PROGRESSIVE PIGMENTARY DERMATOSIS OF SCHAMBERG

(Report of four cases)

By

K. C. KANDHARI, F.R.C.P. (Edin.), F.A.M.S.

and

V. N. SEHGAL, M.B., B.S., M.D.

Schamberg (1901) described a chronic dermatosis which begins as pinhead sized, reddish, points or dots forming irregular patches which slowly extend by the formation of new lesions about the periphery. The puncta in the course of time disappear, leaving behind a brownish yellow, or reddish brown pigmentation, which slowly fades. The disease involves the distant area of integument and is progressive in character. The spontaneous involution occurs in the oldest area, and is possible that complete restoration to the normal condition of the skin may take place. There is an entire absence of subjective symptoms. The course is unknown. The pathologic process has its chief seat in the subpapillary zone of the corium, seemingly in the immediate vicinity of coil gland ducts. LITTLE (1902) reported a similar case, and KINGERY (1918) a typical example. KINGERY reported histologic picture showing low grade inflammatory process, plus pigmentary deposits, the pigment being an iron derivative, and his finding has been corroborated by PAUTRIER (1927) in his contribution on exchange of substance between the dermis and epidermis. McCAFFERTY (1926) and DESAI (1937) have each added a case to the recorded list. GRECO et al (1938) believed it to be due to a budding fungus found within the red blood cells. WISE (1942) stated that annular formation and serpentine capillary lesions do not occur, that lichenoid and desquamative changes are secondary, that hypercholestraemia is found in many of the patients who nevertheless appear to be in good health and that there is no relationship between schamberg's disease and varicose veins. RANDALL, KIERLAND & MONTGOMERY (1951) reviewed the records of 72 patients who had schamberg's disease and stated, "the average age at the time of observation was forty eight years, with extremes of eleven and seventy six years. The longest duration was forty years, and the shortest one year. The disorder was almost 5 times commoner in males than in females. 83 persons had no symptoms and 17 percent complained of mild to moderate pruritus." According to LEVER (1961) it is believed that the basic pathological process underlying the disease is capillaritis of unknown origin, occuring in upper dermis and leading to capillary fragility.

As there are a few case reports of a rare though harmless disorder, from India, the authors feel it worthy of reporting four cases of progressive pigmentary dermatosis of schamberg which have been confirmed by clinical and histological examinations.

From Department of Dermato-venereology, All India Institute of Medical Sciences, New Delhi-16 (India).

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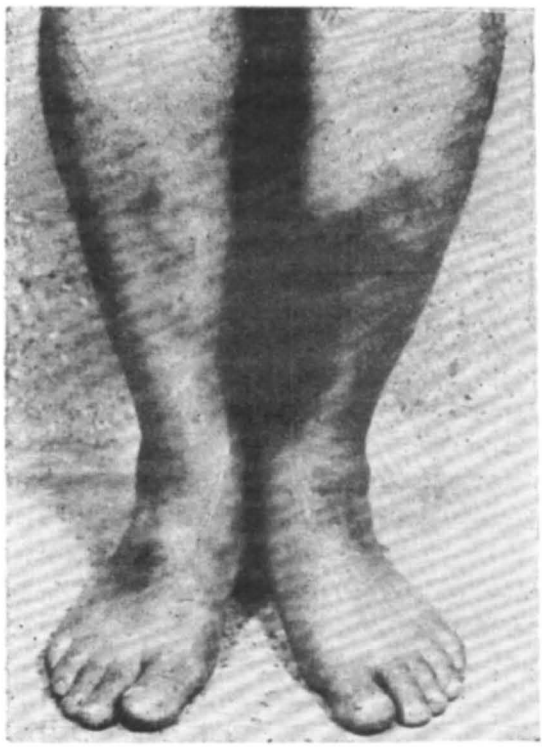

Fig. 1
Schambergs'

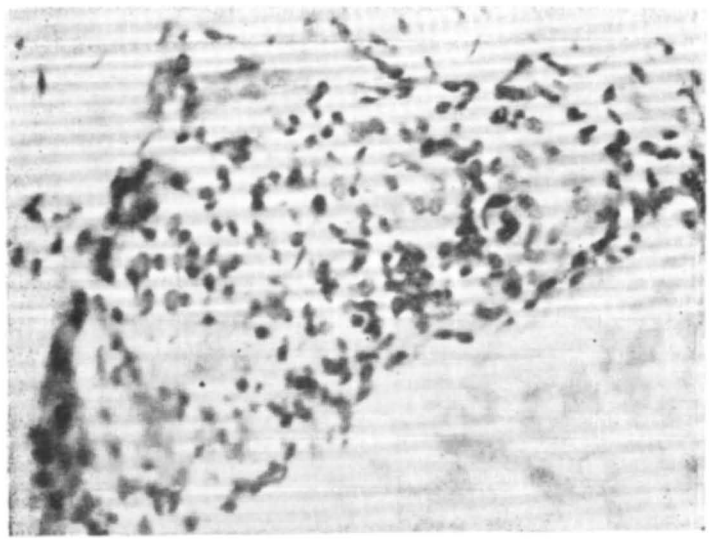

Fig. (1st patient) 2
Subpapillary zone of dermis shows thickening and swelling of the capillary wall with perivascular infiltrate of lymphocytes. (x400)

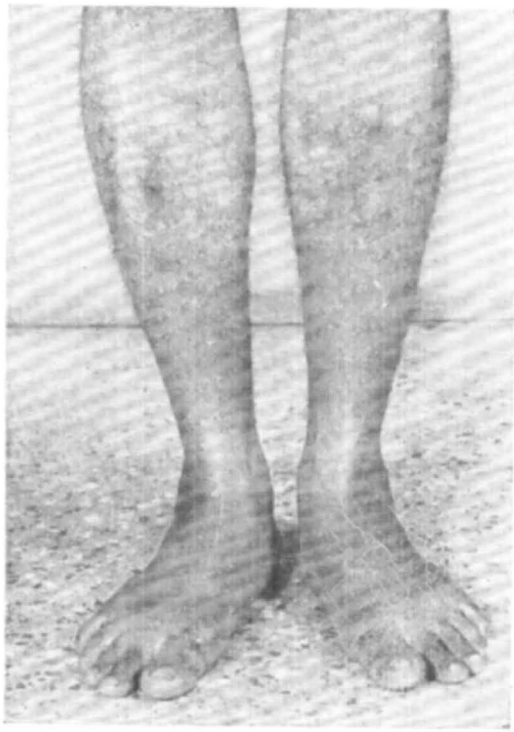

Fig. 3a
Schambergs'

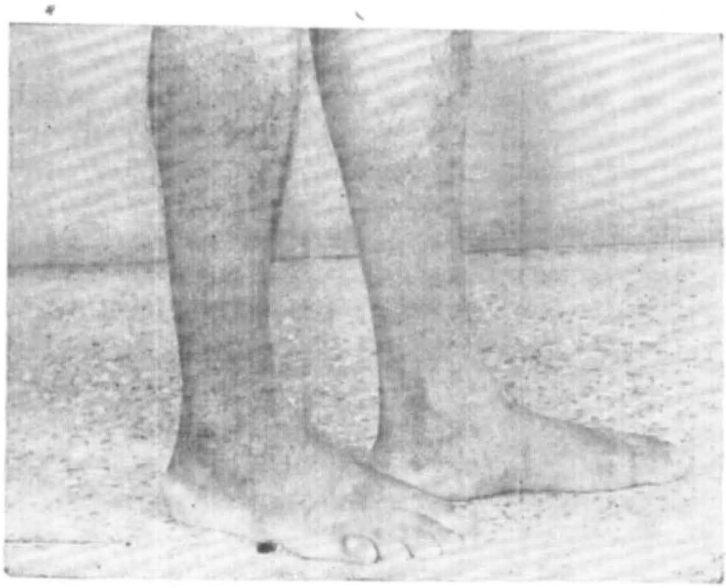

Fig. 3b. Schambergs'

CASE REPORTS

Case No. 1. A patient 35 years old male reported to the sk n out patient department of AIIMS in September, 1962 with reddish eruptions on both the lower legs, the total duration of which was about 6 months. These eruptions were absolutely a-symptomatic but progressive in nature. Nothing contributory was otherwise available in the history.

LOCAL EXAMINATION Fig (1) revealed erythematous macular lesions with well defined margins, some of these showing tendency to coalesce. The associated pigmented macules were also observed. The lesions were distri! uted on the legs bilaterally. No varicose veins were seen. No abnormality was detected on general physical examination. Results of hematologic studies including bleeding time, coagulation time, and thrombocytes were within normal limits. Capillary fragility test was also normal. HISTOPATHOLOGY Tissue sections stained with haematoxylin and eosin showed moderate hyper- keratosis with irregular flattening of reteridges. The subpapillary zone of the dermis showed thickening and swelling of the capillary wall with perivascular infiltration composing chiefly of lymphocytes. Here and there extravastion of red blood cells was also seen. No pigment was seen in the dermis. Fig (2). These findings were compatible with schamberg's disease.

Case No. 2. 39 years' old male reported in September, 1963 with reddish pigmented asymptomatic spots on the legs, which started about 2½ months prior to attending skin out patient department at AIIMS. Ever since these eruptions continued progressing. No other relavent point was available in the history. LOCAL EXAMINATION Fig (3) showed multiple erythematus, pigmented punctate lesions of varying sizes, some of which merging together to give a gyrate appearance, distributed on the extensor aspect of the legs encroaching upon the dorsa of feet bilaterally. No varicose veins were seen. The routine blood studies including bleeding time, clotting time and capillary fragility tests were within normal limits. HISTOPATHOLOGY Tissue section stained with haematoxylin and eosin showed mild hyperkeratosis with flattening of reteridges at places. The capillaries in the subpapillary zone of the dermis showed swelling and proliferation of endothelial cells with perivascular infiltrates, comprising lymphocytes as a chief constituent. There was no extravasation of red blood cells or deposition of pigment. Fig (4). These findings were suggestive of schamberg's disease.

Case No. 3. 15 years old male patient was admitted to the skin ward of AIIMS in October, 1963 with progressive asymptomatic punctate erythematus lesions on both the legs, started about 2 months prior to his admission. A few days previous to these lesions, he developed swelling of the ankle joints, which subsided with local and systemic treatment. On interrogation, he pointed out regarding the recurrent attacks of upper respiratory infection. During his stay in the hospital we noticed stray lesions of similar nature on the upper extremities also. LOCAL EXAMINATION revealed erythematus, pigmented puncta of different

sizes with well defined margins distributed on the lower legs bilaterally. No abnormality was found on general physical examination. With the above findings diagnosis of Schamberg's disease and erythema multiforme was given. The routine hematological studies including bleeding time clotting time, thromocytes counts were within normal limits and so also was the capillary fragility test. HISTOPATHOLOGY—Tissue section stained with haematoxylin and eosin showed minimal changes in the epidermis. The capillaries in the dermis were dilated and increased in number. There was swelling, degeneration and proliferation of the endothelial cells. Here and there, there was extravasation of erythrocyts. No pigment deposits were seen in the dermis. Fig (5). Findings were diagnostic of Schamberg's disease.

Case No. 4 18 years' female reported in September, 1964 to the Skin outpatient department of the AIIMS with the complaint of progressive pigmented reddish eruptions on the legs. The total duration of which was 6 months. These eruptions were unassociated with any symptoms. LOCAL EXAMINATION—Fig (6) revealed pigmented erythematus macular eruptions with well defined margins distributed on the extensor surfaces of the lower legs with a few lesions on the dorsa of the feet bilaterally. No varicose veins were seen. No abnormality was found on general physical examination. The routine blood investigations were normal. So also the bleeding, the clotting time, platelet count and capillary fragility test. HISTOPATHOLOGY—Tissue section stained with haematoxylin and eosin showed that the epidermis was normal, whereas the upper dermis showed changes in the blood capillaries i. e. swelling of the wall with perivascular infiltrate of lymphocytes. No extravasation of erythrocytes or pigment deposits were seen Fig (7). These findings were compatible with Schamberg's disease. PROGRESS REPORT—All the above cases described are being followed and none of the cases so far has shown any sign of regression of the lesions, rather they are progressively increasing and many new lesions are making their appearance. The pigmentation has started increasing in case No. 1,2 and 4 respectively. No symptoms are being compliained of by any of the patients.

COMMENTS—Progressive pigmentary dermatosis of Schamberg is a rare though harmless disease of idiopathic origin. Four cases of disorder are presented with full clinical details and histopathological findings. The clinical features as is evident are quite characteristic and are in conformity with those described by Schamberg (1901) Little (1902) Kingery (1918) McCafferty (1926) Desai (1937) and Randall et al (1951). In none of these cases varicose veins were found as an associated finding, this was also emphasised by Wise (1942) who stated that there was no relationship between Schamberg's disease and varicose veins. However, it has recently been pointed out by Marshall (1960) that the histological picture is that of capillaritis as has also been seen in progressive pigmentary dermatosis of Schamberg. Stasis and anoxaemia could hardly produce such a picture. It is probable that microbic allergens are also implicated. Such an explanation may also be of some value in understanding the origin of a disease under discussion. It may be presumed that they are

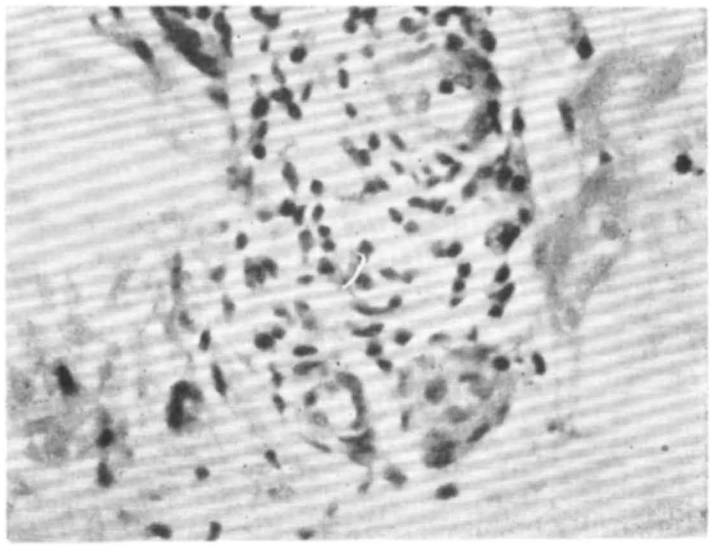

Fig. (2nd patient) 4

The capillaries in the sub-papillary zone of the dermis show swelling and proliferation of endothelial cells with perivascular infiltrates of lymphocytes (x 400)

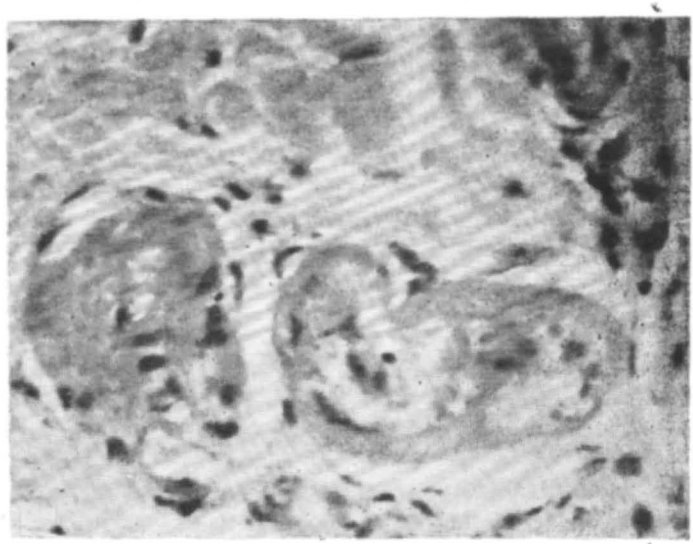

Fig. (3rd patient) 5

The capillaries in the dermis are dilated with, swelling proliferation and degeneration of the endothelial cells. Infilterate composed of lymphocytes. (x 400)

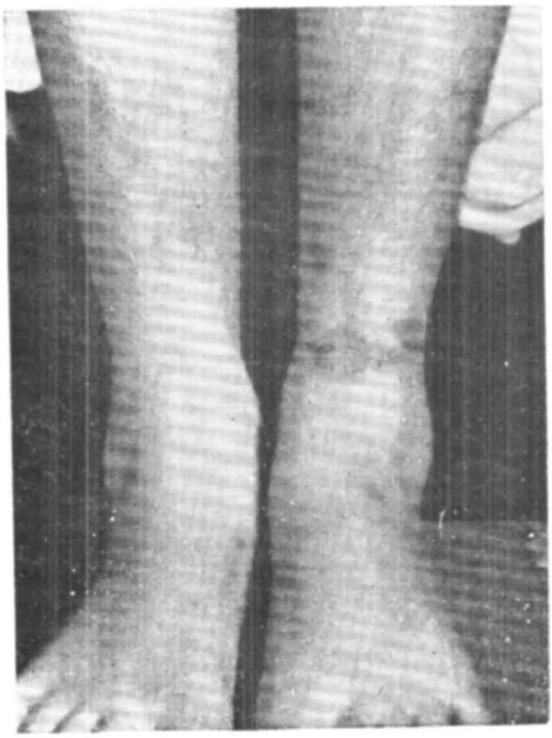

Fig. 6. Schambergs

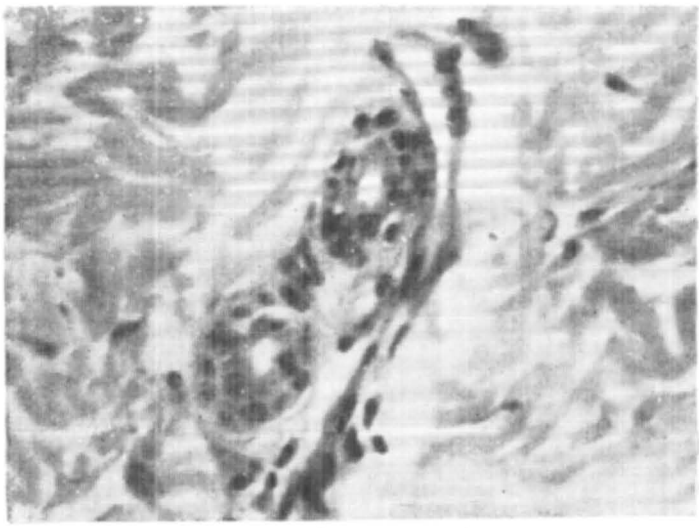

Fig. (4th patient) 7

The capillaries show swellinu and thickaning of wall with perivascular infiltrate of lymphyocytes. (x 400)

allergides and the microbes involved are not always the same. French workers on the other hand believed it to be due to tuberculosis as quoted by Marshall (1960) though it is a question yet open to criticism. The association of diverticulitis has also been observed by Marshall (1960) though it is a point difficult to conceive. Lever (1961) brought out that the basic pathological process underlying this disease is capillaritis which in turn leads on to capillary fragility, thus explaining to some extent the clinical picture. The precise nature of this is still unknown. No symptoms were complained of by any of the patients which again is a point in support of Randall et al (1951) who reported absence of symptoms in 83 percent of cases and rest of them had mild pruritis. He also pointed out the preponderance of the adult males over the female which is also our finding. No evidence of iron pigment deposit were found in the dermis in our cases, probably the reason being that these were fairly early cases and such a change occurs in the later stages of the disease. Spontaneous remission of this conditions is reported by Schamberg (1901) Ormsby & Montgomery (1954) Marshall (1960). It is too premature to advance any comment as regard the course of this disease as we are still following these cases.

SUMMARY

1—Four cases of progressive pigmentary dermatosis of Sceamberg are presented with clinical details with a light on salient histopathological features.

2—Literature on the subject has been briefly reviewed.

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