

Fordyce (J. A.)

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OF THE SKIN (KAPOSI).

BY
J. A. FORDYCE, M.D. ✓

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DR. FORDYCE'S CASE OF IDIOPATHIC MULTIPLE PIGMENTED SARCOMA OF THE SKIN.

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BY

J. A. FORDYCE, M.D.

TYPICAL examples of this affection are sufficiently uncommon to justify me in placing the following case upon record:

The patient, R. B., is a muscular Italian sailor, of medium height, energetic and intelligent, who had been six weeks in America when he came under my observation in September, 1890. He is not addicted to the excessive use of alcoholic drinks. He has been married twice and has a family of healthy children.

About fifteen years ago he had several attacks of malarial fever; in 1862 he had gonorrhoea; in 1876, a venereal sore followed by a suppurating bubo. Twelve years ago, in Malta, he claims to have had quite a profuse hemorrhage from the dorsum of the right hand, which recurred at about the same period for several years, and was not preceded by an injury or a lesion of any kind. His present affliction began two years ago last May, and is attributed by him to a mental shock. He returned home from a voyage and found that his wife had just died from an acute malady. The shock was so great that he lost consciousness. Within a day or two he had attacks of bleeding from the nose, which continued at intervals for some weeks; at the same time he noticed when he rubbed or washed his lips that bleeding took place from them.

On the cessation of these hemorrhages, a few weeks after the mental shock mentioned, he noticed the presence of dark red spots on the calf of the left leg, followed within a day or two by the same appearances on the left thigh, then on the right leg and thigh, and finally on the right and left arms and hands similar tumors appeared. When first seen the tumors were of lighter color than at present, but no change in their size has taken place. Itching has been noticed at times, and now and then is a distressing symptom. In less than one



week after the tumors were noticed on the calf of the left leg all of the others had developed in the order named, and no additional ones appeared until about six weeks ago, when an elevated bluish-red growth developed on the dorsum of the right hand near the ulnar side. Since the onset of his skin eruption he has had no hemorrhages from the nose, lips, or from any mucous membranes. His general health has continued fairly good, but he says that he has lost in weight; to what extent, however, he is unable to state.

During the four months in which I have observed him he has been engaged in selling fruit and working as a day laborer on the streets.

After a day of hard work, I have noted that his hands and feet were markedly œdematous and that their local temperature was elevated. He says that he has not the endurance that he once had, but yet considers himself capable of doing ordinary work.

Present Condition.—On stripping the patient the tumors are seen to be confined to the extremities, the face, neck, and trunk being quite free from them. They are limited, moreover, to certain definite portions of the extremities, extending upward, somewhat beyond the middle third of the thighs, and not beyond the lower half of the upper arms. The almost symmetrical arrangement of the growths is a feature which strikes the observer at a glance, the tumors on one half of the body having almost their counterparts on the other half.

On the left side they are somewhat more numerous and larger than upon the right, this being especially noticeable upon the forearm. The symmetry of the disease is shown in the accompanying colored plate, though illustrated better in the patient, as some of the less pigmented spots are not well shown.

On the dorsal surfaces of both feet narrow bands of bluish-black infiltration, from five to seven centimetres in length, are to be seen, and a diffused infiltration is noticeable on the posterior and outer aspect of the left ankle. The dorsal surfaces of the hands and fingers are the seat of numerous nodular masses, dark blue in color, and on the ulnar side of the right palm a tumor one centimetre in diameter with two or three smaller ones are seen, looking not unlike a papulo-squamous syphilide. The growths vary much in shape, size, color, and consistency; on the legs they are darker, many being almost black, and retaining their color upon pressure. On the anterior aspects of the legs and thighs, farther removed from the extremities, their color is more reddish-brown. The darker tumors on the posterior aspects of the legs and thighs are irregularly oval in outline, firm and somewhat elastic in consistence, and show in places a distinct central depression with slight scaliness, as if in the process of involution. At the periphery of some of these depressed patches more recent nodules of a lighter color are seen, as if the disease were starting

afresh. The growths in general are firmly situated in the cutaneous tissue, movable with the skin and not very sharply defined. On the hands and arms the disease has more the character of a diffused infiltration, though showing distinct nodular formation at the periphery and over many of the patches. Both wrists are almost encircled by bands, three to four centimetres wide, of this nodular infiltration, and several narrower bands extend transversely across the forearms. The tumors here are, as a rule, more elevated than those upon the legs, their color varying from a brownish-red to a dark purple, and fading slightly upon firm pressure. The epidermis is glossy in places, as if from over-distention, and the tumors look not unlike angiomas. No evidence of past or present ulceration could be seen in any of the growths. Many of the tumors on the lower extremities impressed one as having passed their active period of growth; while those upon the hands and arms appeared to be in a more early stage of development. The tumors are slightly painful to touch.

The inguinal and epitrochlear glands were enlarged to about the size of a small marble, quite hard and oval in outline.

No abnormality could be detected in the heart or abdominal organs, and the urine was found to be quite normal.

Histology.—The recent tumor from the dorsum of the right hand and a portion from a patch on the right forearm were removed by the cutaneous punch, hardened in alcohol, and stained in a variety of ways; with borax-carmin, hæmatoxylin and eosin, hæmatoxylin and picro-carmin, and with safranin.

The best results were obtained from the combination of hæmatoxylin and picro-carmin, which gave a beautiful double stain, the cell nuclei being colored by the hæmatoxylin, while the intercellular substance was rendered visible by the diluted picro-carmin.

In the epidermis the only changes noted were a slight thickening of the horny layer, a deep pigmentation of the cylindrical cells of the rete; the papillæ were not well defined, and nowhere was there any ingrowth or proliferation of the rete cells. The cutis and subcutaneous tissue were replaced by a new growth which extended from one-fourth to one-half a centimetre below the surface. It began directly under the epidermis and was lost in the subcutaneous connective tissue. Under a low power the structure of the tumor is seen to be composed of small fusiform cells arranged in bundles extending longitudinally, transversely, and obliquely; their transverse sections looking not unlike round cells.

A striking feature of the microscopic picture was the large number of blood-vessels, around and between which are grouped the spindle cells of the tumor simulating very closely the structure of recent

cavernous angiomata. The endothelium of some of the vessels had undergone active proliferation, showing several concentric layers of round cells.

In one vessel whose calibre was almost obliterated, the several innermost layers were made up of round cells, while without these, but still in the vessel's walls, the cells became elongated and finally spindle-shaped.

In a number of these cells, as well as in the spindle-cells of the growth, the karyokinetic figures could be seen. Blood-pigment and partially degenerated corpuscles were found in the walls of the blood-vessels and scattered throughout the section.

Other blood-vessels showed exceedingly thin walls, while yet others seemed to have no proper walls, but communicated directly with the cells of the tumor.

The enormous number of blood-vessels within the growth, with their peculiar structure, accounted for the occurrence of the blood-pigment and degenerated red corpuscles, as well as for the variety of colors presented clinically by the tumors.

The pigmentation of the tumors is not to be ascribed to the presence of pigment granules within the cells, as in the true melanotic sarcomata, which originate from pigment moles or the choroid coat of the eye, but is due altogether to the hemorrhages mentioned.

The result of my examination accords closely with that of Kalindero and of Babes,¹ of a similar case in which the authors were disposed to look upon the growths as vascular in origin.

The same view is upheld by Babes² in an elaborate article upon the development of the sarcomatous affections.

The almost simultaneous occurrence of the tumors on the four extremities, their independence of a primary growth, and their close resemblance to other infectious diseases, as leprosy and syphilis, have suggested to more than one observer that they might owe their development to some infection which had gained entrance to the general circulation.

Pringle³ in the discussion of the subject before the *Congrès International de Dermatologie et de Syphilographie*, held at Paris in 1889, announced that in two cases of the affection he had found bacilli which were situated in the capillaries, and in one of the cases the bacilli were found in great numbers in the sweat-glands and their ducts. Stimu-

¹ Sarcome cutané pigmentaire multiple idiopathique, avec début par les extrémités.

² "Handbook of Skin Diseases." Edited by Ziemssen. Am. trans. New York: Wm. Wood & Co.

³ Comptes Rendus. Paris: G. Masson, 1890.

lated by this announcement by Pringle, I stained a number of sections after the method of Gram and in various ways with the aniline dyes, but without finding micro-organisms of any kind.

The clinical history and microscopic examination in my case showed it to be a sarcomatous affection and to belong to the special variety which was first described as a distinct affection by Kaposi,⁴ under the name of multiple pigmented sarcoma of the skin. He gave the histories of five cases in his original communication, all of which proved fatal, and in two of them a microscopic examination showed the tumors to be small round-celled sarcomata. An autopsy in one of the fatal cases revealed metastatic tumors in the internal organs. Treatment was without avail in these cases, and the writer states that a fatal termination is inevitable.

In a later work⁵ Kaposi states that he has seen in all twelve cases of this affection, and that internal medication or extirpation of the tumors have proven unavailing in checking the fatal course of the malady.

Following Kaposi's description, other cases of this affection have been described by Vidal,⁶ Wigglesworth,⁷ Tanturn,⁸ Taylor,⁹ Amicis,¹⁰ Köbner,¹¹ Hardaway,¹² Hallepeau,¹³ Funk,¹⁴ Schwimmer,¹⁵ Köbner.¹⁶

The subject of sarcomatous affections in general has been fully and ably considered in a recent work by Perrin,¹⁷ which treats of the various forms of the disease, their differential diagnosis, and relationship to allied affections.

The minute structure and clinical course of this affection would certainly ally it to the sarcomata, but it must be looked upon as an affection having marked characteristics of its own, among which may be mentioned the simultaneous appearance of symmetrical tumors on the extremities, apparently independent of a primary growth, the slight tendency of the growths to soften, and the slow course of the malady.

⁴ "Diseases of the Skin." Hebra and Kaposi. New Sydenham Society translation. London, 1875.

⁵ "Pathologie und Therapie der Hautkrankheiten." Dritte Auflage. Wien, 1887.

⁶ Soc. de Biologie, 1875.

⁷ Arch. of Dermatology. New York, 1876.

⁸ Il Morgagni. 1877.

⁹ Arch. of Dermatology. New York, 1875.

¹⁰ Il Morgagni. Napoli, 1882.

¹¹ Köbner, Berliner klin. Wochschr., No. 2, 1883.

¹² JOURNAL OF CUTANEOUS AND VENEREAL DISEASES. New York, 1884.

¹³ Revue des Sciences méd., 1885.

¹⁴ Monatsh. f. prakt. Dermat., 1889.

¹⁵ "International Atlas of Rare Skin Diseases," II. Hamburg and Leipzig: Leopold Voss, 1889.

¹⁶ Demonstration of a Case of Idiopathic Multiple Pigmented Sarcoma of the Skin (Kaposi). International Medical Congress, Berlin, 1890.

¹⁷ "De la Sarcomatose Cutanée." Paris: G. Steinheil, 1886.

The affection described by various writers under the names of mycosis fungoides, mycosis fungoïde of Alibert, granuloma fungoides, and the inflammatory fungoid neoplasm of Duhring, regarding the nature of which so many divergent views have been expressed, has undoubted clinical relationship with the sarcomata, but can readily be differentiated from this affection by its primary stage of eczema and pruritus preceding the tumor development, the marked disposition of the growths to undergo softening and ulceration, and from their histological structure. The relationship of either mycosis fungoides or multiple pigmented sarcoma with a leucæmic affection of the lymphatic tissue of the skin, described by Kaposi as lymphoderma perniciosa, cannot be regarded as established, inasmuch as this latter affection was found by him in connection with an absolute increase of the white blood-corpuscles and with leucæmia of the spleen and bone marrow.

Prognosis.—The majority of the cases of this form of sarcoma of the skin have terminated fatally within from two to five years, but whether the disease is uniformly fatal, as considered by Kaposi, would hardly seem to be warranted after an examination of the cases recorded.

Hardaway,¹⁸ under the title of a case of pigmented neoplasm of the skin, reported the case of a patient who had at that time multiple tumors of the skin, proven, by a microscopic examination by Dr. Hertzmann, to be alveolar sarcomata, which had existed for ten years without materially influencing the health of the patient. In a further report of this case¹⁹ Hardaway says that after fifteen or sixteen years from the beginning of his disease he remains in good health, and that the sarcomatous tumors have undergone complete involution, leaving behind merely an atrophic condition of the skin.

Taylor²⁰ reported the cases of a colored woman, aged forty-eight, who had multiple pigmented tumors over the abdomen, extremities, palms, soles, and scapular region, in whom the first growth had appeared twenty-four years before over the sternum. The patient's health was good. No opportunity was afforded for microscopic examination, but clinically the tumors were regarded as corresponding to those in Kaposi's disease.

Köbner²¹ reported a case of the disease under consideration in a girl aged eight years, in whom several hundred tumors were scattered over the extremities and trunk. After the hypodermic use of Fowler's solution for a period of several months, all the tumors under-

¹⁸ JOURNAL OF CUTANEOUS AND VENEREAL DISEASES, January, 1883.

¹⁹ JOURNAL OF CUTANEOUS AND GENITO-URINARY DISEASES, Jan., 1890.

²⁰ Loc. cit.

²¹ Loc. cit.

went complete involution, leaving behind depressed pigmented scars, which afterward became white.

An examination proved the disease to be a spindle-celled sarcoma.

At the end of five years Köbner²² showed the case before the Berlin Medical Society, she having remained entirely free from the affection during this time.

While it would appear from the cases just quoted that an absolutely unfavorable prognosis need not be given, it should be remembered that Kaposi, who has seen more cases of this affection than any other observer, has been unsuccessful in obtaining results from the use of arsenic. The true nature of the entire group of diseases classed as sarcomata is so little understood that it would not be surprising if certain cases, which clinically presented similar or identical features, should prove to originate from independent causes.

The patient whose case is described in this paper has been under my close observation for a period of four months, during which time I have given him arsenic both hypodermically and internally, without noting any special action of the drug on the disease. No additional tumors have developed, and the patient's general condition has remained unchanged.

66 PARK AVENUE.

²² Berl. klin. Wochenschrift, No. 12, 1886.

