## Clinical Section

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[February 14, 1936, continued]

Idiopathic Steatorrhæa with Skin Lesions and Megalocytic Anæmia.—Geoffrey Konstam, M.D. and Hugh Gordon, M.D.

Clifford G., aged 55, clerk. History of pigmented scaling skin lesions over trunk and limbs since 1933. In April 1935 he began to pass large pale diarrhœic stools two or three times a day, and concurrently lost weight and became sallow. May 1935: Operation for strangulated right inguinal hernia. When aged 19 attended an orthopædic clinic on account of weakness of knees; at age of 14 was treated at a chest hospital for cough. Has never lived in the tropics.

Admitted to the West London Hospital, 16.10.35.

Condition on admission.—Thin man; normal skeletal development except for slight genu valgum. Papillæ of tongue flattened. No abnormal physical signs in circulatory, respiratory, or central nervous systems. Spleen and liver not palpable.

Complexion sallow with malar flush and dusky red patch on tip of nose.

Skin.—Generally rather dry with tendency to fine scaling on upper arms. Patches of dermatitis are scattered over the body, in a roughly symmetrical distribution. On the extensor surface of each upper arm there is a brightish-red patch of roughly circular shape, about 5 cm. in diameter: its surface is dry, rather shiny, with dried-up vesicles; pigmentation marked at the border.

On the chest are five or six patches of pigmentation, varying from 1 cm. to 3 cm. in diameter. There have been irritable red patches of dermatitis, but these have now subsided. In the right groin there is the scar of a herniotomy (performed in May 1935) which is deeply pigmented along its whole course. Left lower leg: Patch of dermatitis about 10 cm. in diameter; pigmentation marked, particularly at periphery. The skin of the whole left lower leg is generally superficially cracked in fine transverse striations, rather suggesting the appearance of a sulphur dermatitis (see coloured plate). Dorsum of feet: Small circular patches, completely pigmented, with some degree of lichenification. Extending from the anus on to both buttocks is an elongated area, about 5 cm. in width, of typical lichenification. Scattered elsewhere on the body are small patches similar in appearance to those described.

The picture is, therefore, one of scattered patches of pruriginous vesicular

dermatitis of a chronic nature, with a marked tendency to pigmentation.

The general dryness of the skin has been noted by the patient for only eighteen months and the irritation for the same period. The irritation is not very severe, though at times it has worried the patient a good deal; as the appearance suggests, the peri-anal region has been the most troublesome.

Slit-lamp examination of eyes (Mr. R. Davenport): Radiating post-cortical

opacities, left greater than right. Adult nucleus well marked.

Blood-count (25.7.35): R.B.C. 3,540,000; Hb. 75%; C.I. 1.05; W.B.C. 4,400. Differential: Polys. 69%; lymphos. 23%; eosinos. 1%; monos. 7%. R.B.C.'s regularly macrocytic and hyperchromic. Reticulocytes scanty.

Table of subsequent blood-counts (Dr. R. Elworthy):—

Date	R.B.C. in millions	Hb.	<b>C</b> . I.	W.B.C.	Reticulo- cytes	Remarks	
15.8.35 30.8.35	3⋅50 2⋅80	80 70	$1 \cdot 14 \\ 1 \cdot 25$	4.200	0·2% 0·3%	Hog stomach started 8.8.35	
19.9.35 $21.10.35$	$3.79 \\ 3.72$	82 80	1.05 1.08	4,500 3,600		Marmite 3 i	wtde
28.10.35 5.11.35	3·71 4·15	82 84	$1.11 \\ 1.02$	<b>ə</b> ,400	0.25%	mailine Ji	v 0.a.s.
15.11.35	3.89	80	1.02	4,200 4,500	Scanty	Intramusc. 10 c.c.	Liver alt. die.,
21.11.35	$3 \cdot 44$	74	1.07	3,600		10 0.0.	
3.12.35	3.46	74	1.07	4,500	_		
30.12.35 7.1.36	3∙ <b>73</b> 3∙95	78 84	1·03 1·06	5,200 4,700	<del></del>		

Corpuscular volume index: 1.09—1.2; serum calcium 8.4 mgm. per 100 c.c. plasma inorganic phosphorus 2.3 mgm. per 100 c.c.

Van den Bergh reaction (18.11.35) direct negative; indirect, weak positive. (Bilirubin less than 0.5 units.)

Blood-sugar curve: Fasting, 0.07%; half an hour after 50 grm. of glucose, 0.07%; one hour after, 0.08%; one hour and a half after, 0.08%; two hours after, 0.09%. Blood-urea 0.034%.

Stools.—Bacteriological examination: No tubercle bacilli found. Fat-analysis: Total fat 69.37% by weight of dry stool; neutral fat 12.04%; free fatty acids 32.94% fatty acids as soaps 14.39%. Nitrogen content over a three days' period is equal to 9.15 grm. (3.05 grm. per twenty-four hours.)

Fractional test meal: Hydrochloric acid and total acidity, normal.

X-ray and barium enema examination: Dilated transverse, descending, and sigmoid colon; loop in descending colon. Barium meal: Stomach and duodenum, no evidence of organic lesion.

Control skiagram of tibia and fibula: Slight osteoporosis.

Basal metabolic rate +2%.

The general condition of the patient has improved considerably since a fat-free diet and extra calcium (calcium gluconate 3 ii t.d.s. and calciferol mgm. ii b.d. for three days every week) have been given. As is seen from the table of blood-counts, there has been practically no response to hæmatinics, including large amounts of intramuscular liver, hog's stomach, and marmite, per os.

The dermatological picture is not particularly uncommon; it is of considerable interest and importance in this case, however, for it appears that these findings are not uncommon in idiopathic steatorrhæa. Bennet, Hunter, and Vaughan [1] mention "small, abraded, erythematous areas occurring on the dorsal surfaces of the limbs" as being relatively common in the condition. In one case, which they describe in detail, the skin condition was the presenting symptom, and consisted of

"large, moist, red, abraded areas of 2.8 cm. diameter, on all limbs and to a lesser extent on the trunk. Many of these lesions showed scaly, brown, pigmented borders, and on the calf had a deep brown periphery, and moist red abraded centre, the whole appearance being not unlike that of pellagra."

A case described by Constam and Partch [2], with similar skin lesions associated with tetany and osteomalacia, was in fact diagnosed as pellagra, though it is almost certain that it was, in fact, idiopathic steatorrhæa. The term "pellagralike lesion," which appears to have been adopted in the literature, is open to criticism. The skin lesions in pellagra are practically confined to the exposed parts, since an undue sensitivity to sunlight is an important factor; they present a bright red, sharply marginated patch, often with a raised border, looking like an acute sunburn, which desquamates and may be followed by pigmentation and slight atrophy. There is no irritation.



KONSTAM and GORDON: Pigmented Skin Lesions in Idiopathic Steatorrhæa.

The lesions in idiopathic steatorrhea appear to favour the covered parts; they are pruriginous, and look and behave like a nondescript chronic eczema with lichenification. Excessive pigmentation is a striking feature, particularly the tendency to form a sharp ring of pigmentation round the periphery of the patch of dermatitis.

In this particular case, the rapidity with which pigment has been deposited in an operation scar suggests that trauma is the immediate cause of the pigment—surgical trauma in the case of the scar, and prolonged scratching in the case of the patches of dermatitis.

As suggested by Bennet, Hunter, and Vaughan [1], it seems probable that the skin condition is caused by the faulty intestinal absorption, resulting in a condition of avitaminosis. The skin lesions in their case, which closely resembles this one, cleared up rapidly with large doses of marmite.

In this patient improvement has been slower, but the decrease in pigmentation has been striking.

The absence of history of alimentary trouble in childhood suggests that the steatorrhea has been acquired in adult life.

## REFERENCES

BENNET, I., HUNTER, D., and VAUGHAN, J. Quart. Journ. Med., 1932, n.s., 1, 603.
 CONSTAM, G. R., and PARTCH, W. T. Minn. Med., 1929, 12, 40.

Dr. Parkes Weber said he thought that the skin lesions in this case could be classified as a kind of pruriginous dermatitis which was not specific for idiopathic steatorrhea, but might occur in association with various nutritional disorders in adults. The pruritus was an essential feature and led to frequent rubbing and lichenification, for instance, on the forearms. Between the buttocks it might give rise to an intertrigo-like lesion.

## Idiopathic Steatorrhœa with Osteoporosis, Tetany and Megalocytic Anæmia.—Geoffrey Konstam, M.D.

Emily M., aged 55. Pot-belly was first noticed at the age of 10 and since then she has been in and out of various hospitals with attacks of vomiting and ill-defined abdominal pain. Constipation has been the rule and only rarely have there been diarrheeic periods. From the age of 25 to that of 44 she suffered from "fainting attacks" and for the last ten years she has become increasingly deaf.

One year before coming under observation (13.11.34) she had complained of painful spasms of the hands, pains in the legs and loss of  $1\frac{1}{2}$  stones in weight. Diarrhea had been present for two months.

Condition on examination (November 13, 1934).—Small thin pallid woman. Weight 6 stone 7 lb. Chvostek's sign present. Upper limbs rather spastic; tetany of hands observed. Considerable tenderness over long bones but no signs of past rickets. Abdomen distended. Stools yellowish-grey and bulky. Systolic murmur over base of heart.

Investigations (November 1934).—Blood-count: R.B.C. 3,980,000; Hb. 74%; C.I. 0.9. Tendency to megalocytosis, anisocytosis and poikilocytosis. W.B.C. 4,900. Differential: Polys. 58%; lymphos. 41%; large hyalines 1%.

Fat analysis of stool: Total fat 54.8% (by weight of dried stool); neutral fat 8.2%; free fatty acids 29.1%; fatty acids as soaps, 17.5% (by weight of dried stool).

Barium meal X-ray examination: Enlargement of colon with much gaseous distension.

Control skiagrams of tibia, humerus, and femur showed osteoporosis with thinning of the cortex (see figs. 1 and 2).

Sugar tolerance curve: Fasting level, 0.09%; one hour after 50 grm. glucose, 0.10%; two hours after, 0.114%.