LETTERS TO THE EDITOR

CURE IN PEMPHIGUS–A POSSIBILITY

Recently,[1] we proposed pulse therapy with dexamethasone and cyclophosphamide for pemphigus and observed the following two advantages over the conventional treatment schedules :

(1) The hospital stay of the patients was drastically reduced to 4-5 days in a month, enabling the patient to continue with his job or other social responsibilities, and (2) the side effects of prolonged treatment with corticosteroids and/or immunosuppressive drugs, including osteoporosis, obesity, diabetes etc were virtually absent. Of the 10 patients reported earlier,[1] cases 1, 3, 4 and 9 have been lost to follow up, while the remaining 6 have been followed up for another 2 years or so, the results of which are as follows :

Case 2 had received 10 dexamethasone cyclophosphamide pulses (DCP) till February 1984, and was free from pemphigus lesions since March 1983. During the next 9 months, he continued to take only 50 mg cyclophosphamide orally and remained free of the lesions. In November 1984, he developed a mild relapse and was given DCP again. Since then, he has received 12 monthly DCP and 5 more DCP at approximately 2-month intervals. All through this period, he has been free from pemphigus lesions, till last seen in April 1986.

Case 5 had received 7 DCP till November 1983, and was free from the lesions. He continued to take 50 mg cyclophosphamide orally daily and remained free from the lesions till June 1984. Since then, the disease has got reactivated and till February 1986, he had received 17 more DCP with continued recurrences of the disease. This patient however, has been quite irregular in taking the treatment.

Case 6 had received 14 DCP till March 1984 and had been free from pemphigus lesions since December 1982. In March 1984, she had a mild relapse treated with 20 mg prednisolone a day for a few days. Till February 1985, she received another 8 DCP, but had remained free of pemphigus lesions. Subsequently, she stopped all treatment and has remained free from pemphigus lesions.

Case 7 had received 23 DCP till April 1984, and had been free from pemphigus lesions since August 1983. He continued to receive DCP till November 1984 (29 DCP) after which he continued only 50 mg cyclophosphamide a day orally. In June 1985, he stopped even oral cyclophosphamide and has still been free from pemphigus lesions.

Case 8 had received 9 DCP till April 1984 and was free from pemphigus lesions since January 1983. Till April 1985, he received 5 more DCP (total 14), after which he continued to take only 50 mg cyclophosphamide a day orally till Febraury 1986. He has not had any pemphigus lesions ever since.

Case 10 had received 20 DCP till April 1984, but was continuing to develop recurrences in between the DCP. This continued till November 1984, though the relapses were progressively milder. Between November 1984 and June 1985, he stopped treatment. DCP was started again in June 1985 and after 2 more DCP, he stopped having further lesions. Once again he stopped having further DCP, though 50 mg cyclophosphamide a day was continued. In February and March 1986, he has been given 2 more DCP, but he continues to be free from pemphigus lesions since August 1985.

Thus out of these 6 patients, only 1 patient (case 5) is continuing to have recurrences of pemphigus lesions, while 2 patients (cases 6 and 7) have already given up all treatment for the last 14, 12 months respectively, one patient (case 8) has been taking only 50 mg cyclophosphamide a day for the last 12 months, while two more patients (cases 2 and 10) are still being given DCP; and all these 5 patients are not having pemphigus lesions for periods varying from 10-40 months. It is to be noted that we have so far depended upon only clinical criteria, intercellular antibody results are not yet available with us. This trend of the course of disease is highly encouraging, because there seems to be a hope for cure of the disease. Although a longer follow up of a larger number of cases will be necessary before any conclusions can be drawn, during the last 2 years, 26 more cases of pemphigus have been recruited to this mode of therapy, and a similar trend has been observed in these patients as well. Eleven of these patients are already free from any clinical recurrences, five for periods varying from 6-18 months, and another 6 for periods less than 6 months. The remaining 15 patients are still having clinical exacerbations in between the DCP.

From the observations on these patients, the course of pemphigus with this therapy can be arbitrarily divided into the following 4 phases :

Phase I

The phase of clinical recurrences in between the DCP : During this period, the patient continues to develop lesions after a variable number of days following each DCP. This phase lasts 6 months to 1 year in a majority of the patients.

Phase II

The phase of continued DCP without clinical lesions : This phase follows the first phase, when the patient stops having any clinical lesions. The DCP however, are to be continued at monthly intervals to prevent any relapse of the disease. The duration of this phase has been arbitrarily fixed at a minimum of 6 months, though further experience will be necessary to establish the exact duration.

Phase III

The phase of oral cyclophosphamide only : In the third phase, the DCP are stopped, but the patient continues to take 50 mg cyclophosphamide orally per day as a maintenance. Arbitrarily, a duration of one year for this phase seems to be alright.

Phase IV

Treatment-free follow up : The duration of this phase can be as long as possible, because the longer the duration of treatment-free disease-free follow up, more certain the cure of pemphigus.

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A SPECIAL TECHNIQUE TO TREAT NEVUS UNIUS LATERIS

Nevus unius lateris when located on the exposed parts, poses a great cosmetic problem, particularly in marriageable girls. The treatment so far available includes excision and grafting, dermabrasion, electro-desiccation, chemical peeling and keratolytics, but therapy is often unsatisfactory, recurrences are common with most of these methods. We treated a case of nevus unius lateris with deep electrodesiccation who has not shown any recurrence after a follow up of $1\frac{1}{2}$ years. The patient was a 24-year-old woman, having asymptomatic linear warty lesions extending from the right thumb to the shoulder, from early childhood. The lesion became more prominant with age and started becoming more verrucous after 17 years of age.

All treatments taken earlier resulted in recurrence of the lesions. We used electro-desiccator 755 Blendtoms (Birtcher, California), using mainly cutting loop and needle. Treatment of the entire lesion was completed in five sittings treating an area of 10 cm at each sitting. Two percent xylocaine was used for local anaesthesia. At first, the verrucous hypertrophic portion was desiccated and then the deeper tissue upto the white fascia underlying the lesion was destroyed. Except for slight oozing, there was no bleeding. The treated area was dressed with sofra-tulle, till healing occurred, in 8-10 days. The wound healed with normal scarring. The patient accepted the scar better than the verrucous lesion. No recurrence has been noted during one and half year follow up in the treated area. Part of the lesion on the shoulder and the infraclavicular region was left untreated due to the approaching date of her marriage.

Solomon and Esterly[1] had suggested that the dermis is in some way intimately associated with the pathogenesis of epidermal nevus and unless the underlying dermis and the deep tissue is removed, nevus is likely to regrow. This is probably the reason that recurrences are commonly seen after removal by various methods.

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PSEUDOGRANULOMA INGUINALE/CHANCROIDAL ULCER

It is indeed intriguing to go through 'Pseudogranuloma Inguinale', published in the Ind J Dermatol Venereol Leprol, 1985; 51 : 219-220. I am afraid, the criteria adopted to indicate this condition are rather arbitrary. Apparently, Khare and Bansal seem to have overlooked the essential diagnostic criteria of culturing the *Haemophilus ducreyi*, as advocated by Kraus et al[1] and Werman et al.[2] In its absence, in the present cases, it is indeed hard to accept the capstion.

Should the authors care to look at the recent article by Sehgal and Shyam Parsad,[3] they will be able to sort out the issue in favour of chancroidal ulcers.

Chancroidal ulcers are single, well-defined, soft, tender, non-indurated with weakening edges. Absence of lymphadenopathy is cardinal. These ulcers have a longer incubation period of 8-11 days. They occur in persons of low socio-economic status and in the sexually vulnerable age-group. The prepuce, coronal sulcus and glans penis are the common sites of affliction. It is, however, imperative in such situations, to undertake dark-ground microscopy for *Treponema pallidum*; Gram staining of smear for *Haemophilus ducreyi*; tissue smear for Donovon bodies, and Tzanck smear for multi-nucleated giant cells.

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2. Werman BS, Herskowitz LJ, Olansky S et al : A clinical variant of chancroid resembling granuloma inguinale, Arch Dermatol, 1983; 119 : 890-894.
3. Sehgal VN and Shyam Parsad AL : Chancroid or chancroidal ulcers, Dermatologica, 1985; 170 : 136-141.

REPLY

Thank you very much for your interest in our article and valuable comments. We wish to repeat that our cases had solitary, soft, ele-

vated, bright red, round to irregular and non-tender ulcers without bubo. However, one case had slight tenderness only. Because of these classical clinical features, a provisional diagnosis of granuloma inguinale was made in all the three cases. The crush preparations, Gram staining for *Haemophilus ducreyi*, dark ground examinations, Tzanck smear and VDRL test were negative. When Donovan bodies could not be seen from these granuloma inguinale-like lesions, the possibility of pseudo-granuloma inguinale was suspected. The absence of tenderness and granulomatous nature of the lesions was not in favour of chancroidal ulcers. These cases were then compared with those reported previously and a marked similarity was observed. It is not correct to say that we over-looked the criteria of culturing *H. ducreyi*. In fact we have very clearly mentioned twice in our article that a culture for *H. ducreyi* could not be done because of lack of facilities. We expressed our belief solely on clinical and therapeutic grounds. The main interesting feature in our cases, which prompted us to report, was presentation of granulomatous genital sores resembling granuloma inguinale but not showing Donovan bodies in the crush preparations.

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HYPOHIDROTIC ECTODERMAL DYSPLASIA

This letter is in reference to a case report on hypohidrotic ectodermal dysplasia, Ind J Dermatol Venereol Leprol, 1985; 51 : 229-233. It is stated that dominantly inherited form of hidrotic ectodermal dysplasia is decidedly more common in males. However, no reference is given for this statement. According to Hurwitz,[1] hidrotic ectodermal dysplasia has equal sex distribution. It is also stated that transmission of hypohidrotic ectodermal dysplasia (anhidrotic ectodermal dysplasia) is thought to be autosomal recessive. This statement does not correspond to those reported in standard text books. Hurwitz[1] states that it is an X-linked recessive disorder in which 90% of the patients are males. A much rarer autosomal recessive form of anhidrotic ectodermal dysplasia is described in which males and females present with similar clinical features.[2] At the end of the article, it is stated that the disease in three male siblings can be explained on the basis of autosomal recessive inheritance. I think it can be explained more correctly on sex linked recessive basis. In the clinical features, it is stated that there was nail dystrophy and plantar hyperkeratosis and paronychia, features more often seen in the hidrotic variety. Actually, it is stated that in hypohidrotic (anhidrotic) variety, the nails are usually normal, and the skin of the palms and soles is also normal.[2]

I think it would have been better, if along with the side view, front view of the patients was also included. In the hypohidrotic type of ectodermal dysplasia, facies is distinctive, and as in progeria, unrelated patients look alike. Depressed bridge of the nose is the characteristic which is missing in these patients.

Summing up, I would like to know from my colleagues whether these patients are sweating or not sweating; or are we dealing with the rare form of hidrotic ectodermal defect with autosomal recessive inheritance.[3] Finally, I think much confusion can be avoided by not using the term hypohidrotic, because even though anhidrotic patients do sweat to some extent, hidrotic patients also have reduced sweating. Traditional usage, though not completely correct, atleast avoids confusion.

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2. Carter DM and Keefe EJO : Hereditary cutaneous disorders, in : Dermatology, Vol II, 2nd ed, Editors, Moschella SL, Hurley HJ, WB Saunders, Philadelphia, 1985; p 1211-1213.
3. Verbow J : Hereditary ectodermal dysplasia; in : Current Dermatologic Therapy, Editors, Maddin S, WB Saunders, Philadelphia, 1982; p 210-213.

REPLY

I am grateful to Dr. Gharpuray for the interest he has shown in our cases of hypohidrotic ectodermal dysplasia. One of the objections is to the mode of transmission of hypohidrotic ectodermal dysplasia which we have mentioned as autosomal recessive. I would invite his attention to a review article by Solomon and Keuer[1] in which the authors quoting the work of Freire-Maia,[2] have mentioned the mode of transmission of hypohidrotic type as autosomal recessive and that of anhidrotic variety as X-linked semi-dominant. Because most of the patients have some degree of sweating rather than complete absence, the term hypohidrotic has been suggested by Felsher[3] as quoted in our text. I wish the division between anhidrotic and hidrotic was perfect and complete. Dr. Gharpuray has himself mentioned about an autosomal recessively inherited situation which would equally involve the males and females. It is difficult to presume that our patients belong to that group because they don't have a sister and no other female in the family is involved. I would gladly agree with Dr. Gharpuray that the dermatology text books only mention the mode of transmission as mentioned by him, (unfortunately none of these two text books has given any reference after 1970) except for the situation referred to above. About the clinical features, there was hardly anything for the diagnosis of hidrotic variety to be considered. Reduced sweating with dry, thin, wrinkled skin, almost total absence of hair since birth, hypodontia, nasal mucosal involvement with rhinorrhoea, decreased salivation and skin biopsy showing sparse and rudimentary apocrine, eccrine (only a few normal glands) and sebaceous glands, and ill developed hair follicles all point towards its being more of hypohidrotic than the hidrotic variety. Eye involvement is likely to occur in hypohidrotic variety. Nails are defective (brittle, thin and ridged) in half of the cases of hypohidrotic (anhidrotic) type.[4] Hyperkeratosis was present in the soles only and palms were not involved. Inclusion of front view of the patients was not giving any extra information except that the size of the faces was different according to the age, otherwise they all looked alike.

I would however, agree with Dr. Gharpuray that technically, there would be equal distribution among sexes in the dominantly inherited hidrotic ectodermal dysplasia. Similarly the presence of disease in all males can be better explained by sex-linked inheritence.

Bhushan Kumar

References

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4. Rook A : Genetics in dermatology, in : Text book of Dermatology, Vol I, 3rd ed, Editors, Rook A, Wilkinson DS and Ebling FJG : Blackwell Scientific Publications, Philadelphia, 1979; p 113-118.

Reference my article "Sharma VK, Kumar B, Kaur I and Kaur S : Side lab diagnosis of chromoblastomycosis, Ind J Dermatol Venereol Leprol, 1985; 51 : 157-159," I want to rectify two errors. Firstly, the name of our mycologist Dr. (Mrs.) P Talwar was missed by over-sight. Secondly, the causative fungus reported as *Cladosporium carrionni*, was identified as *Fonsecaea pedrosi V. pedrosi* on subsequent testing. I am very sorry for the inconvenience caused.

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