 70% of FAD in PRP specimens, warrants greater awareness of this possible association in future studies.

TREATMENT

Evaluation of the treatment for PRP has been difficult because of its natural remitting course and the lack of a standardized assessment of severity. When reading treatment studies of PRP, one must consider the possibility of erroneous inclusion of psoriasis in the study population, unless clearly defined clinico-pathological diagnostic inclusion criteria for PRP was established.

Systemic therapy

Systemic treatment is usually required in patients with extensive involvement. Although a multitude of systemic therapies were described, controlled trials involving large numbers of subjects were rare.

Table 3. Gelmetti et al.'s classification of juvenile PRP (localized or diffuse)

Gelmetti et al.'s classification of juvenile PRP (localized or diffuse)	
Type	Duration
Acute	<6 months
Acute with prolonged course	<1 year
Chronic	>1 year

Retinoids

Some of the largest studies that achieved the best clinical response involved the use of retinoids. Systemic Vitamin A had been used with considerable effectiveness.[11,32] The advent of synthetic retinoids has largely supplanted vitamin A therapy. Isotretinoin (13-cis-retinoic acid) was demonstrated to be efficacious in a large number of patients. In a multicenter trial, Goldsmith et al.[39] treated 45 PRP patients with isotretinoin. Patients initially received up to four months of treatment with a mean daily dose of approximately 1-2 mg/kg. More than 90% of patients demonstrated significant treatment response as judged by evaluation using a rating scale for global improvement. Borok and Lowe [40] observed that seven out of fifteen patients who received isotretinoin in daily doses of 0.42-2.2 mg/kg for PRP cleared, usually within seven months of starting treatment. Dicken[41] found that 10 out of 15 patients taking 40-80 mg daily cleared with isotretinoin therapy, given on average for 25 weeks

Etretinate was also found to be effective and doses of 0.5-1.0 mg/kg/day were typically used. Borok and Lowe[40] observed complete clearing in three out of four patients after five months of etretinate therapy with a daily dose ranging from 0.27-1.0 mg/kg. They stated that patients treated with etretinate cleared slightly faster than those in the isotretinoin group. Dicken[41] found that four out of six patients receiving an initial dose of 50-75 mg of etretinate per day cleared, with an average clearing time of eight months.

Methotrexate

Although its use has been largely supplanted by synthetic retinoids, it remains an excellent second-line drug for the treatment of PRP.[5,41] Dicken treated eight patients with methotrexate for an average of six months, all of whom showed significant improvement on 10-25 mg/week.[41]

Phototherapy

The results of phototherapy in patients with PRP are much less dramatic than in psoriasis. Ultraviolet B (UVB) treatment seemed to be ineffective[5,41] and was reported to exacerbate PRP.[42] Narrowband UVB resulted in lesional blisters in one case.[43]

Treatment with systemic psoralens and ultraviolet A (PUVA) gave variable results. Most reported a lack of response.[18,41]

While the combination of PUVA and etretinate therapy (Re-PUVA) was reported to be ineffective, Kirby and Watson recently successfully treated a case of juvenile PRP with acitretin and narrow-band UVB.[44]

Other treatments

Other treatments reported in the literature with variable success include stanozolol, azathioprine and cyclosporin A.[13,18,19,45]

FUTURE

A number of issues remained unresolved and future studies on PRP should focus on these particular areas. The first issue refers to the enigmatic aetiology of PRP. Better understanding of its pathogenesis would be invaluable for developing specific diagnostic test or therapy. Its discovery in patients with HIV led to hypothesis that PRP might be related to abnormal immune response to antigenic triggers. Whatever started this disease process, it resulted in abnormal epidermal keratinization, as evidenced by its response to retinoids. Perhaps one of the first studies that could be done was immunoblot analysis of epidermis from non-familial cases of PRP. This would determine if expression of an additional 45-kd acidic keratin (K17), seen in familial PRP,[17] was also present in the more commonly encountered non-familial cases.

The second issue refers to its classification. The relationship of classical PRP (Types I and III) and the other types of PRP (Types II, IV, and V) remains obscure. The atypical PRP (Types II and V) may represent a form of follicular ichthyosis and may ultimately acquire other labels. PRP Type IV stands alone both in its clinical appearance and its behavior. Until specific laboratory investigations or genetic markers are found, the relationship of these diseases to each other will remain obscure. Selecting individual groups for special genetic and keratin studies seems to be the next logical step.

The third issue that requires further study is the role of focal acantholytic dyskeratosis. Multi-center prospective and retrospective studies of histological specimens from unambiguous PRP cases would help to yield the true incidence of FAD in PRP. Its potential role as a histological discriminating feature would be better defined when larger comparative studies with biopsies from control patients were performed.

Lastly, and perhaps most importantly, a standardized diagnostic criteria for PRP has yet to be established. The lack of consistency in patient selection had made comparison of study results between different groups impossible and occasionally meaningless. In order to facilitate discussion and progress in the study of PRP, a standardized, universally accepted clinico-pathological diagnostic criteria should be drawn up and based upon in all future studies. This would provide a uniform standard by which patients and treatments can be evaluated.

References

1. Griffiths WAD. Pityriasis rubra pilaris – an historical approach: clinical features. Clin Exp Dermatol 1976;1:37-50.

2. Lam WS. Pityriasis rubra pilaris or psoriasis? Social Hygiene Service Bulletin 1995;3:59-63.

3. Chan LY. A woman with pityriasis rubra pilaris. Hong Kong Dermatology and Venereology Bulletin 1996;4:240- 3.

4. Fung AY. Pityriasis rubra pilaris: a study of 21 cases in Hong Kong. Dissertation submitted for Hong Kong College of Physicians, 2000.

5. Lim JT, Than SN. Pityriasis rubra pilaris in Singapore. Clin Exp Dermatol 1991;16:181-4.

6. Jacyk WK. Pityriasis rubra pilaris in black South Africans. Clin Exp Dermatol 1999;24:160-3.

7. Sanchez-Regana M, Creus L, Umbert P. Pityriasis rubra pilaris. A long-term study of 25 cases. Eur J Dermatol 1994; 4:593-7.

8. Griffiths WAD. Pityriasis rubra pilaris. A clinical and laboratory study. University of Cambridge, MD thesis 1976;1-180.

9. Sehgal VN, Jain MK, Mathur RP. Pityriasis rubra pilaris in Indians (Correspondence). Br J Dermatol 1989;121:821-2.

10. Clayton BD, Jorizzo JL, Hitchcock MG, et al. Adult pityriasis rubra pilaris: a 10-year case series. J Am Acad Dermatol 1997; 36:959-64.

11. Griffiths WAD. Pityriasis rubra pilaris. Clin Exp Dermatol 1980; 5:105-12.

12. Gelmetti C, Schiuma AA, Cerri D. Pityriasis rubra pilaris in childhood: a long-term study of 29 cases. Pediatr Dermatol 1986; 3:446-51.

13. Griffiths WA. Pityriasis rubra pilaris. In: Champion RH, Burton JL, Ebling FJ, editors. Textbook of Dermatology. Blackwell Scientific Publication, 1998; 35:1539-45.

14. Griffiths WA, Pieris A. Pityriasis rubra pilaris – an auto-radiographic study. Br J Dermatol 1982;1107:665-7.

15. Niemi KM, Kousa M, Storgards K, et al. Pityriasis rubra pilaris: a clinicopathological study with a special reference to autoradiography and histocompatibility antigens. Dermatologica 1976;152:109-18.

16. Piamphongsant T, Akaraphant R. Pityriasis rubra pilaris: a new proposed classification. Clin Exp Dermatol 1994;19:134-8

17. Vanderhooft SL, Francis JS, Holbrook KA, et al. Familial pityriasis rubra pilaris. Arch Dermatol 1995;131:448-53.

18. Albert MR, Mackool BT. Pityriasis rubra pilaris. Int J Dermatol 1999;38:1-11.

19. Goldsmith LA, Baden HP. Pityriasis rubra pilaris. In: Freedberg IM, Eisen AZ, Wolff K, editors. Fitzpatrick's dermatology in general medicine. McGraw Hill 1999;46:538-40.

20. Finzi AF, Altomare G, Bergamaschini L. Pityriasis rubra pilaris and retinol-binding protein. Br J Dermatol 1981;104:253-9.

21. Miralles ES, Nufiez M, De Las Heras ME. Pityriasis rubra pilaris and human immunodeficiency virus infection. Br J Dermatol 1995;133:990-3.

22. Martin AG, Weaver CC, Cockerell CJ. Pityriasis rubra pilaris in the setting of HIV infection: clinical behaviour and association with explosive cystic acne. Br J Dermatol 1992;126:617-20.

23. Blauvelt A, Nahass GT, Pardo RJ. Pityriasis rubra pilaris and HIV infection. J Am Acad Dermatol 1991;24:703-5.

24. Auffret N, Quint L, Domart P, et al. Pityriasis rubra pilaris in a patient with human immunodeficiency virus infection. J Am Acad Dermatol 1992;27:260-1.

25. Menni S, Brancaleone W, Grimalt R. Pityriasis rubra pilaris in a child seropositive for the human immunodeficiency virus. J Am Acad Dermatol 1992;27:1009.

26. Gonzalez-lopez A, Velasco E, Pozo T. HIV-associated pityriasis rubra pilaris responsive to triple antiretroviral therapy. Br J Dermatol 1999;140:931-4.

27. Resnick SD, Murrell DF, Woosley J. Pityriasis rubra pilaris, acne conglobata, and elongated follicular spines: an HIV-associated follicular syndrome? [letter]. J Am Acad Dermatol 1993;29:283.

28. Misery L, Faure M, Claudy A. Pityriasis rubra pilaris and human immunodeficiency virus infection-Type 6 pityriasis rubra pilaris? Br J Dermatol 1996;135:1008-9.

29. Griffiths WA, Hill VA. Zidovudine in HIV-negative pityriasis rubra pilaris. J Dermatol Treat 1997;8:127-31.

30. Sanchez-Regana M, Lopez-Gil F, Salleras M, et al. Pityriasis rubra pilaris as the initial manifestation of internal neoplasia. Clin Exp Dermatol 1995;20:436-8.

31. Tannenbaum CB, Billick RC, Srolovitz H. Multiple cutaneous malignancies in a patient with pityriasis rubra pilaris and focal acantholytic dyskeratosis. J Am Acad Dermatol 1996;35:781-2.

32. Cohen PR, Prystowsky JH. Pityriasis rubra pilaris: a review of diagnosis and treatment. J Am Acad Dermatol 1989;20:801-7.

33. Griffiths WA. Pityriasis rubra pilaris: the problem of its classification [letter]. J Am Acad Dermatol 1992;26:140-2.

34. Shahidullah H, Aldridge RD. Changing forms of juvenile pityriasis rubra pilaris-a case report. Clin Exp Dermatol 1994; 19:254-6.

35. Soeprono FF. Histologic criteria for the diagnosis of pityriasis rubra pilaris. Am J Dermatopathol 1986;8:277-83.

36. Howe K, Foresman P, Griffin T. Pityriasis rubra pilaris with acantholysis. J Cutan Pathol 1996;23:270-4.

37. Magro CM, Crowson AN. The clinical and histomorphological features of pityriasis rubra pilaris: a comparative analysis with psoriasis. J Cutan Pathol 1997;24:416-24.

38. DiMaio DJ, Cohen PR. Incidental focal acantholytic dyskeratosis. J Am Acad Dermatol 1998;38:243-7.

39. Goldsmith LA, Weinrich AD, Shupack J. Pityriasis rubra pilaris response to 13-cis-retinoic acid (Isotretinoin) J Am Acad Dermatol 1982;6:710-5.
40. Borok M, Lowe NJ. Pityriasis rubra pilaris. Further observations of retinoids therapy. J Am Acad Dermatol 1990;22:792-5.
41. Dicken CH. Treatment of classic pityriasis rubra pilaris. J Am Acad Dermatol 1994;31:997-9.
42. Yaniv R, Barzilai A, Trau H. Pityriasis rubra pilaris exacerbated by ultraviolet B phototherapy [letter]. Dermatology 1994;189:313.
43. Khoo L, Asawanonda P, Grevelink SA. Narrowband UVB-associated lesional blisters in pityriasis rubra pilaris [letter]. J Am Acad Dermatol 1999;41:803.
44. Kirby B, Watson R. Pityriasis rubra pilaris treated with acitretin and narrow-band ultraviolet B (Re-TL-01). Br J Dermatol 2000; 142:376-7.
45. Usuki K, Sekiyama M, Shimada T. Three cases of pityriasis rubra pilaris successfully treated with cyclosporin A. Dermatology 2000;200:324-7.

Answers to Dermato-venereological Quiz on page 42

Answer (Question 1)

1. This condition is known as Harlequin fetus.

2. The clinical features illustrated are generalized erythroderma, severe skin fissurings, ectropion of the eyes, eclabium of the lips and deformed ears. The baby born was usually of low birth weight, prematurity and died shortly after birth.

3. Systemic retinoids like etretinate had been used successfully to control the condition. Our illustrated case had been treated with oral acitretin and liberal topical emollients. The side effects of systemic retinoids were regularly monitored.

   Harlequin fetus is a very severe and rare neonatal condition of the skin. Its mode of inheritance is thought to be autosomal recessive. The term Harlequin referred to the diamond-like costume of the fetal skin as a result of the severe fissures and hyperkeratosis. It is usually a fatal condition but systemic retinoids may control the condition. Nowadays, authorities believe that Harlequin fetus is a heterogeneous skin disorder. The baby if survive may evolve into an ichthyosis resembling congenital ichthyosiform erythroderma. Molecular studies suggested that the features of the condition may be due to a biochemical block in the conversion of filaggrin from profilaggrin. Hyperproliferative keratin 6 and 16 had also been identified in the abnormal keratinocytes.

Answer (Question 2)

1. The diagnosis is Naevus lipomatosis superficialis (classical type as described by Hoffman and Zurhelle).

2. Histologically, there is a proliferation of adipose tissues in the dermis of the skin. The adipose tissues surround the blood vessels and the vessels may rise up from the subcutaneous layer and spread out to form the subpapillary plexus.

3. Naevus lipomatosis superficialis should be distinguished from focal dermal hypoplasia. The latter condition is an important genodermatosis which can be associated with widespread ocular, dental and skeletal dysplasia.

   Naevus lipomatosis superficialis is a benign connective tissue naevus of the adipose tissues. It is divided into two types: the classical type is the Hoffman Zurhelle variant which is characterized by unilateral multiple skin colored cerebriform plaque, mostly occurred over the buttock and upper thigh of the patient. The second type is the solitary form in which the lesion appears as domed shaped, single papule. The site of predilection is the arm, axilla, head and neck other than the lower trunk. Both conditions are asymptomatic. No specific treatment is required except surgical excision for cosmetic reasons.