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CONFLUENT AND RETICULATED PAPILLOMATOSIS (GOUGEROT-CARTEAUD)

An Abnormal Host Reaction to Malassezzia Furfur

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Abstract. Three cases which satisfied the clinical and histological criteria for the diagnosis of confluent and reticulate papillomatosis (C.R.P.) are reported. The three patients, all males, showed distinct yellow fluorescence in skin lesions under Wood's light and were colonized by *Pityrosporon orbiculare*. In one case, filament formation as in pityriasis versicolor was found in addition. Treatment with 2 1/2% selenium sulphide shampoo (Selsun, Abbott) completely cleared up one case; in one case therapy had to be stopped after 10 days due to intolerance to the topical application and in the third case vitamin A injections alone cleared the C.R.P. lesions. The authors discuss the possibility that C.R.P. may represent an abnormal host reaction to *Malassezzia furfur*.

In 1927 Gougerot & Carteaud presented a case for diagnosis under the name papillomatose pigmentée innominée in a healthy woman of 21 years in whom the lesions had started at the age of 5 years (3). As a rule, the initial lesion, which is a papillomatous papule, appears shortly after puberty and is greyish-brown in colour. Abrasion of the papule produces a mealy deposit. By coalescing, the papules form confluent and reticulated patches which fade imperceptibly into normal skin. The condition usually begins in the intermammary and epigastric regions and extends downwards over the pubic area and upwards over the breasts and axillae. Intergluteal and crural folds may be slightly involved. Concomitantly the skin of the neck becomes thick, rough and dark. In age of onset, pigmentary disturbance and distribution there is close similarity to pityriasis versicolor. Histologically there is hyperkeratosis, keratotic plugging, atrophy of the malpighian layer with elongation of the rete pegs with a slight, non-specific perivascular infiltrate of lymphocytes in the dermis (7).

Beurey et al. (1) reported a case of C.R.P. with associated pityriasis versicolor. Kesten & James (4) reported 5 cases collected over a period of 4 years, 3 of whom were believed at first sight to have pityriasis versicolor. In their very first case Gougerot & Carteaud (3) mentioned a close resemblance between C.R.P. and pityriasis versicolor. Marshall (5) has also recorded a similarity between the two conditions. Roberts & Lachapalle (8) were the first to associate C.R.P. with *P. orbiculare*.

REPORT OF CASES

Case 1

A 24-year-old Moslem male patient sought advice for lesions on his skin of 8 years' duration which were mildly pruritic but mainly of cosmetic embarrassment. The lesions consisted of dark brown papules coalescing to form a plaque on the interscapular region and reticulate pattern over the scapulae (Fig. 1). Similar lesions were also present on the anterior chest wall (Fig. 2). He had typical scaly hypopigmented macules of pityriasis versicolor over both upper arms. He had applied several ointments in the past including Whitfield's ointment which he used to discontinue within a few days because of skin irritation.

On scraping the C.R.P. lesions from different sites a mealy deposit was obtained which after potassium hydroxide (KOH) clearing showed a few clusters of spores but no filament formation. The hypopigmented patches on the upper limbs showed many hyphae but no spores. Under Wood's lamp all the lesions, both the C.R.P. as well as the hypopigmented macules, showed a distinct yellow fluorescence.

Histology showed marked hyperkeratosis, keratotic plugging, slight atrophy of stratum malpighii and elongation of the rete pegs with a perivascular lymphocytic infiltrate in the dermis. Periodic acid schiff stain (PAS) did not show any spores in the stratum corneum (Fig. 3). Treatment with 2 1/2% selenium sulphide shampoo was

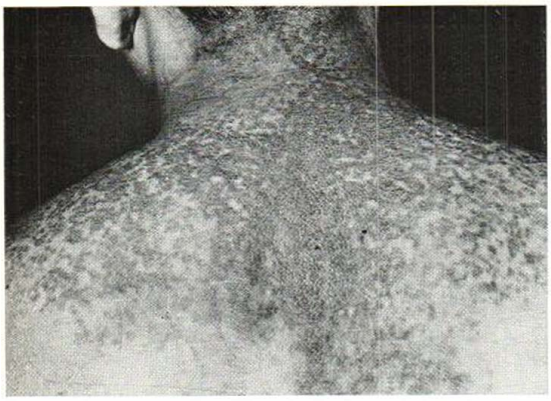

Fig. 1. Case 1. Interscapular area showing confluent papules with reticulated appearance towards the periphery.

commenced and within 10 days most of the C.R.P. lesions had started to clear, leaving the hypopigmented macules of pityriasis versicolor more clearly visible (Fig. 4). Unfortunately at this stage patient developed erythema and intense pruritus at the sites of application of selenium sulphide and was lost for follow-up for 4 months. At the end of this period the patient returned in the same condition as he was at his first attendance. However, he refused treatment for his skin condition, having come to the hospital for low backache.

Case 2

A 20-year-old Hindu male attended the Skin Out-patient Department for skin lesions of 9 months' duration. The lesions were not itchy and were only of cosmetic disability.

On examination he had brownish black papillomatous papules over the interscapular region, the right clavicle and face. The patient had evidence of avitaminosis in the form of Bitot's spots and angular stomatitis. Under

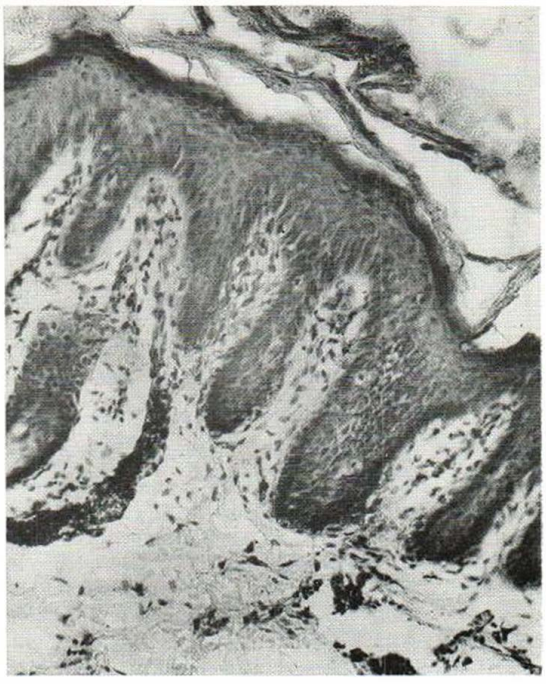

Fig. 3. Case 1. Hyperkeratosis with papillomatosis and slight perivascular dermal infiltrate. (PAS, × 125.)

Wood's lamp both the trunk and face lesions showed yellow fluorescence.

Scrapings from the back mounted on KOH showed abundant clusters of spores but no hyphae. The spores were much more numerous than in case 1.

Biopsy of the dorsal lesion was similar in appearance to case 1 and PAS stain revealed a few spores in the stratum corneum as well as in the keratotic plugs.

Treatment with selenium sulfide completely cleared up the lesions from the back and face within 2 weeks and the patient remained clear when reviewed 3 months later.

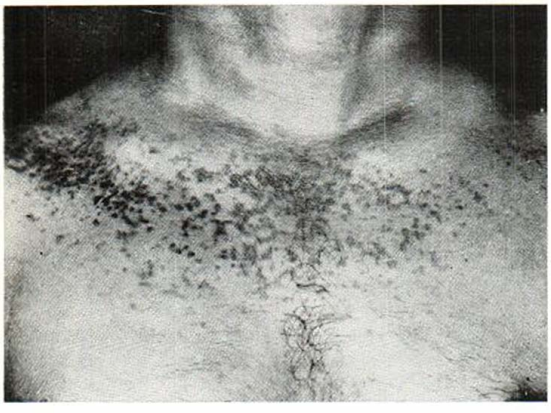

Fig. 2. Case 1. Presternal and infraclavicular papules in net-like pattern.

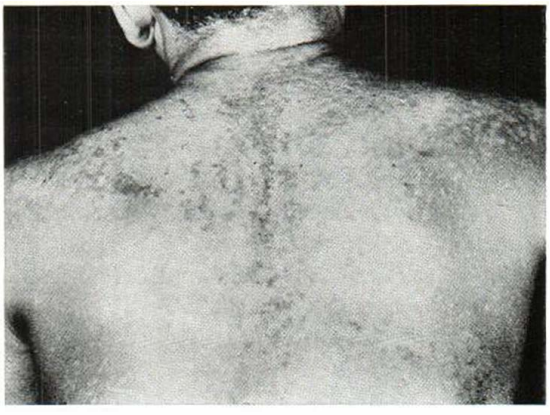

Fig. 4. Case 1. After 10 days' treatment with Selsun. Note hypopigmented macules over right shoulder.

Case 3

A 22-year-old Hindu male was admitted to the Skin Ward for an acute eczematous condition of his scrotal skin of 2 weeks' duration. On examination of the rest of his integument he was found to have classical hypopigmented scaly macules of pityriasis versicolor on his anterior chest wall with closely set brownish black papules over the manubrium sterni and coalescing to form confluent patches over the nape of the neck. When his attention was drawn to these lesions the patient stated that they had been present for only about 3 months. He had in addition pronounced avitaminosis as evidenced by angular stomatitis, glossitis, phrynoderma and Bitot's spots. Examination under Wood's lamp showed yellow fluorescence of the classical pityriasis versicolor lesions as well as the C.R.P. lesions.

Scrapings from the C.R.P. lesions on the neck and the hypopigmented macules on the chest were mounted separately on KOH. Both showed abundant spores and also numerous hyphae.

Biopsy from the neck lesion was similar in appearance to cases 1 and 2. PAS stain showed numerous hyphae and spores in the stratum corneum.

Treatment consisted of vitamin A injections, 100 000 units intramuscularly daily for 10 days. By the tenth day of treatment the C.R.P. lesions had vanished and only the classical pityriasis versicolor lesions remained. No external treatment was given.

Routine urine examination and blood sugar estimations were normal in all 3 cases described above. No family history of similar skin lesions was obtained in any of the 3 cases.

DISCUSSION

The exact aetiology of C.R.P. is not known. Some authors have attributed it to a genodermatosis but only in the report by Braden (2) was a familial incidence noted. Endocrine disturbance has also been postulated since many cases are associated with obesity. Wise (11) reported a case of C.R.P. with Cushing's disease. Marshall (5) mentions a Bantu boy with gynaecomasia and C.R.P. Watkins & Lockwood (10) reported a case in a 28-year-old Negro who was obese and who had diabetes mellitus. None of our 3 cases had diabetes mellitus or obesity.

The close clinical resemblance of C.R.P. to pityriasis versicolor has been alluded to by many authors but dismissed as insignificant because hyphal elements were not seen by the usual KOH technique. In fact, Beurey et al. (1) recorded a case of C.R.P. with pityriasis versicolor but since treatment with sodium thiosulphate did not clear up the C.R.P. lesion they regarded the association as fortuitous. Roberts & Lachapalle (8) were the first to consider the possibility of the association between C.R.P. and *P. orbiculare* as having an aetiological significance. Although they could not detect any hyphal elements, numerous clusters of *P. orbiculare* were found when scrapings were stained with the Parker ink/KOH technique and allowed to stand overnight. Since the whole eruption vanished after treatment initially with 10% sodium thiosulphate and later with 2.5% selenium sulfide shampoo they argued that C.R.P. was an abnormal host response to colonization of the skin by *P. orbiculare*.

From the study of our 3 cases we have reached a conclusion somewhat similar to Roberts & Lachapalle. The first case, which is the one of longest duration (8 years) showed spores of *P. orbiculare* but no hyphae. The spores were not numerous and a careful search had to be made before detecting them. PAS staining did not disclose spores in the sections studied, probably due to the sparse colonization by *P. orbiculare*. In case 2 of 9 months' duration no difficulty was experienced in detecting the spores in the scrapings. Spores were also seen easily in PAS sections. The third case of 3 months' duration not only showed spores in abundance but also numerous hyphae and PAS sections of the skin also had hyphae and spores in the stratum corneum.

The 3 cases thus appear to demonstrate the evolution of C.R.P. To start with, both filaments and spores are seen as in many ordinary pityriasis versicolor macules, but as the lesions develop, the hyphal elements seem to disappear or perhaps become transformed into *P. orbiculare* and still later the population of spores also diminishes as though the substratum of C.R.P. is no longer suitable for the existence of pityriasis orbiculare. The only inexplicable fact is that while pityriasis versicolor is a fairly common disease, C.R.P. is very rare; less than 50 cases have been recorded in world literature since Gougerot first described it. The only way to explain this discrepancy is to postulate, as Roberts et al. did, an abnormal host response. We feel, however, that this abnormal host reaction is not against the normal inhabitant pityriasis orbiculare but to the disease producing *Malassezzia furfur*. The cause of this abnormal host reaction remains obscure. Avitaminosis may play a role. Cases 2 and 3 showed evidence of avitaminosis. Case 3 was treated with vitamin A injections alone and this resulted in the disappearance of the C.R.P. lesions. In this context it is

interesting to note in the literature that the cases recorded by Miescher (6) and Waisman (9) also improved considerably with vitamin A therapy. Another unusual feature of our series of cases is that all the affected patients are males, whereas in literature more than two-thirds of the recorded cases have been females.

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