FOLLICULITIS CRURIS PUSTULOSA ET ATROPHICANS

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Introduction

Follicular diseases resulting in permanent alopecia and atrophic scarring were not unknown to dermatologists. A number of these conditions were described by French dermatologists more than 80 years ago under various names: Pseudopelade (Brooq, 1885) Folliculitis decalvans (Quiqaud, 1888), Acne decalvante (Lailler, 1884), Lupoid sycosis (Brooq, 1888) and epilating folliculitis of the glaborous skin (Arnozan, 1892 and Dubreuilh, 1894). The disease described by Arnozan and Dubreuilh was characterized by therapy resistant symmetrical follicular pustules of both legs resulting in alopecia, atrophy and scarring. Though many more such cases were reported in the European dermatological literature, Clarke (1952) was the first to describe a similar condition in English literature. He reported it to be a prevalent skin disease among the negroes of Nigeria and called it Dermatitis Cruris Pustulosa et Atrophicans (DCPA). Miller (1961) described the first case from North America and suggested the name lupoid sycosis of the legs. Though he reviewed the literature upto 1961, he failed to mention Clarke. Desai et al (1964) studied 30 patients suffering from 'therapy resistant pyogenic folliculitis of the legs' (which was later reported to be identical to DCPA; Desai 1969) and reported the occurrence of hypergammaglobulinaemia in them. After a systematic study of 15 patients from Western Nigeria, Harman (1968) reviewed the pertinent literature and justified Clarke's original assumption of DCPA being a distinct entity. He called it "Nigerian Shin Disease" and gave an excellent clinical description. He called attention to the well defined borders, presence of pitting oedema, equal prevalence in both sexes, occurrence in patients below the age of 30, absence of post inflammatory hypo or hyperpigmentation and a wiry roughness of the hairs in the affected area. It was also pointed out that itching was an inconsistant feature and scratch marks were never seen. He also suggested that local application of vegetable oils had a possible aetiological role.

Symmetrical, pruritic, follicular pustules of the legs associated with varying degree of alopecia and atrophy is a widely prevalent dermatological disorder in Kerala. The striking resemblance of these cases to that of DCPA stimulated a more systematic investigation and the following report is based on our findings in 79 such patients.

Material and Method

All patients who attended the Dermatology out-patient department of the Kottayam Medical College Hospital with complaints of symmetrical pruritic follicular pustules of legs, between 10th July 1969 and 27th August 1970 were included in this study. They were interrogated on the basis of a proforma regarding duration and mode of onset of the illness, its course, nature and response to previous therapy, personal habits, details of occupation, aggravating factors and family history of

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hereditary or metabolic diseases. Since the dominant clinical picture varied from case to case each patient was graded by more than one observer as follows:

Grade 1. Patients having only follicular pustules, few of them showed well defined perifollicular erythema.

Grade II: Predominant lesions were follicular pustles but there were also significant number of infiltrated papules around broken hairs. The papules showed excoriation marks, crusting or a peripheral rim of white scales. Alopecia was observable but not marked. Palpating hand cou d detect the 'wiry roughness' due to the broken or irregular hair.

Grade III: This stage was characterized by number of infiltrated scaly papules though few scattered pustules were also seen at the periphery. There was atrophy and shininess and marked alopecia (Fig. 1)

Grade IV: This was characterized by almost complete alopecia and atrophic shiny and scaly skin. Pustules were no more seen but few discrete scaly papules were seen at the periphery. (Fig.2)

Investigations included total and differential leucocyte count, haemoglobin (Sahli's), erythrocyte sedimentation rate (Westergrenns), STS, blood grouping, serum protiens including paper electrophoresis, stools for ova and cyst and complete urinalysis. Pus from the lesions were directly innoculated into blood agar plates and antibiotic sensitivity tests done. Biopsy was done from representative lesions. Fifteen patients were admitted to observe the results of selected therapeutic regimes. Others were treated as out-patients and reviewed fortnightly and progress recorded.

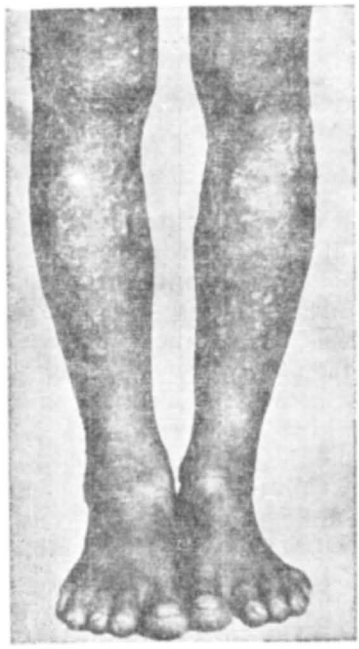

Fig. 1

Shows typical grade 3 stage of Nigerian Shin disease.

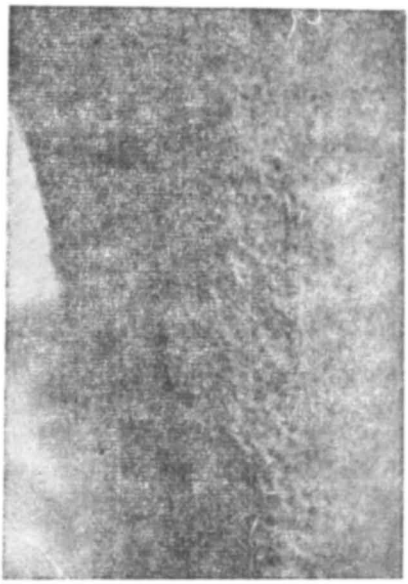

Fig. 2

Shows grade 4. Note pitted scarring.

Observations

The period of study was one year and 17 days. Out of 79 patients there were 72 males and 7 females. The youngest patient was aged 17 and the oldest was 58 years old. Though the majority of patients belonged to the 16–31 age (Fig 3 and 4) group the arithmetic mean age was 30.8 years (Standard deviation 9.8 years). These patients represented people from all walks of life.

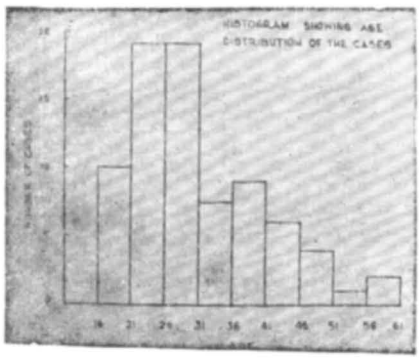

Fig. 3

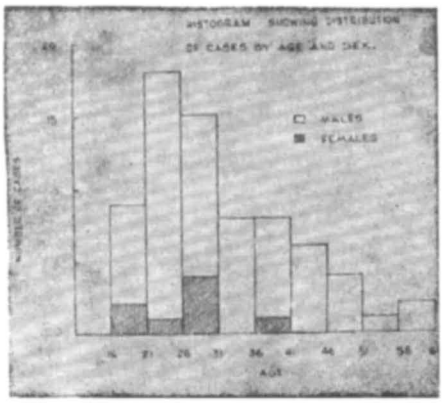

Fig. 4

Shows the age and sex distribution of cases.

The duration of the illness varied from 2 months to 22 years. More than 5 years duration was reported by 28 patients. Simultaneous onset in both legs were reported by 19, whereas 60 patients had an unilateral onset. Among the latter the time taken for the involvement of the other leg varied from 1 month to 8 years (Median average 0.9 years). Six patients were found to have grade I, 16 grade II, 48 grade III and 9 grade IV of the illness. However the extent of involvement had no relationship to the grade or duration of the illness. But there was a significant relation between the grade of the illness and the duration (P 0.05). The nature of onset also did not decide the future course.

Among the 79 patients only 21% (17) had the lesions confined to the "classical sites". Fifty two percent (41) had involvement of the front of the thighs in addition to the legs. Involvement of the thighs and forearms were seen in 22.8% (18), and facial involvement in addition to the above was seen in 3.8% (3). All patients who showed involvement of the thighs had bilaterally symmetrical involvement of the legs. The lesions were strictly confined to the hairy areas but the pubic region, the trunk and the scalp were never affected. The borders were not always well defined. All the female patients except one had well developed terminal hairs. None among them showed any lesions above the knees.

Pitting oedema and scratch marks were always seen in grade II and III of the illness. 'Wiry roughness' of the hairs could easily be detected in all patients who complained of severe itching. Scarring and atrophy showed certain preferential localization. The shiny appearance was mostly seen on the anterior and anterolateral aspects of the lower two third of the legs. On the lateral aspect of the calves and on the anterior aspects of the thighs numerous, well-defined, small, depressed, roundish, atrophic scars; measuring 2–4mm in diameter were seen in the place of the hairs. Few unaffected downy hairs were seen in those areas on closer scrutiny. Post inflammatory depigmentation was observed only in 1 and another showed lichenification.

Subjective and objective aggravations were reported due to a variety of factors; ingestion of animal protein 30.3% (24), alcoholic drinks 24% (19), sleeplessness 30.3% (24), contact with wet soil 26.5% (21), excessive sweating, contact with cowdung and even sexual indulgence! 5% (4). Seasonal aggravation was reported by 66, of which 50.6% (40) were worse in summer, 20.2% (16) during winter and 12.6% (10) during monsoon. Among the 70 patients only 47 regularly applied coconut oil on their body. It did not influence the course of the disease in any way.

Blood group studies revealed that 55.6% belonged to 'O' group, 18.9% in group B, 14.3% in group A and 3.7% in AB. It could not be done in 7.5%. All the cultures yielded coagulase positive staphylococci. Antibiogram (disc method) showed that the most effective antibiotic was streptomycin. Kanamycin tetracyclines, and chloramphenicol were also effective in vitro.

None of the patients showed any systemic disorder usually associated with resistant pyogenic infections like Diabetes mellitus, Tuberculosis, Malnutrition or Malignancy. Anaemia was not a constant feature and leucocytosis above 10,000/- cmm was observed in only 51 patients. Serum paper electrophoresis was done in 51 patients. Significant hypergammaglobulinaemia was observed in 26 patients (Fig 5) STS (VDRL test) was negative in all.

Histopathological examination of the earliest lesion showed only nonspecific focal round cell infiltration of the dermis and focal infiltration of the mouth of the hair follicle by polymorphs. The well developed pustular lesion showed a "Wine glass" shaped abcess involving the upper part of the hair follicle without involvement of the bulb or root of the hair (Fig: 6). The hair shaft was seen passing through the abcess cavity. Flakes of keratin and several acantholytic cells were seen in the abcess cavity. There was also lymphocytic and plasma cell infiltration of the sweat glands and hair follicle. One remarkable observation was the uniform absence of the foreign body giant cells around the hair follicle described by Miller and others. No

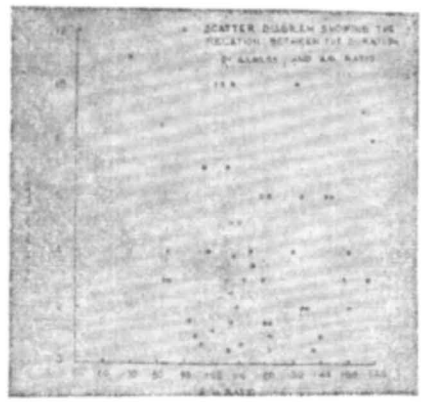

Fig. 5

Scattergram showing incidence of hypergammaglobulinaemia in relation to age and duratin of the disease.

Fig. 6

Shows typical "wine glass" shaped abscess involving the upper 3rd of hair follicle.

sebaceous glands were observed in any of the biopsy specimens. Another notable feature was the marginal acanthosis around the abcess. In the cicatricial stage (Fig: 7 & 8) the rete ridges were atrophic and dermis showed fibrosis with focal collection of round cells around the appendages. The arrector pilorum muscle was seen lying horizontally in the upper part of the skin.

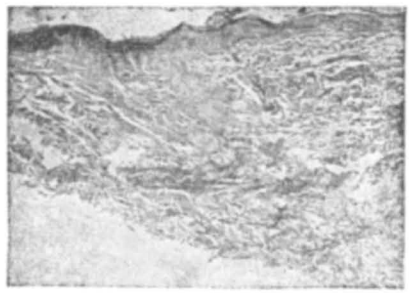

Fig. 7

Fig. 8

Shows early and late stage of scarring. Note the complete absence of hair follicles atrophy of epidermis and relatively horizontal position of arrectores pilorum muscle.

Treatment

Therapy resistance of this condition had been stressed by all the previous workers. After onset the disease progresses relentlessly inspite of all therapeutic efforts, till the hairs fall off and scarring set in. Our experiences were also not different. On the basis of the *in vitro* studies a number of antibiotics were used alone and in combination. Dyes, and antiseptics, and even the empirical medicines of pre antibiotic era were tried in desperation but the results were unsatisfactory.

Discussion

The disease called by various names such as epilating folliculitis of the glaborous skin, lupoid sycosis of legs, sycosis cruris, DCPA and Nigerian shin disease has a number of unique features which separate it from banal pustular folliculitis. The outstanding clinical features are its peculiar localization, symmetrical involvement, therapy resistance, extreme chronicity and inevitable alopecia and atrophy of the skin. It is predominantly a disease of men and women who had showed well developed terminal hairs. The presence of thick mature hairs thus seems to be a prerequisite. However even in those with extensive involvement, the scalp, trunk and the pubic region were never affected. Harman (1968) stressed the occurrence of this disease in patients below the age of 30, but in our experience 36 patients were above the age of 30 at the time of examination. Among these 36 patients 22 developed the disease only after the age of 30.

Desai et al (1964) reported a male predominance. Our findings are in agreement with this The most distressing symptom of our patients was itching and scratch marks could be readily detected in grade II and III. The wiry roughness is entirely due to scratching and has no other diagnostic significance. Pardo-Castello (1956), (1962) had implied injury with sugar cane needles as a possible cause and claimed it to be an occupational disease. This is rather unlikely. Our patients represent people from all walks of life. Further, those who had changed their profession after the onset of the disease continued to have it. The local application of coconut oil being a widely prevalent custom among the local population, it is unlikely to have any bearing on the initiation or perpetuation of this disease.

The complete absence of any observable effects even in the presence of

extensive lesions was surprising. There was no lymphadenopathy, or fever. The state of nutrition and standard of hygiene were adequate. The presence of hyper-gammaglobulinaemia had no correlation with the extent of involvement or duration of the illness. We also therefore agree with the suggestion of Desai et al (1964) that it reflects only the presence of a chronic infection. Thus the aetiology of this disease remains to be understood.

The existing names of this disease are misleading or inadequate. As it starts purely as a follicular pustule and dermatitic changes are secondary the term DCPA is misleading. Since it is prevalent even in countries other than Nigeria and the lesions are seen at other anatomical sites the term Nigerian Shin Disease is also inadequate. The term lupoid sycosis, sylosis cruris and epilating folliculitis are also unacceptable for obvious reasons. Since it starts as follicular pustules and end in atrophy and predominantly seen in the legs we propose the modified term 'Folliculitis Cruris Pustulosa et Atrophicans'. An alternate term might be Clarke's Disease.

Acknowledgments

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TRUE or FALSE?

Koplik's spots the diagnostic clinical lesions of Measels lack specific histological features by which they can be detected.

(Answer page No. 44)