Erythema elevatum diutinum / by H. Radcliffe Crocker and Campbell Williams.

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Crocker

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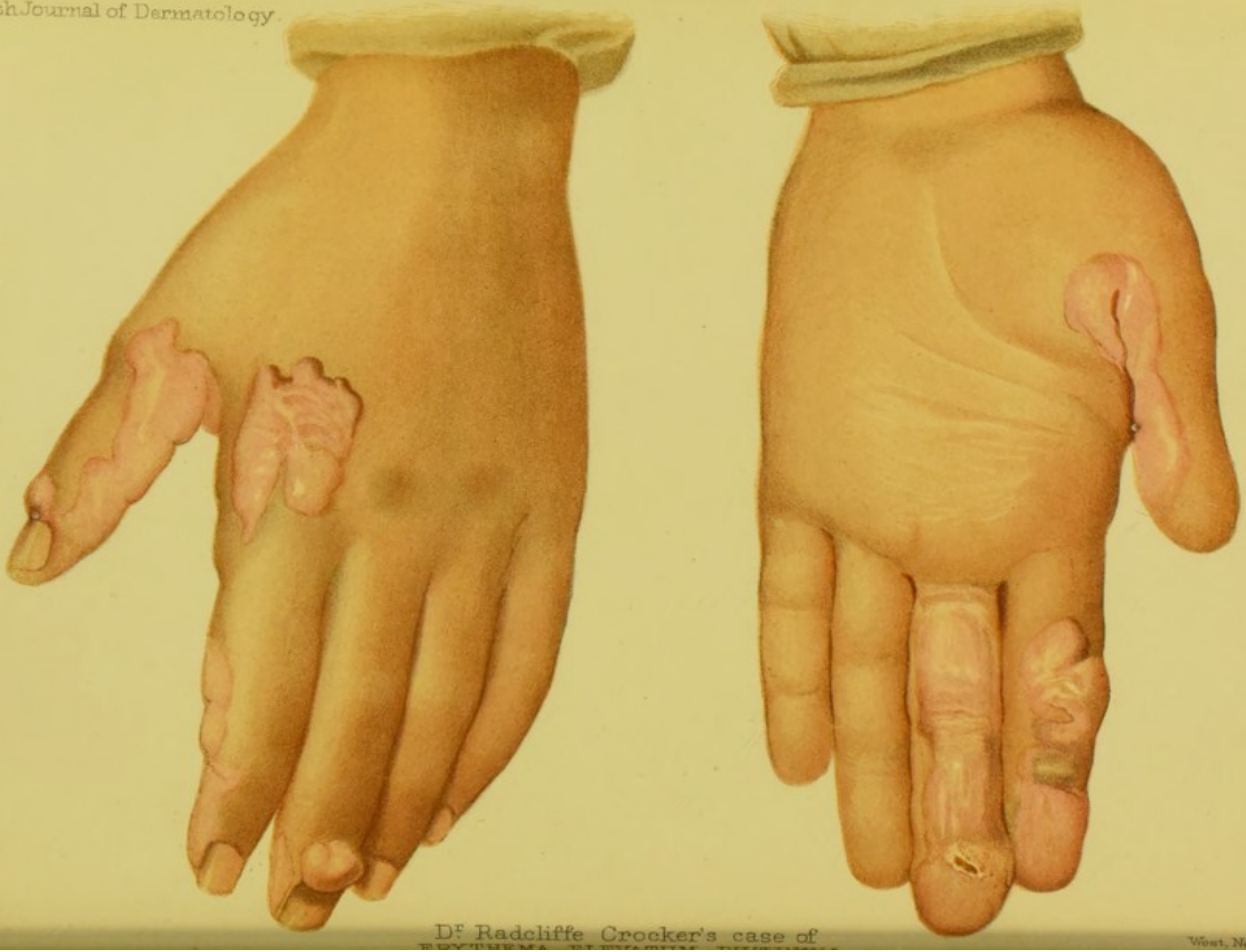

Dr Radcliffe Crocker's case of

West, Newman chromo.

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ERYTHEMA ELEVATUM DIUTINUM.

By H. RADCLIFFE CROCKER, M.D., F.R.C.P.,

AND

CAMPBELL WILLIAMS, F.R.C.S.

The advantage of a pictorial record of rare cases in producing the recognition of similar instances when they are met with by other observers is universally acknowledged. It is in the hope, therefore, of eliciting fresh facts that the following case is described and figured, and comparison made between it and other recorded cases more or less resembling it.

Amy H——, æt. 6 years, was brought first to the Victoria Hospital for Sick Children in October, 1893, and on the 21st was sent on from there for diagnosis, to University College Hospital. There was a very strong history of gout and rheumatism on the mother's side:—the mother's grandmother suffered badly from rheumatism; the mother's father and brother had gout badly, and her sister had rheumatism. The father's family history was unknown, but he himself was in good health.

The patient's general health had always been quite good, and she had never had any serious illness except measles in infancy.

The disease of the skin began simultaneously on both knees five months before she came under notice; next, it developed on the buttocks, elbows, and finally on the hands, which have been affected for two months.

Positions.—The lesions are situated on the left hand:—on the first

knuckle, and on the first terminal phalanx at the border of the nail of the thumb, first and second fingers, on the palmar surface of the same fingers, over the whole of the second finger, and over all but the proximal phalanx of the index finger; on the thumb, it affects only the bend and the proximal phalanx.

On the right hand:—the same fingers but not the thumb are affected, all up the palmar surface of the index finger, and the distal end of the second finger; the back is only slightly affected on the terminal phalanx of the index finger and the joint between the proximal and second phalanx of the first and second fingers.

The most highly developed lesions are over the first knuckle of the left hand and the inter-phalangeal joints of the right hand.

The one on the right index finger may be taken as a type; here the lesion was raised one-eighth of an inch, convex, sharply defined, pale purplish-red in colour, with a few dilated vessels over it, somewhat whiter on pressure, but diminishing very little in size; firm to the touch, very tender; but not painful except when pressed upon. It did not itch or burn.

Over the elbow-joints there were lesions evidently of a similar character, but retroceding. At the border the skin was wrinkled, and there was a brownish discoloration.

On the knees involution was still more complete; the left was almost well, except some slight discolorations and thickening in one part.

On the right there were still three prominent lesions; the rest had involuted, leaving a dull, slightly reddened tint.

The buttocks were quite well, only a faint purplish stain remaining, and on the right side lower down, where the mother said there had been several tumours, even the stain had faded.

While the lesions on the back of the hands were conspicuously nodular, those on the palmar surface were more diffused from coalescence of the component nodules, which were only just discernible, and the whole infiltration was less raised and defined. The affected parts were hotter to the touch, very slightly scaly here and there, but most of the nodules were quite smooth.

The child was not subject to chilblains nor to factitious or other variety of urticaria, but was a well-nourished, apparently healthy girl. She was treated at the Victoria Hospital for Children with liquor

arsenicalis and tinctura lupuli. The arsenic was at first given in two-minim doses three times a day, gradually increased to four minims, and locally a lotion of liquor carbonis detergens, ʒ j. to ℥ j. of water, was sponged on twice daily.

Under this treatment there was improvement, commencing on the elbows and knees, and later on the hands. On December 1st chloroform was given and a piece of skin removed from the left border of the patch over the first knuckle of the left hand, together with a little healthy skin. The wound was sewn up with horsehair sutures and dressed with wet mercuro-zinc cyanide gauze; then a pad of salicylic wool was put on, and the whole hand, except the thumb, enveloped in carbolic gauze.

On December 15th the dressing was taken off, the stitches removed, and the wound was healed. Marked improvement was observed in all the lesions of the left hand, but not so much on the right hand. On December 18th she came again by request to University College Hospital, when the following condition was noted:—All the patches were shrunk and flattened. The diseased area on the left forefinger was scarcely raised. The second finger thickening was diminished, but in a less degree. The nodule at the root of the nail of the second finger was greatly diminished. The thumb also was much improved, on the flexor surface it was only just perceptible to sight, and scarcely at all to the touch. The nodules on the back of the right knuckles looked shrivelled, but still projected, though to a less degree. The right forefinger also presented much less infiltration on the flexor side; on the right elbow there was still one bean-sized nodule left, and a portion of another above it, and the skin for a good half-inch round had a deep brownish stain. On the left elbow, there was nothing but staining and the remains of a small nodule on the left border, which had been knocked, and was consequently covered with a small scab. The left knee was well with the exception of staining. The right one still presented some signs of nodules, but for the most part only the purplish-brown stain was left.

The question is whether the operative or the general treatment produced the improvement, or whether it was not really spontaneous. The fact that improvement had set in before the operation suggests that it was the treatment adopted before, that was beneficial, viz., the arsenic and liquor carbonis detergens lotion.

The portion of skin removed was hardened in equal parts of alcohol and a $\frac{1}{6}$ per cent. solution of chromic acid and subsequently in pure alcohol. The result of the microscopical examination will be given in the concluding portion of the paper.

The case clearly did not fit into any known category, and our patient was therefore shown at the Dermatological Society on November 11th to elicit opinions, but no one had met with any case corresponding to it. Subsequently Mr. Hutchinson suggested that it resembled a case published with coloured plate by Dr. Judson Bury in the *Illustrated Medical News* of February 23rd, 1889, and republished in Mr. Hutchinson's *Archives of Surgery*, Vol. II. No. 8, plate lxi. Although there are some differences, careful comparison shows that the resemblances outweigh them, and that the two cases are clearly of the same nature.

For purposes of comparison I subjoin a very full abstract of Dr. Judson Bury's case, a girl æt. 12 years, who came under his care first in July, 1888. She was the youngest of twelve children; all the other children and the parents were quite healthy, and there was no history in the whole family of rheumatism, gout or any skin disease. The patient herself had measles when five years old, scarlet fever when nine years old, followed by rheumatic fever. Her general health was good; she was well developed, and all the viscera were healthy. She had never suffered from chilblains. The eruption began early in July, 1888, in the toes, then attacked the knees, then the elbows, and lastly the hands. She had constant hyperidrosis of the palms, the sweat having an acid reaction. The hands were swollen, and there was much thickening of the superficial tissues on the palmar aspect, where the skin had a peculiar purple tint. The purplish thickenings had well-defined raised edges, especially marked along the radial side of the thumb, and even at this time the thickened edge was nodular, but this feature became more marked at a later stage. There were also raised patches of erythema on the backs of the elbows, and to a less extent over the knees. The toes were slightly livid and swollen; there was a small erythematous spot at the right corner of the mouth. The patient complained of much itching and tingling of the hands, especially at night when she got thoroughly warm. While in the hospital her temperature fluctuated between 98° F. and 100° F.; though as a rule the maximum, which occurred in the evening, was 99·6°. Ammoniated mercury ointment locally, and small doses of

arsenic internally had no effect. The distribution of the eruption in November was as follows :—

Right-hand Dorsum.—The skin over the joint between the first two phalanges of the index finger was thickened and purplish. Passing over the radial side of this thickened skin was the raised defined margin of the projecting portion of eruption from the palm ; the radial side of this finger more distally was also purplish and a little thickened. On the ring finger, there were two large nodules on the back of the terminal phalangeal joint ; they were oval in shape, one-third of an inch in length, and their distal ends converged ; in appearance they reminded one strongly of gouty tophi. There were three smaller nodules on the ulnar side of this joint ; all these nodules were thickenings of the skin, and moved with it ; they were purplish in colour. The ends of the phalanges forming this joint felt distinctly thickened. The little finger was free except for a patch on the back of the metacarpal joint, which was nearly one inch square, and had a defined edge with a purple tint passing into yellow. It turned round to join the eruption on the palm. There was another patch $3\frac{1}{4}$ by $2\frac{1}{2}$ inches on the inner side of the wrist, extending a little above and over the styloid process of the ulna, and forwards over the inner fourth of the wrist. Just below the styloid process towards the palm, the tissues were much thickened, forming a ridgy nodular condition similar to the scar thickening of a burn. There was a second patch over the middle two thirds of the anterior aspect of the wrist.

On the palmar aspect the centre was free, and also a tract of skin passing to the ulnar surface of the hand ; but even here the skin was a little thickened and of yellowish tint. The most deeply congested part was the ball of the thumb and the fore part of the metacarpus ; there was a defined curved edge, concave to the palm, and raised edges along the sides of the fingers, abruptly separating the eruption from the healthy skin on the posterior aspect. This defined edge was much thickened in several places, especially on the radial border of the thumb, where, in fact, it was made up of a series of irregular nodules. The eruption passed back over the knuckle of the thumb to within a quarter of an inch of the raised edge of the eruption, which projected back from the palm to line the sides of the interval between the thumb and first finger. On the left hand, the eruption had a similar distribution. On the dorsum, the metacarpus and the ring-

finger were free, but the middle finger on this side was decidedly thickened, not only in the superficial tissues, but also the bone near the base of the second phalanx.

There was much induration of the skin over the last joint of the index finger, and a small thickened patch over the knuckle. The dorsum of the little finger presented nodular indurations at the sides of the last joints, and over the middle of the first phalangeal joint. At the fold of the thumb, the eruption from the palm projected back as in the right hand.

As regards the palmar surface, there was more nodular thickening on the radial side of the left than of the right index-finger; and the end of the left index-finger was also much thickened.

There were also well defined symmetrical erythematous patches on the backs of the elbows. Some parts were bright in colour, others were fading and becoming yellowish in tint; on the margin of the latter, two or three slightly raised pinkish spots were visible, and a portion of the eruption on the right elbow appeared to be made up of purplish elevated spots; while over the left olecranon there were well marked nodules about the size of peas. Gradations could be traced between the pinkish spots and the large purple nodules, the latter appearing to result from a blending together of the former. There was still some purplish induration of the plantar aspect of the toes, but faint stains and roughness were the only relics of the knee eruption. The changes in colour, in thickness, and in consistence of the skin just described, were the only ones present; enlarged venules were visible; there was no œdema; in no part of the eruption was there any sign of desquamation; the nodules showed no tendency to soften; on the contrary, most of them were as firm as leather, and some as hard as cartilage. It is to be noted also, that the skin in other parts, even in the immediate neighbourhood of the eruption, appeared to be quite healthy, and free from all suspicion of induration, of thickening, or of atrophy; the affected always felt warmer than the unaffected parts. They were not tender to the touch; there was no anæsthesia, the cutaneous sensibility was indeed perfectly natural.

During the period, November 13th to Decembor 24th, that the girl was an in-patient for the second time, the urine was found to contain albumen. The albuminuria was intermittent, being absent some-

times for days; it had no constant relation to meal times, though it occurred oftener after breakfast than at other times of the day; the amount of albumen varied from the merest trace to a measurable quantity. The specific gravity of the urine fluctuated between 1·018 and 1·027.

The deposit from the urine of twenty-four hours was carefully examined, but no renal elements or casts were discovered. There was never a trace of œdema of the legs or other parts. The heart's impulse and sounds were natural. It was again noted that the evening temperature occasionally reached 100° Fahr., but the girl always felt and looked quite well. Since leaving the hospital she attended off and on as an out-patient. During the last three months the eruption had faded in colour, thus the erythematous patches at the elbows and wrists, bright red in August, were in March represented by yellowish stains, and the purplish hue of the palms was much less noticeable. But while the colour had become fainter, the nodular induration had increased, as is well depicted in the photographs taken by Dr. Owen on March 15th, 1889. The lumpy, knotty appearance of the fingers, together with the purplish tint of the palmar aspect of the hands, with the exception of the centre of the palms, were very striking features. The leathery thickening of the skin over the fore part of the palm near the fingers was very remarkable, especially near the root of the right middle finger, and separating this thickened tissue from another portion. Passing back to the ball of the thumb, was a deep groove which was crossed by two cicatricial-like bands. Other parts of the growth also, as over the palmar aspect of the first phalanges, felt, when grasped, like pieces of leather. But although most parts of the original eruption had progressively thickened, and become harder and shrunk at their margins, it was important to observe that in other parts, involution had occurred. Thus, to give one example, it was recorded in November that of the right fingers the end of the middle finger was the most affected, whereas in March, 1889, this finger was quite tapering, and except for the slight change of colour, looked perfectly natural.

Three years later, as recorded by Mr. Hutchinson in the *Archives*, very little change had occurred. Some of the patches had become decidedly thicker and more nodular, and a few had entirely disappeared. The general health was good. In answer to inquiry, Dr.

Bury states that a year ago, *i.e.*, December, 1892, she was in much the same condition.

Bury refers to two cases described by Mr. Hutchinson as resembling his own. The first was described in the Illustrations of Clinical Surgery, plate viii. page 42. The second in the *British Journal of Dermatology*, for November, 1888, under the title of "Symmetrical purple congestion of the skin." But they do not appear quite to correspond with our case, and that of Bury. Still the recapitulation of the leading points may be useful.

The first case was a stout florid man, æt. 58, who had suffered much from gout, but was in otherwise good health and comfortably off. The patches of disease of the skin began two years before he came under notice in January 7th, 1869, remained unaltered during twelve months treatment, and as far as could be ascertained for the remaining six years of his life, his death having been produced by kidney and bladder disease. The lesions of the skin extended rather than declined. The skin lesions began on the left leg, the right having been affected only two months. The distribution was fairly but not strictly symmetrical, on the front of each leg, on the back of each middle finger, involving nearly the whole of the right one, while on the left the patch was limited to the dorsum of the finger just above the knuckle. There were small separate nodules on the backs of the hands, especially on the right. On the left arm there was a small patch above the elbow and another below it, but there were none on the right arm. Both hands were slightly swollen.

The patches began as distinct nodules, but became confluent and lost their nodular character. They were of a dark purple colour, paling by pressure at the margin only; they were irregular in size and shape, distinctly raised with irregular but abruptly defined edge, with usually smooth surface, but sometimes covered with a thin dry epidermic scale. The elevation was chiefly due to œdema, as almost all the thickening could be removed by continued pressure, and there was slight œdema in the neighbouring skin. They were neither tender nor painful. Whilst under observation some fresh patches appeared on the legs; a few at the margins of the earlier patches, but others came separately on the back of the right calf and only one on the left, and once he had gout in the left forefinger. Colchicum, magnesia, arsenic acid, iron mixture, iodide of potassium, and

simple alkaline mixture, were all given without effect on the eruption.

Mr. Hutchinson records that he has seen a drawing belonging to Professor Boeck of Christiania, exactly resembling his case and equally persistent. The patient was an adult sailor apparently in good health.

The case recorded in the first volume of the *British Journal of Dermatology* is regarded by Mr. Hutchinson as a much more developed example of the same character as the first.

Mr. B. was a florid healthy looking farmer, æt. 65, without any apparent disease besides that of his skin. He ascribed his illness to a deep cut by a chaff-cutting machine received in the right wrist twelve years before, which led to ankylosis of the joint. Soon after this the right hand became dusky, and the left only several years later. Some years after the first injury he sustained another incised wound on the right leg, and this too was followed by lividity of the limb, but the scar did not became indurated.

Two more cases of an earlier and therefore milder form are recorded by Mr. Hutchinson, in the first volume of his Archives of Surgery, p. 372. One was a florid large framed man, æt. 65, who had lived freely and often suffered from gout. He was a great rider, and on the inner side of the left knee where it gripped the saddle, a purple patch had formed within a couple of years, and had recently increased to the size of the palm. It was raised so that it was a quarter of an inch thick, had well defined borders, and was of a dusky purple tint. It itched a great deal at times, but there were no other symptoms or lesions.

The next case was also a florid man, æt. 56, who had had good health with the exception of repeated attacks of slight gout in his foot and hand. He also inherited gout from both sides.

For two years a dusky purple patch had been forming in the middle of the forehead where the hat pressed. It had thickened, well-defined borders of a dusky purple tint, and perfectly smooth surface. The natural wrinkles were obliterated, but the orifices of the hair-follicles were more conspicuous than normal.

Near the major patch was another the size of a shilling which had recently developed and was exactly like the original one.

Two little patches of doubtful nature had formed quite lately on the wrist-joints.

All four of these cases of Mr. Hutchinson possess the following points in common :—

They were all men of florid complexion over 50 years of age (the youngest was 56). All were chronic sufferers from gout; all had one or more raised purple or plum-coloured patches, much of which was due to venous congestion, this feature, Mr. Hutchinson thinks, being the direct outcome of the gouty diathesis ; they resisted treatment of all kinds, and slowly spread over the body. In two, pressure or friction was thought to be the localizing cause ; in one, the patches first came below an incised wound. In Boeck's case particulars are too scanty for discussion, beyond that he was an adult sailor, and therefore very liable to injury. They did not appear to select the articulations, most of them coming out over the tibia, forehead or similar flat surface.

Since the first portion of this paper was written, I have seen the drawing of another case like Bury's and my own, in a woman of about 22, in whom it had been present, I believe, about five years, but it will no doubt be published before long.

The cases of this form have the following features in common :—

All of them were of the female sex, and the disease commenced in childhood or early youth. Of the two published cases, Bury's had had acute rheumatism; and in mine there was a strong family history of gout and rheumatism, though the patient herself had not had any rheumatism. Bury's case had intermittent albuminuria. In both, the lesions developed over the articular prominences of the fingers, the elbows and knees, and also on the palms. In addition, in Bury's case, the toes were affected ; in mine, the buttocks.

In both, the lesions were primarily nodular, with a tendency to coalesce into an elevated infiltration, most marked on the palmar surface.

In both, there was a tendency to involution on the elbows and knees, but on the hands in Bury's case the tendency was to further development, while in mine involution occurred in the hands also, and there is some hope of complete recovery.

Nevertheless, the persistence of many of the lesions is a very

striking feature in the disease, and on the whole greatly exceeds the tendency to involution.

While the primary tint of the lesions is pink, they soon acquire a purplish hue, which becomes more marked the longer the duration of the lesions; how very long that may be, is strikingly shown in Bury's and the unpublished case, well earning the title of 'diutinum', or persistent. The older lesions become much firmer to the touch, almost cartilaginous, but at no time are they otherwise than firm and incompressible. The sharp line of demarcation between the diseased and healthy skin is to be noted.

There can be no question about the identity of the nature of these two published and the one unpublished case, but with regard to Mr. Hutchinson's type, the differences between that and what in justice to the first recorder I may call the Bury type, are considerable, and we may compare them in parallel columns as follows:—

Bury Type.	Hutchinson Type.
All Females.	All Males.
All Young.	All Elderly.
Gouty or Rheumatic in Self or Family.	Very Gouty personally.
Lesions situated over articulations and on the palmar surface.	Lesions not any special localization or over flat bony surfaces.
Began as nodules, but became confluent, still showing nodular character.	Began as flattish nodules, became confluent, and lost their nodular character.
Erythematous at first, but becoming purplish.	Purplish from the first.
Lesions, some very persistent, others involuting. Never spread over a large area.	All lesions persistent throughout life, and spreading widely.
Lesions paled but very firm, and otherwise unaffected by pressure.	Lesions paled only at the border. The elevation was chiefly due to œdema, and could be almost removed by continued pressure, and there was œdema in the neighbourhood.

The two types differ in the age and sex of the patient, in the position of the lesions, and in the older cases the nodular character was less developed, being œdematous, and lacking the firmness of the Bury type of the nodules and patches.

They resemble each other in the gouty diathesis of the patients, their being primarily nodular in character, becoming confluent and

forming patches, and in the purple tint, the age of the patients perhaps acounting for the deeper hue of the Hutchinson type.

I think it is evident, therefore, that at present it is not safe either to affirm or deny that the two types may be phases only of one affection; but until, and perhaps even after, we have discovered connecting links, it will conduce to clearness of conception to regard them as distinct types, and not to endeavour to weld all the symptoms of the two varieties into one comprehensive description.

It may be safely assumed that the number of cases is too small to afford us a complete symptomology of these forms of disease, which can so clearly be traced to what French writers would term "arthritism."

The microscopic examination of the portion of skin excised from

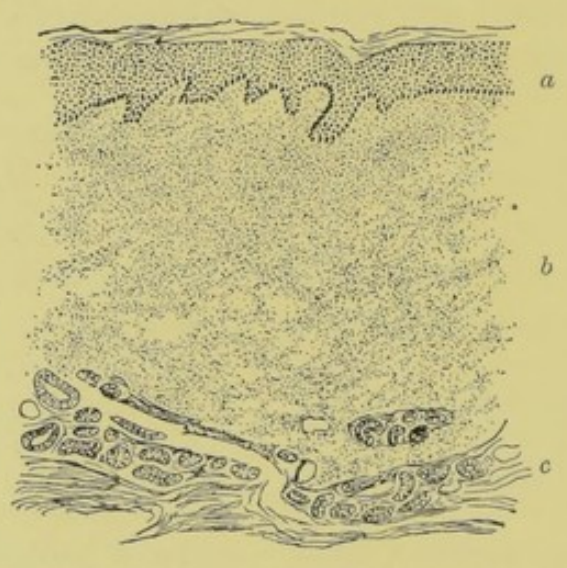

FIG. 1.—*a*, Epidermis normal; *b*, Area of morbid process; *c*, Coil glands and corium below, normal. The fibrous tissue between the cells is not visible with the low magnifying power. × about 40.

the knuckle showed that the epidermis was unchanged, except, perhaps, a slight thickening of the rete mucosum and elongation of the interpapillary processes in some parts. The seat of the morbid process was between the epidermis and the deep portion of the corium immediately adjacent to the coil-glands, all below these glands being normal. (Fig. I.)

Within these limits was a fibro-cellular structure, which in great part replaced the normal fibres of the corium, such portions as were present taking the carmine stains more deeply than the new tissue.

The fibres ran in various directions, following the course of the vessels. They were horizontal above, immediately below the papillary layer, vertical or oblique, and then branching horizontally in the deep part.

The cells permeated the interstices between the fibres, sometimes aggregated into clumps, and in these parts the cell structure predominated, in other parts, the cells were placed singly between the meshes of the fibres, and then these latter were the predominating feature. (Fig. II.)

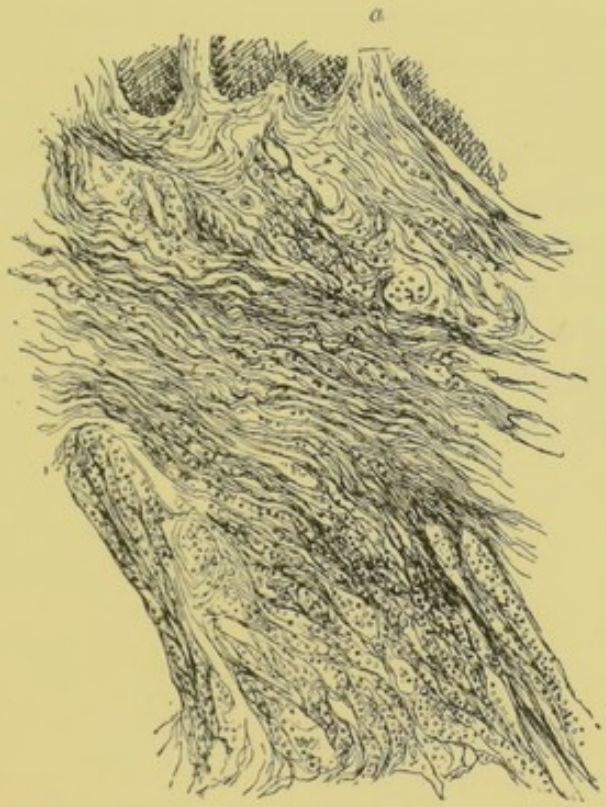

FIG. 2.—To show fibrocellular structure. The cells were somewhat more abundant than depicted in the upper half of the drawing. *a*, Lower end of interpapillary processes of the rete.

The greater part of the section was in the same plane as the fibres, but in some places it was transverse to their direction, and then an adenoid appearance was presented, single cells lying in a fine fibrous meshwork. There was great variation in the density of the cells in different portions of the section. In the centre, they were rather closely aggregated, but still much less than in an acute form of inflammation, and towards the periphery, they were more disseminated between the fibres.

There was slight periglandular infiltration in a few of the top segments of the sweat-coils, but practically the sweat-coils as a whole might be considered to be normal. There were no hair-follicles or sebaceous glands to be seen in the sections.

The walls of the vessels were normal, except slight cellular infiltration in the outer coat, but there was no thickening of the walls or marked dilatation of the lumen.

On the whole, therefore, the microscope reveals only what might have been anticipated from the clinical characters of the lesion, viz., a chronic inflammatory process in which both plexuses of the corium took part, but only the upper wall of the deep plexus was concerned. The older the lesion, the more would the new fibrous tissue be developed, until, in cases like that of Bury, one would anticipate that many of the patches would be composed almost entirely of fibrous tissue, and perhaps not unlike a keloid in structure.

Practically, the microscope does not explain the real pathogeny of the process, and the name placed at the head of the paper is justified simply by the clinical features, which are those of an erythematous, raised, persistent lesion.