FORDYCE (J. A.)

ANGIOKERATOMA OF THE SCROTUM
RAYNAUD'S DISEASE OF THE EARS
LUPUS ERYTHEMATOSUS DISSEMINATUS DISAPPEARING DURING PREGNANCY

BY

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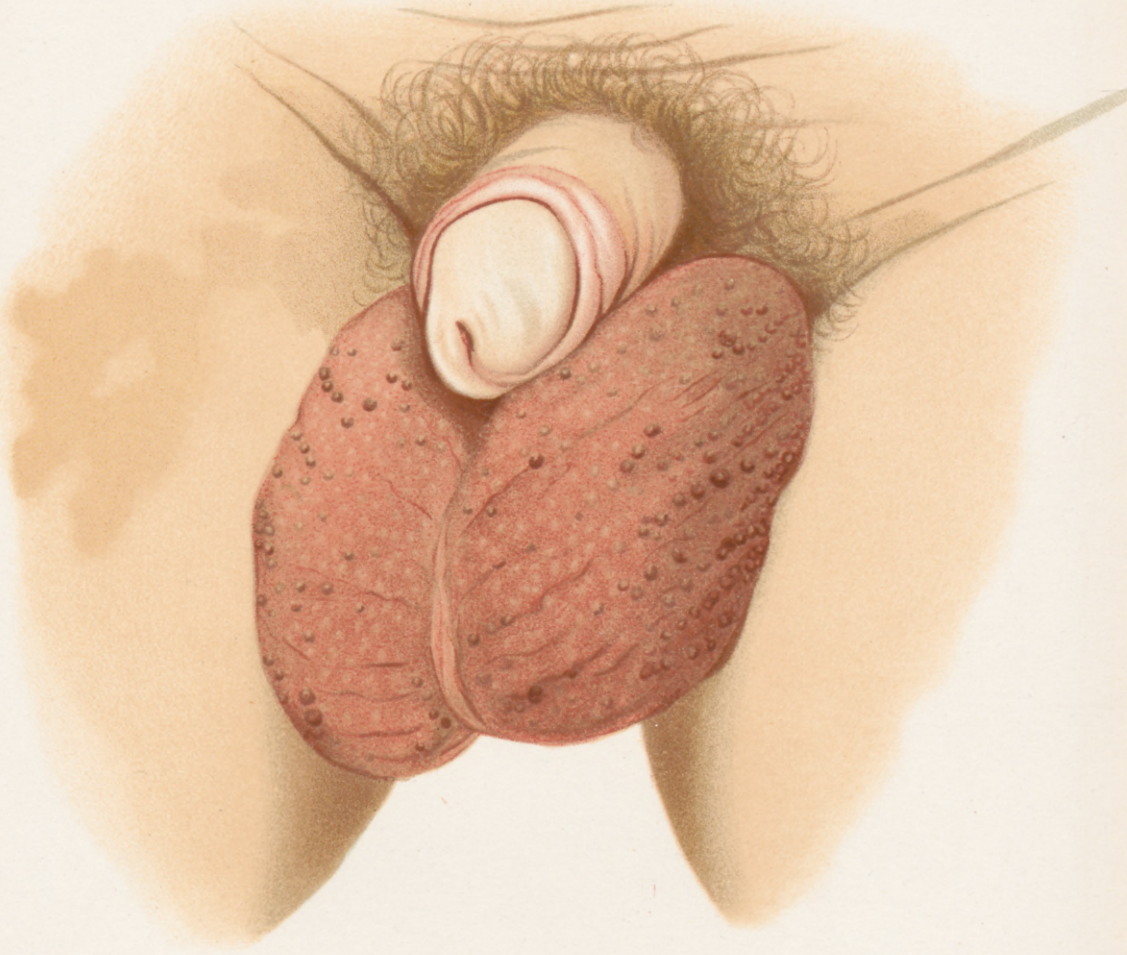

DR. FORDYCE'S CASE OF
ANGIOKERATOMA OF THE SCROTUM

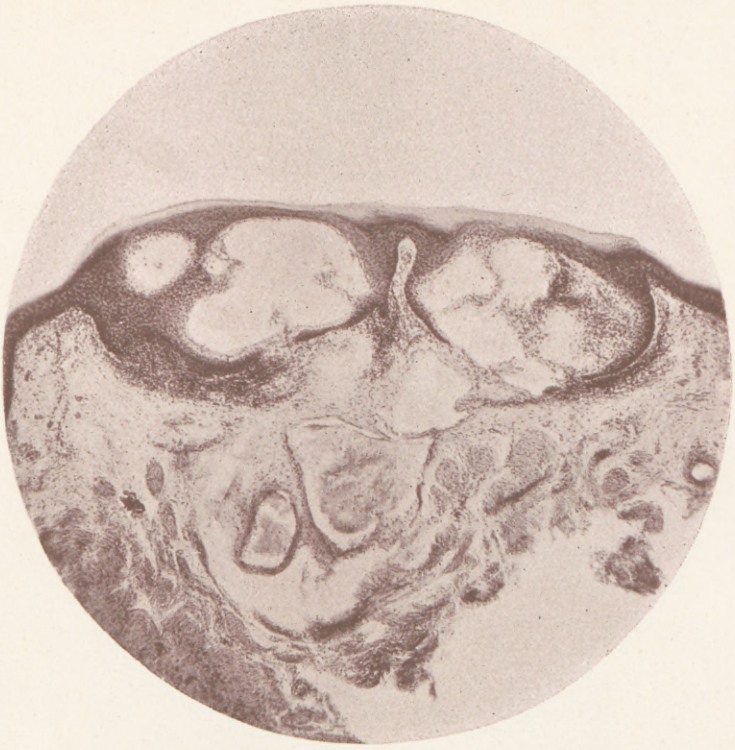

FIG. 1.—Section through two adjacent tumors.
Spencer 1 in. *Zeiss* projection ocular 2. × 50.

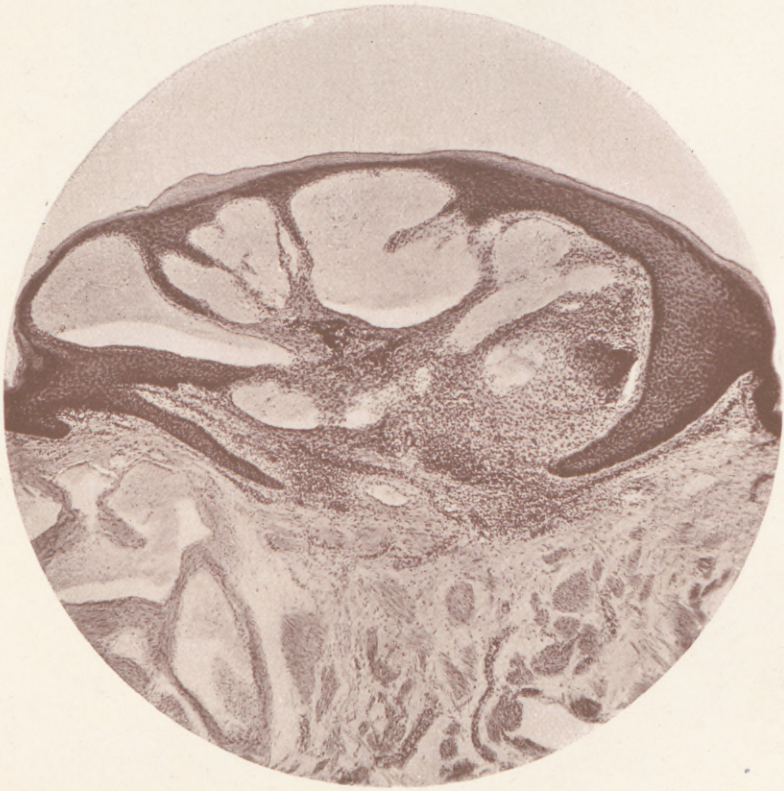

FIG. 3.—Section through larger tumor, showing papillary vascular dilatation and hypertrophy of the rete Malpighii.
Spencer 1 in. *Zeiss* projection ocular 2. × 50.

ILLUSTRATING DR. FORDYCE'S ARTICLE ON
(Photomicrographs b

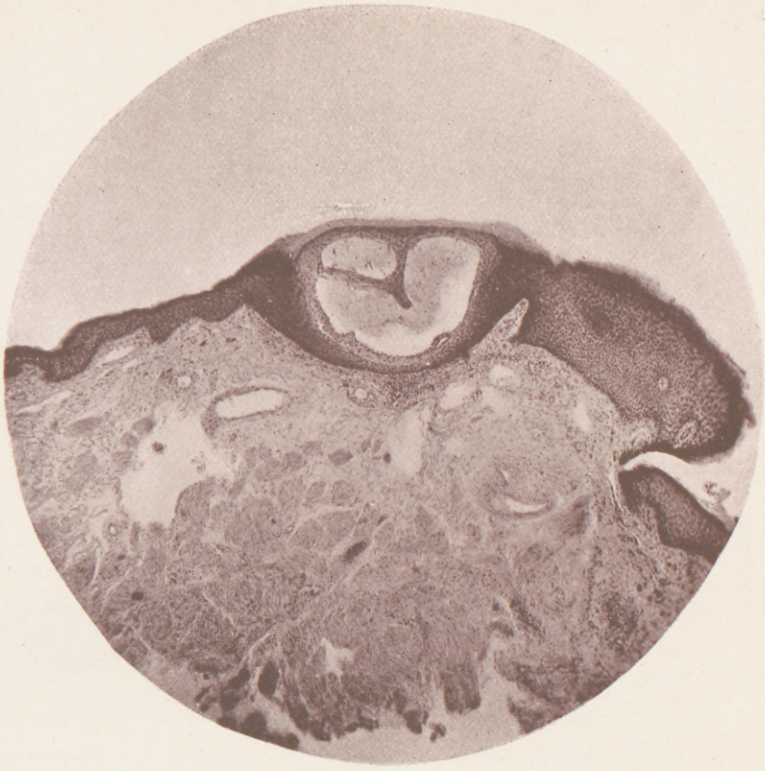

FIG. 2.—Section through small tumor completely inclosed by the hypertrophied rete Malpighii.
Spencer 1 in. Ocular 1¼ in. × 60.

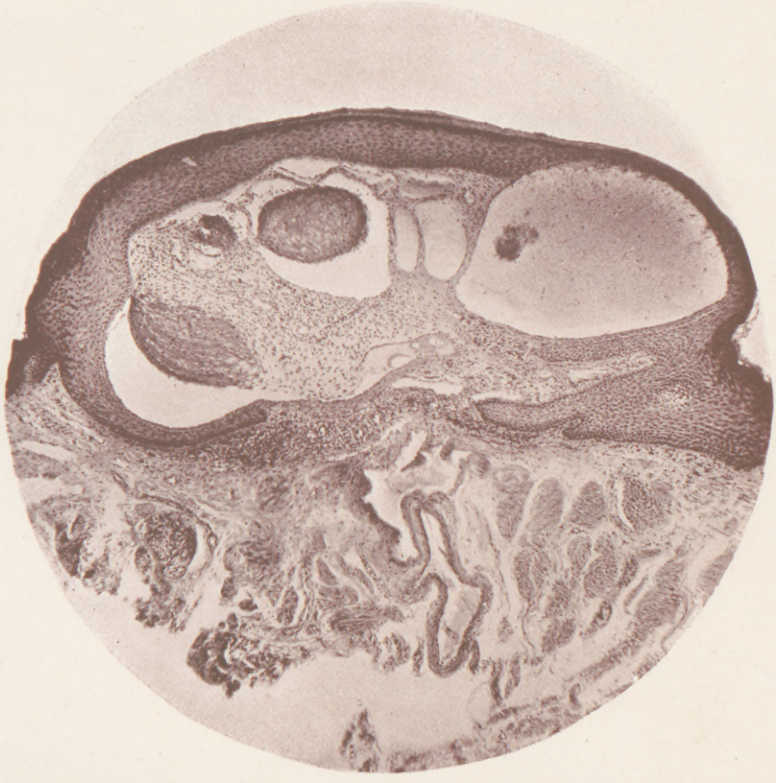

FIG. 4.—Section through larger tumor, showing cavernous spaces divided by septa, and containing organized blood clots.
Spencer 1 in. Ocular 1¼. × 75.

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ANGIOKERATOMA OF THE SCROTUM. RAYNAUD'S DISEASE OF THE EARS. LUPUS ERYTHEMATOSUS DISSEMINATUS DISAPPEARING DURING PREGNANCY.*

BY J. A. FORDYCE, M. D.,

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THE patient, a male aged sixty, was admitted to the City Hospital in the spring of 1894 for some urinary trouble. He was somewhat feeble both mentally and physically, and could give little information regarding the eruption which was present on his scrotum, except to say that he had had it for a number of years.

He feared that some harm would come to him if he remained in the hospital, and demanded his discharge before a careful study could be made of the interesting skin lesions which he presented.

An opportunity was afforded, however, of excising a number of the small tumors for microscopic purposes, and of having made the colored sketch, which is reproduced in connection with the present article.

The skin covering the thighs and lower part of the abdomen was the seat of a number of patches of vitiligo. The patient was also affected with a double varicocele, which is interesting in connection with the superficial vascular dilatation. His hands and feet were free from any skin affection, and he denied having suffered with chilblains. The scrotum, especially on its lateral and posterior surfaces, was the seat of a great number of small, spherical-shaped, dark purple tumors (see colored plate). They were arranged in a linear manner as if following

* Read at the nineteenth annual meeting of the American Dermatological Association, Montreal, Canada, September, 1895.

the superficial vascular supply of the parts. The small growths were from a pin's head to several times that size, their dimensions being pretty uniform. Pressure caused the color of the majority of the tumors to disappear. In some of them, however, the color was only partially lost.

The small growths were distinctly elevated above the surface of the scrotum, seeming to rest on it rather than to be imbedded in the skin. Some of them were covered by a slightly thickened horny layer, under which minute dark-red points could be seen, which gave the tumors a wartylike appearance.

Composite lesions were formed by the union of two or more of the smaller ones, which, however, preserved all the characteristics of the original growths. No pain, pruritus, or other subjective sensations were complained of by the patient. Puncture of the tumors with a needle was followed by slight hemorrhage.

In the cases heretofore reported the eruption has been, with few exceptions, confined to the extremities.

Dr. Zeisler's case * presented, in addition to characteristic lesions on the hands and feet, nævuslike patches and pedunculated vascular tumors on the forearms, over the patellæ, the legs, thighs, and auricles.

It is probable that the same pathological condition produced the vascular dilatation in these various localities, the hypertrophy of the epidermis covering the lesions on the extremities being favored by the anatomical condition of the parts and exposed position of the vascular tumors.

The presence of patches of vitiligo in Dr. Zeisler's case and my own, while probably a coincidence only, is yet worthy of notice. In most of the cases reported chilblains preceded the appearance of the angiomatous tumors, and the damaged state of the blood-vessels is looked upon as a strong predisposing factor to their development; the localization of the affection on the hands and feet is therefore readily explained.

The unusual site of the affection on the scrotum in my case demands a careful study of the histological appearances, and a comparison with the minute structure of the cases which have occurred in typical localities. The usual ætiological factor, chilblains, can not, of course, be invoked to explain the occurrence of the localized vascular tumors on the scrotum. The tendency to dilatation of the blood-vessels as manifested by the double varicocele, and the degenerative state

* *Transactions of the American Dermatological Association*, Seventeenth Annual Meeting, 1893.

of the vessels and surrounding connective tissue incident to old age, were probably the most potent causes in bringing about the condition in question.

The dilatation of the capillary vessels is looked upon by all who have studied the affection as the primary and essential condition, the change in the epidermis being of a secondary nature.

If we adopt this view, which is doubtless the true one, any dilatation of the papillary loops should in time be followed by thickening of the epidermis and the other anatomical changes which are found in this affection.

Mibelli * gave the first anatomical description of the condition found in this affection, and proposed the name "angiokeratoma" for the disease. The lesions which formed the basis of his observation occurred on the dorsal surface of the fingers of a fourteen-year-old girl and had existed for several years. They were preceded by chilblains.

The stratum corneum was so much thickened in this case that the gross appearances of many of the lesions suggested the diagnosis of keratoma. The name proposed by Mibelli is a very appropriate one, as it accurately indicates the pathological condition which exists.

Before Mibelli's careful investigation, cases of the same affection had been noted by other writers under various names; the true character of the lesions had not, however, been determined.

Cottle † seems to have made the first observation regarding the disease. Crocker ‡ refers to a case which he described in the first edition of his text-book under the heading of Verruca, a further history of which he gives in the *British Journal of Dermatology* for 1891.

Colcott Fox # described a number of cases of the disease in 1886 and in 1889; in the latter year, under the name of Cases of Lymphangiectasis of the Hands and Feet in Children. Since the publication of Mibelli and Pringle's articles he admits the inaccuracy of his observation regarding the histology of the lesions, and agrees fully with these investigators regarding the morbid changes and the name proposed. Dubreuilh ‖ described the disease under the title of Telangiectatic Wart. This case is in every way a typical one, the patient affected being a young girl who had previously suffered with chilblains.

* *Giornale italiano delle mal. ven. e della pelle.* Fasc. iii, September, 1889. *Internat. Atlas of Rare Skin Diseases*, No. II, 1889.

† *St. George's Hospital Reports*, vol. ix, 1877–'78, p. 758. Quoted by Crocker.

‡ *Diseases of the Skin*, second edition, 1893.

These cases are referred to at length in Pringle's article on Angiokeratoma, *British Journal of Dermatology*, 1891.

‖ *Annales de la Polyclinique de Bordeaux*, 1889.

We are indebted to Pringle's* article for a most accurate and painstaking description of the clinical appearances and morbid anatomy of the affection, as well as for an analysis of most of the cases which had been met with up to the time of its publication.

Pringle reported two cases affecting girls with chilblains, and his histological description agrees in all essential points with that previously made by Mibelli.

Since the publication of this article cases have been reported by Audry and Deydier,† Brocq,‡ Thibierge,# Joseph,‖ Tommasoli,^ Renault,◊ and others, in addition to which several instances of the disease have been observed but not published.

Histology.—For microscopic study a number of tumors of various sizes were excised from the scrotum, hardened in absolute alcohol, and stained with different coloring agents.

A section through two small tumors situated side by side is shown under a low power in Fig. 1 (plate).

Two cavernous spaces containing blood are seen directly beneath the epidermis, divided by irregular septa and bounded on all sides, with the exception of the low portion of one of them, by epidermic cells.

Directly above the lacunar dilatation the rete layer is thinned as if from pressure of the distending cavity, while the stratum corneum is shown somewhat hypertrophied. At the lateral margins the rete mucosum is considerably hypertrophied, which is more evident in Fig. 2 (plate), representing a single tumor entirely surrounded by the proliferated rete layer.

At the right of the blood cavity the thickened rete is well shown.

The cavernous spaces are filled with red and white blood-corpuscles in normal proportions, and under a low power their lining membrane seems to be made up of epithelial cells.

With high amplification, however, they are found to be lined with a greatly distended connective-tissue layer which separates the blood from the epidermis. In certain sections blood spaces are found to exist in the epidermis, surrounded only by disintegrated epithelium, as if the blood had penetrated between the cells and produced the cavities in

* *British Journal of Dermatology*, 1891.

† *Lyon méd.*, 1892. *Ann. de dermat. et de syph.*, 1893, p. 381.

‡ *Ann. de dermat. et de syph.*, 1892, p. 819.

Ann. de dermat. et de syph., 1892, p. 1139.

‖ *Berl. klin. Woch.*, 1892, p. 493. *Dermat. Zeitschrift*, 1894, p. 16.

^ *Commentario clinico delle malattie cutanee e genito-urinarie*, 1893.

◊ *Ann. de dermat. et de syph.*, 1894, p. 1248.

question by a destructive action on the epidermis rather than by stimulating the growth of the rete layer, as seems to be the case in the formation of most of the cavernous spaces.

Beneath the lower margin of the proliferated rete a round-celled infiltration is present, which is distinctly shown in Figs. 1, 2, and 3 (plate).

Blood pigment is intermingled with the cell infiltration, and there is some indication of fibroid-tissue formation about some of the subpapillary blood-vessels. There is also to be observed in this area a considerable vascular dilatation of the same character as shown in the papillary region (Figs. 3 and 4, plate).

Figures 3 and 4 represent sections through larger tumors, the cavernous blood spaces being more distinctly shown, as is also the elevation of the tumors above the level of the skin.

The lower border of these tumors is not completely encircled by the hypertrophied and proliferating rete Malpighii, being formed by the subpapillary portion of the derma in which are seen greatly dilated blood spaces.

On the right of Fig. 4 a large cavernous space is seen to be filled with blood-corpuscles, which have by pressure caused a marked atrophy of the epidermis.

On the left of this section the circulation has been obliterated, as the lacunæ are occupied by concentric layers of fibrin containing blood-corpuscles and pigment.

Fig. 5 represents a more enlarged view of the cavernous spaces with their dividing septa. The stratum corneum is also shown to be considerably thickened.

An examination of the sections shows that the lesions consist of lacunar spaces filled with blood occupying the papillary portion of the derma, some of which are inclosed in the rete Malpighii. These cavernous spaces are evidently the essential feature of the disease, and are probably the primary pathological condition.

It is the opinion of Mibelli and Pringle that these lacunar dilatations result from changes in the papillary vessels, gradually brought about by repeated attacks of chilblains which impair the contractility of their walls and cause permanent telangiectases.

While in my case such a cause had not been present, it is not unreasonable to suppose that the tendency to vascular dilatation, and the impairment of the normal connective-tissue support to the vascular walls which was shown in a marked atrophy of the scrotal tissues, were the active agents in bringing about the condition described.

It is the opinion of both the gentlemen formerly quoted that the

primary pathological change is vascular and the altered appearance of the epidermis a secondary phenomenon.

My own sections would lead me to coincide in this view and also in Pringle's hypothesis that the blood spaces in the rete Malpighii are caused by a downgrowth of the cells of this layer, producing a constriction of the terminal loops and their resulting distention.

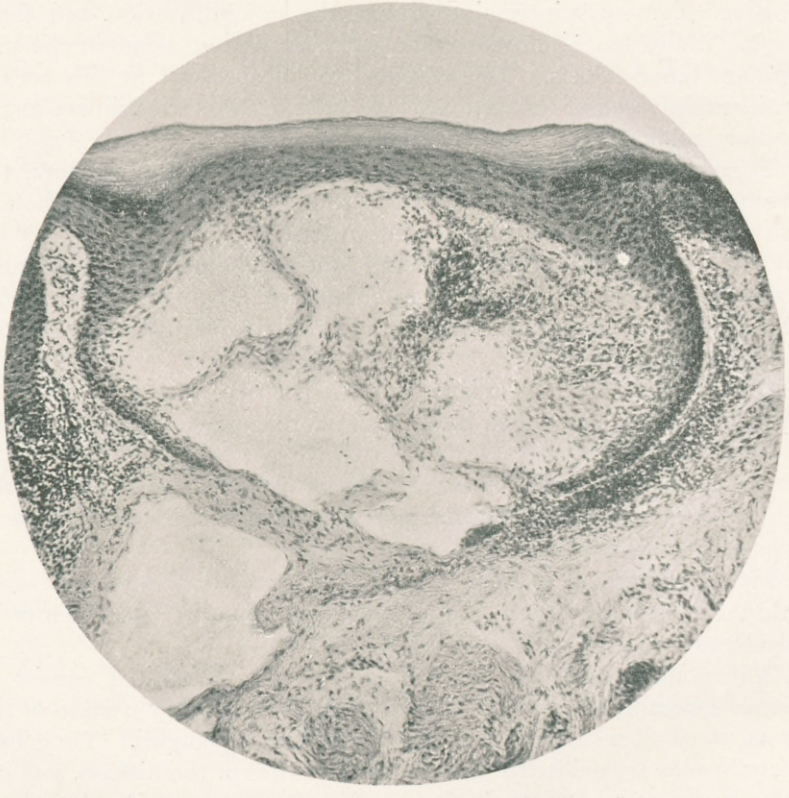

FIG. 5.—Spencer, half inch. Projection ocular 2, Zeiss. × 175. Cavernous space filled with blood corpuscles and divided by fibrous septa. Hypertrophy of the stratum corneum and rete Malpighii.

My case differs from those previously reported in the peculiar localization of the tumors, the absence of chilblains as a cause, and in the minor grade of keratosis.

When we compare the normal thickness of the horny layer over the hands with that on the scrotum, this discrepancy can readily be explained. In all the essential histological features, viz, the presence of blood spaces in the epidermis and papillary region, the prolifera-

tion of the rete Malpighii and in the presence of an inflammatory infiltration in the derma, it corresponds very closely with the classical cases reported by Mibelli and Pringle.

The tumors have been successfully treated by electrolytic puncture with the negative needle and by the galvano-cautery.

Raynaud's Disease of the Ears.

The patient, a male, aged thirty-nine, was first seen in March of the present year (1895.) His habits and family history are good. He is able to do physical work, and does not suffer with headaches. He describes a venereal sore eight years ago, which he states was followed by loss of hair, sore mouth, and sores on his legs. He also had some affection of the eyes after his venereal infection. Had gonorrhœa thirteen years ago, and again five years ago, both of which attacks were followed by rheumatism in the wrist and small joints of the fingers. He has never suffered in a marked degree with cold hands or feet.

His present trouble began in August, 1894, on a warm day. The ears at this time became cold and blue, and remained so for a period of several hours. They then gradually resumed their normal color.

Such attacks were frequent until January, 1895, when that portion of the auricle shown in the illustration assumed a permanent bluish-black color, which persisted up to the time he came under my observation in March of the present year. He states that the ears feel numb and cold, and that a burning pain is frequently present in them.

Present Condition.—The colored illustrations which I show will give a better idea of the symmetrical location of the affection than any description could accomplish. (Illustrations shown at the meeting.)

Both ears were distinctly cold, and of a purplish-blue color for some distance around the seat of the gangrene. The gangrenous area had formed at the junction of the upper and middle thirds of the auricle, and occupied a surface about half an inch in diameter (see cut).

It was bluish-black in color, dry, and without odor or suppuration. On scraping it, the diseased tissue could be readily removed, and appeared more like coagulated blood than like a sloughing mass. Little or no pain was produced by attempting to remove the dead tissue.

The purplish color extended for a considerable distance around the imperfect line of demarcation.

Both eyes showed the scars of an interstitial keratitis which the patient refers to the syphilis of eight years ago.

The patient was given no internal medicine; he was directed to use a simple ointment and report at the clinic within a week.

He was not seen again until August 30th, when no trace of the

disease was found except superficial cicatrices on the ears, the scar on the left side being more noticeable than on the right.

The interest of the case, aside from the somewhat unusual site of the gangrene, lies in the possible ætiological relationship of syphilis to

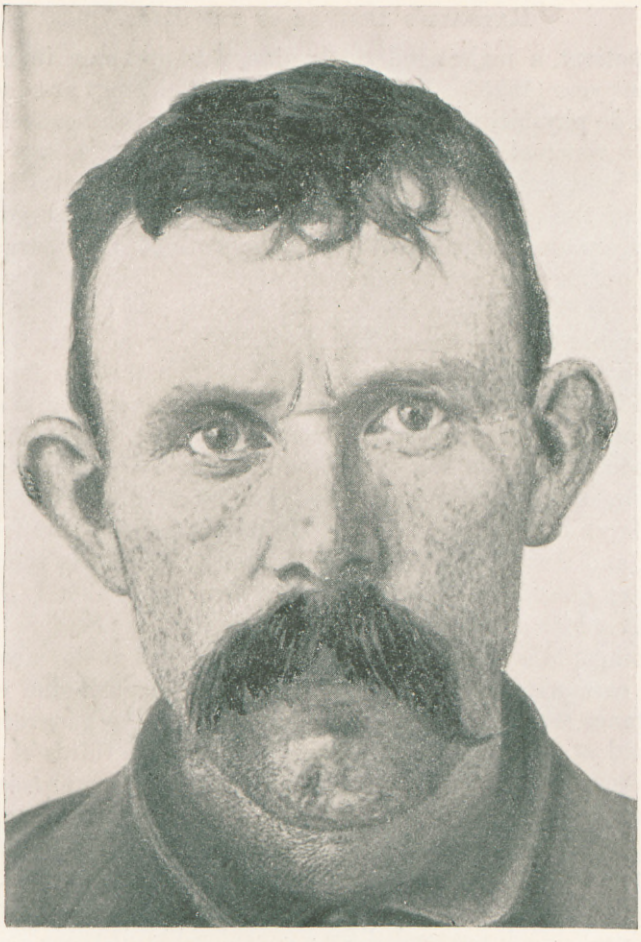

FIG. 6.—Raynaud's disease of the ears; showing the gangrenous areas.

the local disease. The interest of this case is strengthened by the observation recently made by an English surgeon, Mr. Wherry (*Clinical Sketches*, August, 1895), of the occurrence of Raynaud's disease of the fingers, toes, and ears in a fourteen-year-old boy, the subject of hereditary syphilis, who also presented the scars of an interstitial keratitis.

The implication of the arterial coats in both early and late syphilis, leading to lessened and obstructed blood supply, is doubtless responsible for many of the cases of Raynaud's disease in patients with syphilitic disease. When in addition to this obstructive endarteritis there is an added element of vascular spasm, due to cold or other causes, all the conditions necessary to bring about the localized gangrene are present.

It is not the intention of the writer to enter into a consideration of the ætiology of Raynaud's disease, which is doubtless the result of many conditions, and may be due in certain instances to a vascular spasm, pure and simple, brought about through disturbance of the central or peripheral vaso-motor apparatus.

Lupus Erythematosus Disseminatus disappearing during Pregnancy.

Mrs. F., aged twenty-five, married eight months. Six months pregnant. Has always enjoyed good health. No skin disease before the present one. She had an erythematous eruption on the face for some months before her marriage, which was attended by marked pruritus.

On the hands and arms the affection first appeared, two months after her marriage, at the beginning of her pregnancy.

Examination shows a general hyperæmia of the face, uniform in character, which is free from any evidence of atrophy, except at the margins of the erythema, below the ears, and on the neck. Even here the cicatricial depression is so slight as to escape notice, except on very careful examination.

There are no spots on the scalp, and none on the body, except those shown in the colored drawings. (Drawings shown at meeting.)

On the backs of the hands and forearms the eruption is symmetrical, showing distinct atrophy of the skin in the larger patches, and to a less extent in some of the smaller ones. The edges of the patches show very slight infiltration and no scaling. The entire process impresses one as a mild type of hyperæmia, which has resulted slight atrophy of the central parts of the lesions.

The patient was examined again shortly before these notes were taken, when almost at term. At this time all evidence of the disease had disappeared, except the atrophied spots, which were surrounded by a pigmented zone.

In this connection I recall the case of a woman in the South whose husband wrote me in regard to the treatment of his wife for a lupus erythematosus of the face. He stated that the eruption had entirely disappeared during a recent pregnancy, but had recurred in a form equally severe after her confinement.