

SARCOMATOSIS CUTIS.*

DAVID LIEBERTHAL, M.D.

Professor of Skin and Venereal Diseases, Illinois Medical College;
 Attending Dermatologist Michael Reese Hospital and United
 Hebrew Charities' Dispensaries.

CHICAGO.

It would far exceed the time allotted should we attempt to consider the class of true sarcomata occurring within the skin, or all those forms belonging to the class of sarcoïd tumors, as mycosis fungoids, lymphodermia perniciosa and leucemia cutis. We will limit ourselves to the discussion of the idiopathic multiple pigmentary or hemorrhagic sarcoma (Kaposi) and the so-called sarcomatosis cutis (Kaposi) and their relation to each other, after citing cases of the first type from our own observation, which, differing somewhat from the usual type, offers some interesting points which tend to show that in the two affections we probably have to deal with an identical process.

A. IDIOPATHIC MULTIPLE HEMORRHAGIC SARCOMA.

CASE 1.—C. L., German, 40 years old, married, has three healthy children. His parents died well advanced in years from causes unknown to him. He never suffered from any illness nor had venereal disease. His present affliction dates from the summer of 1896, when he bruised his left leg on stepping off a street car. Soon afterward his feet began to swell and to become very painful; after a month or two there appeared dark spots and lumps on his feet. In the course of time these increased in number and spread up the legs, while spots developed on the arms and lastly in the groins and armpits. Pain in the feet continued for about two years, especially when he attempted to stand up, so that he was confined to bed, and after that time it subsided so as to be felt in cold weather and then in the lumps only. Some of these lumps disappeared, while others developed.

Examination.—I saw the patient first in October, 1899. He was tall and strong. The mucous membranes were well injected, the nervous and vascular systems, the various internal organs, the blood and the urine were normal, the integument was affected on the extremities only, the feet and limbs were thickened nearly up to the knees, the toes were broadened and clumsy and at the interdigital folds were densely arranged filamentous, horny outgrowths, and on the dorsum of the toes, pea to bean-sized, bluish-red, smooth and moderately firm nodules. On the inner side of the sole of the right foot were three plaques of the size of a quarter, arranged behind each other. The middle one was slightly raised and of a bright red color, which disappeared on pressure. It was beset with horny, thorn-like excrescences. The two others were of a dirty grayish-brown color and were saucer-shaped, smooth, hard and horn-like. Around the malleoli were similar ones. On the dorsa of the feet and over the tibiae were various round or oval nodules and nodes from the size of a pea to that of a nickel, some bluish red, firm, smooth and round, and others dark blue, soft and compressible, also dark brown, slightly depressed spots and blue ones level with the skin. On the outer aspect of the right tibia in its upper half was a group of about forty dark-bluish pea to bean-sized nodules closely arranged, and bearing on their surface lamellous, easily removable scales. The nodules were compressible. There were two groups of nodules of precisely the same character on the inner aspect of the left tibia. On the inner aspects of the thighs and in the axillae were bluish-red streak-like spots arranged parallel to Poupart's ligament and to the axillary arch respectively. They were perfectly smooth, not elevated, but the skin beneath was densely infiltrated and scarcely movable above the subcutis. This infiltration extended throughout the affected area and beyond the spots. On the right external epicondyle of the humerus was a bluish-red spot of the size of a dime, smooth, not elevated nor the skin beneath infiltrated and its color did

not change on pressure. There was a spot of the same nature on the ulnar side of the left forearm.

Subsequent History.—The patient was soon lost sight of until January, 1902, when he reported that he had suffered from phlegmon on the left leg in the fall of 1899, and from erysipelas of the face in the spring of 1900. Of the lumps some had disappeared without leaving any trace, while new ones had developed. His health during all this time had continued good. The examination of the patient resulted in the same favorable general findings as at the first presentation, but the local symptoms of the skin had somewhat changed. The pictures which I show you were taken at this time. The spots on the arms had increased in size, were darker and slightly elevated, but still movable with the skin. There was also found one red-blue spot of the size of a bean above the one previously described on the epicondyle. It was not infiltrated.

The thickened feet and legs presented the following appearance: Of the three discs on the inner side of the right sole, the middle one lacked the spinous processes in its periphery and was dirty yellowish brown, cornified and of saucer-like appearance, like the other two.

The plaques on the outer aspect of the right tibia which consisted, at the previous inspection, of numerous blue compress-

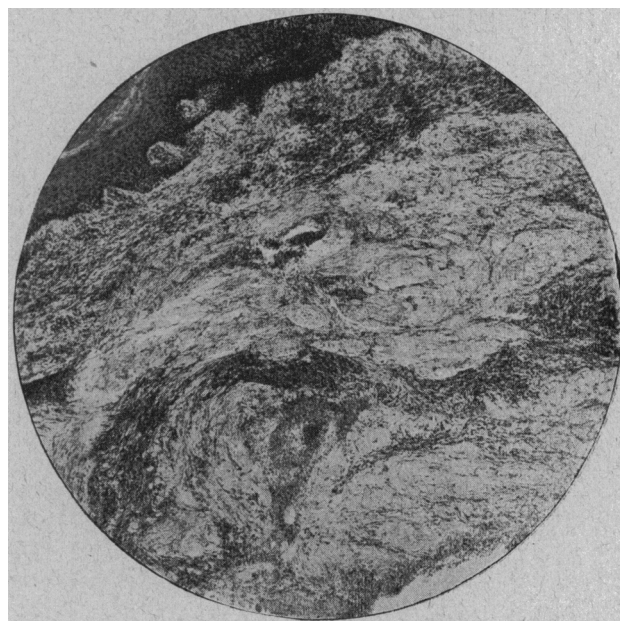


Fig. 1.—Case 1.

sible nodules now present a brown and hardened area, flattened in the center, bearing a horny knob-like outgrowth and lamellous yellow-brown scales. The two plaques on the inner aspect of the left tibia had changed in their centers to corneous cauliflower-like formations with dirty yellowish scales on their surface. In the periphery of these plaques are two zones, the inner consisting partly of flat, hard nodules and partly of compressible tumors; the outer, of depressed pinkish skin surrounded by a dark brown line level with the surrounding skin. Besides there were found over the tibiae brown atrophic spots, flat and dense nodules and compressible ones, and also areas of the extent of a dime to a nickel of normally appearing skin, but which on touch revealed dense infiltration.

Over the posterior aspects of the femurs were a number of brown, irregularly outlined, smooth, non-elevated spots of the size of a bean to a half dollar, the skin beneath some of these and somewhat beyond being densely infiltrated. Below the right scapula is a reddish-blue spot of bean size, the last one to appear until January, 1902.

In the groins and in the axillae the streak-like spots became transformed into chocolate-brown, flat, firm nodules of the size and shape of a bean; with the underlying infiltration remaining.

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About May 10 of this year the right sole became more swollen and very painful. I saw him a week later and discerned on the diffusely swollen and bluish sole in the metatarsal region five violet nodules from the size of a pin-head to a pea, very painful on pressure, not raised, but distinctly palpable as spherical bodies. During the past year he had felt pain in the older lesions during cold weather only.

Microscopic Examination.—A bean-sized, moderately flat nodule was excised from the leg for histologic examination, the wound healing promptly. Histologic findings as follows: Stratum corneum, lucidum and granulosum well developed, stratum Malpighii somewhat narrowed. The papillary layer is obliterated, especially in the central part of the nodule. The corium contains an abundance of coarse, fibrous connective tissue and within this thin and coarse bundle-like masses of medium-sized or small fusiform cells, especially in the upper strata. These bundles cross each other in various directions.

On the other hand, there are found numerous aggregations of the same cells with scanty fibrous tissue, and more so in the deeper strata. Throughout the corium are numerous lymph-clefts.

Of the blood vessels, which are very numerous within the cell accumulations, some show thickened walls, which thicken-

urination and frequent and painful evacuation of his bladder. This condition became more aggravated until I first saw him in August, 1898.

Examination.—He was short, very emaciated and of sallow complexion, the mucous membranes pale, the lungs slightly emphysematous, the heart hypertrophic, the spleen normal, the prostate gland hypertrophied. There was retention of urine, chronic interstitial nephritis, sclerotic arteries and slightly elevated temperature. The blood showed hemoglobin 65 per cent., reds normal, whites slightly increased in number. The integument was normal except on the penis and lower extremities. On the skin of the under surface of the penis, just behind the glans, was a bean-sized, bluish-red, roundly elevated, firm nodule, movable with the skin and not painful. On the legs, and especially numerous over the tibiae, were nodules and tumors of sizes from a pea to that of a hazelnut, bluish or brownish, more prominent or flat, solitary and in groups, some soft and compressible with scales on their surface and others firm, while between these were blue and brown spots of various sizes. These were not elevated, but the skin beneath was infiltrated. Some of the brown spots were atrophic.

On the dorsa of the enlarged and clumsy feet were flat, dense, brown nodules of the size of a split pea to a bean. The inter-

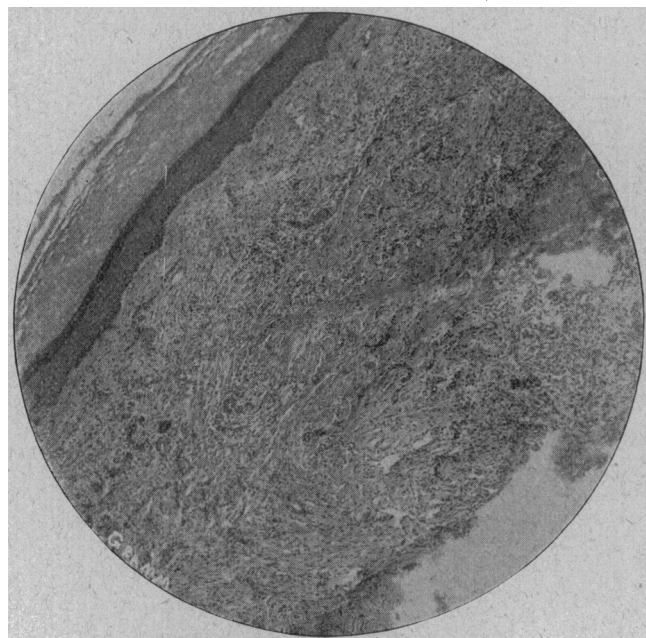


Fig. 2.—Case 2. Smaller nodule.

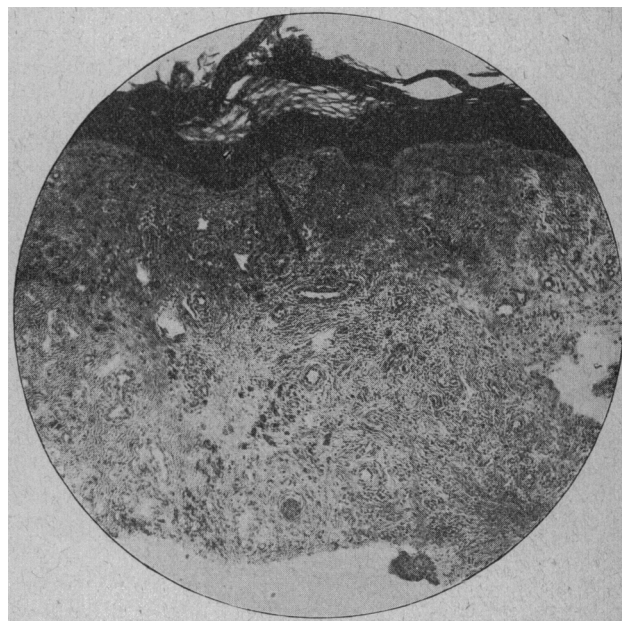


Fig. 3.—Case 2. Larger nodule.

ing is partly due to infiltration of the adventitia to which frequently are attached the streaks of cells and partly to a swelling and proliferation of the endothelium, while other blood vessels have a thinner wall looking as if ruptured. Some are densely filled with red corpuscles which may also be seen extravascularly.

Large masses of a yellowish-brown pigment are seen among the tissues, especially within the cell accumulations throughout the corium, besides pigment granules intra- and extra-cellularly, all this pigment giving the test of hemosiderin. Elastic fibers stained with Weigert's fuchsin-resorcin were found to be normal throughout the sections. Round cells are found here and there in small numbers intermingled with the others, while they are quite numerous around the sudoriferous and sebaceous glands and follicle sheets.

CASE 2.—H. G. Nothing in his family history has any bearing on his skin affections. He had no venereal disease. Ten years before I saw him his feet swelled and became painful, especially when walking. The swelling subsided to a certain degree, but after a time there appeared dark lumps on his feet and gradually ascending also on the legs, and at last one came on the penis. From time to time certain ones disappeared and others occurred. These lumps were painful on pressure. He complained of nothing else until five years after the skin first became affected, when he experienced difficulty in

digital folds of the toes were densely beset with rod-like, horny outgrowths like a brush. The inner side of the soles is the seat of dirty yellowish flat tumors, hard and cornified, elevated two lines above the level of the skin, with depressed center and smooth surface and of the size of a nickel to a quarter.

There was a similar one, bean-sized, anterior to the right internal malleolus and two, of the size of a quarter, below the left outer malleolus. There was no enlargement of the lymphatic glands. The patient died from uremia two months after I first saw him.

Microscopic Examination.—Two nodules from the leg, one pea-sized, of recent development and one bean-sized, of quite long standing, both moderately firm, were excised and prepared for examination.

1. In the sections of the smaller nodule: Stratum corneum slightly thickened, stratum granulosum narrow, as is also the stratum Malpighii. The papillary layer has vanished, the subpapillary layer contains wide lymph spaces, as does also the reticular layer of the corium. Throughout the corium, between sclerotic or normal connective tissue, there are round cells intermingled with sparse fusiform ones, arranged mostly in irregular streaks, and here and there in irregular aggregations. Around the glands is abundant round-cell infiltration. In many places the cell bundles are attached to the infiltrated adventitia of blood vessels. These are partly thickened, partly dilated.

There are no hemorrhages nor are there found masses of pigment.

2. Sections of the larger nodule show nearly the same conditions as those of the smaller, but besides there are found blood vessels with very thin walls, and densely packed with red corpuscles. Other free ones are found outside the vessels. There are also numerous masses of yellow-brown pigment and intra- and extra-cellular pigment granules. The cells, of which the round predominate, were arranged in a similar manner to those of 1.

COMPARISON OF CASES.

Comparing the anatomic findings in the two cases we observe in the first a prevalence of spindle cells, in the second of round cells. Both have a hazy, coarse, granular nucleus and a narrow zone of protoplasm. In the upper strata of the corium, in both cases, the bundle-like arrangement of the cells is more pronounced. While we find in the first case blood vessels with very thin walls and in places appearing as if ruptured, and numerous masses of pigment, and the same two conditions in the older nodule of the second case, these are wanting in the sections of the small recent nodule of the second case. In neither case was exulceration of the nodules observed.

Considering now the clinical as well as the microscopical findings of both cases, the diagnosis of *sarcoma multiplex hemorrhagicum idiopathicum* is justified.

OPINIONS OF WRITERS.

Since Kaposi¹ first reported these cases and applied to them the name *sarcoma pigmentosum*, the literature of the subject has increased considerably. A very animated discussion arose in regard to the name, inasmuch as this affection in some respects does not correspond to the requirements of a true sarcoma, that is to say, the tumors have a limited growth and may heal spontaneously.

On account of these and from differences afforded by a study of the histologic findings, Kundrat and Paltauf were of the opinion that these cases should be accredited to the group *granuloma*, which view is shared by other observers. Selli² goes even so far as to propose for them the name *granuloma multiplex hemorrhagicum*. As if to do justice to both sides of the controversy Kaposi subsequently classed his pigmentary or hemorrhagic sarcoma in the group of *sarcoid tumors*.

The etiology of this disease is unknown. As early as 1882 de Amicis³ expressed the opinion that an infectious agent might play a rôle in the development of these cases. Other authors inclined to the same view, but hitherto no proof has been afforded by bacteriologic examination of the diseased areas or by inoculation. Another suggestion as to the etiology has been offered by Campana in his discourse,¹¹ in which he records a case showing neoplastic infiltration of the peripheral nerves in diseased areas, and therefrom drew the conclusion that the changes in the skin are trophic disturbances.

Semenow⁴ arrived at the same conclusion from the findings in one of his cases. There was accumulation of pigment in the spinal ganglia, swelling of the nuclei in the neuroglia and proliferation of the perineurium of peripheral nerves. He assumed to be supported in his view by the symmetrical arrangement of the lesions and the severe pain frequently occurring within them. However, it is well to question this theory on the ground that the skin is affected by the same cause and at the same time as the nervous system, or the latter is affected secondarily.

It seems plausible that this process represents a slowly growing sarcoma which may change secondarily and be-

come malignant and that the question of its etiology will not be solved before that of typical sarcoma becomes known. To speak with Perrin:¹⁰ "We are scarcely in the beginning of our knowledge of this affection." Therefore, we should rather accept the interpretation and the name which the genial diagnostician, Kaposi, has offered and so splendidly defended⁵ (1894), and not welcome other names or explanations unless they can be better substantiated.

This affection occurs mostly in males over forty years of age, but has been observed in a patient as young as five years by de Amicis.⁶ The diagnosis of idiopathic multiple hemorrhagic sarcoma offers as slight difficulty as its differentiation from other similar affections. The prognosis is unfavorable, although the administration of arsenic improves some cases temporarily, while there are even cases on record⁶ where permanent cure has been effected by the hypodermic administration of arsenic. It is of interest just here to refer to a case of Semenow's,⁴ in which an erysipelas migrans caused its cure.

B. SO-CALLED "SARCOMATOSIS CUTIS."

Let us now consider Kaposi's so-called *sarcomatosis cutis*. Kaposi describes it as follows: "On the trunk



Fig. 4.—Case 1.

and extremities there appear numerous bluish, flat or more prominent spots, on the palpation of which there are felt in the depth of the skin firm, elastic, flat roundish plaques, not sharply defined. Besides these, firm plaques are felt over which the skin is neither protruding nor discolored. In one case the plaques were painful; in another on pressure only, and in a third case there were found bluish-red prominent, smooth and firm nodules of sizes from a pea to a hazelnut, besides larger flat plaques with central depression. There were no enlarged lymph glands, nor any alteration in the blood. These cases are curable by arsenic."

The histologic findings are, in Paltauf's opinion, somewhat similar to round-celled sarcoma. Cases of this type were further recorded by Joseph,⁷ Spiegler,⁸ Tandler⁹ and others, but their number remains in the minority in comparison with the numerous recorded cases of hemorrhagic sarcoma.

The histology of different cases differs somewhat, although in most of them similar conditions are encount-

ered, as in the hemorrhagic variety. The horny layer is broadened, the stratum Malpighii nearly normal, the papillary in some places vanished. The subpapillary layer shows sclerotic connective tissue and numerous wide lymph clefts. In the deeper strata of the corium, the real seat of the affection, between scanty, or in places sclerotic, tissue bundles there are embedded irregular masses of round cells (in Joseph's⁷ case fusiform cells arranged in rows, infiltration-like), or such cells arranged in bundles or streak-like.

Within the areas of cell accumulation are wide lymph clefts and numerous blood vessels, the endothelium of some of these latter swollen and in proliferation. The elastic fibers are preserved throughout.

COMPARISON OF TYPES A. AND B.

Although he insists upon strict division between the hemorrhagic sarcoma and the so-called sarcomatosis cutis, Kaposi admits that there might be transitional forms between the two, as between all other forms

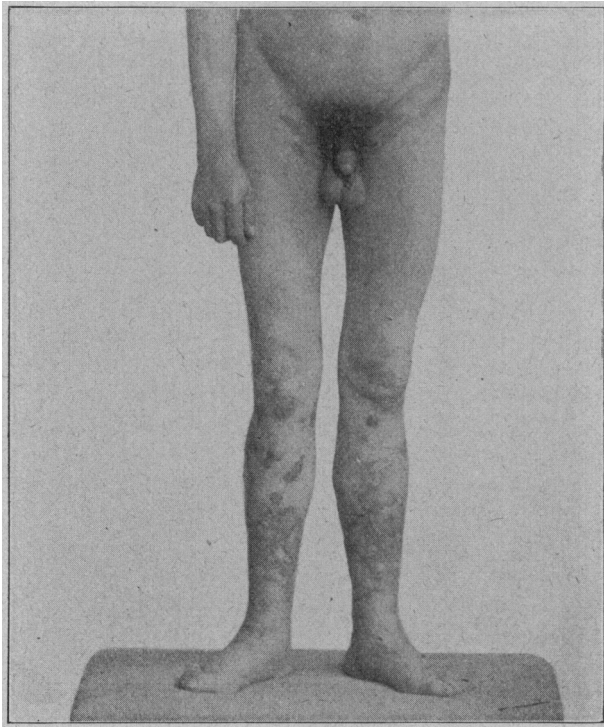


Fig. 5.—Case 1.

classified by him as sarcoid tumors. But we may go further and assume that this division can not be strict, but that both forms are the same process, with some modifications.

This view is supported by a study of our own cases. The character and arrangement of cells are in both types nearly the same. While it is assumed that the cells are arranged more in irregularly grouped masses throughout the whole corium in the hemorrhagic, and more especially in the deeper strata, and band-like in the form sarcomatosis cutis, yet we really find cases of hemorrhagic sarcoma, as our second one, where the upper part of the corium is only slightly infiltrated. In both of our cases the band- or streak-like arrangement of the cells is prevalent throughout. It is further claimed (Joseph l.c.) that the elastic fibers are wanting throughout the hemorrhage node, while they are preserved in that of sarcomatosis. The fact is that they are unaltered in hemorrhagic cases, too, as is shown in our first case. Let us inquire in regard to the hemorrhages and

masses of pigment. We should find them in all cases of hemorrhagic sarcoma, but not in the other form. As above recorded, there were found no strong indications of such in the more recent and small nodule of our second case, while the older and larger nodule showed an abundance of them. From this we may infer that the hemorrhages and pigmentation are of secondary occurrence. The papillae are in a similar condition in the two forms, and the same is true of the lymph spaces. In our second case the latter were numerous throughout the sections; in the first they were found in the deeper strata of the corium only.

There is as much similarity in the clinical conditions as there is in the histologic. Concerning the clinical symptoms, we may state that in our first case there were found bluish-red spots in non-infiltrated skin, and bluish-red, rounded and firm, flat, or prominent nodules, as well as compressible ones, solitary or grouped, typical of the hemorrhagic form.

On the other hand, there were found characteristics of sarcomatosis cutis, such as bluish or brownish spots on densely infiltrated bases, as those in the axillae and inner and posterior aspects of the thighs, which infiltration reached into the subcutis and extended beyond the spots in the periphery, also irregular and dense infiltrated areas over which the skin looked perfectly normal.

Blood and lymphatic glands are found to be normal in both varieties. In the hemorrhagic form the skin becomes affected first on the distal ends of the extremities, but there are cases on record where another region took the lead. In the sarcomatosis cutis the skin lesions occur irregularly on the body, but they ultimately affect the extremities. The prognosis is favorable in the sarcomatosis, the cases yielding readily to arsenic, while in the hemorrhagic variety a few cases only are recorded as cured,⁸ temporary improvement only being the rule. There seems to be no doubt that the prognosis is rendered less favorable in the latter type by the occurrence of hemorrhages.

CONCLUSIONS.

The result of all the foregoing comparisons is that there is no great distinction between the two forms. It rather impresses one that they represent the same process. While some of the cases do not change secondarily, and are, therefore, more amenable to treatment and cure, the majority are subject to secondary changes, rendering the course unfavorable.

103 State Street.

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DISCUSSION.

DR. W. S. GOTTHEIL, New York—I have had a number of similar but more extensive cases under observation for a long time. The rather careful clinical and microscopic study to which I have subjected them would lead me to conclude that there is not so much difference between them as the elaborate nomenclature of their subvarieties would imply. It is a question whether classification is not rendered more difficult by the multiplication of Latin terms, of which our German colleagues

are so fond. Synthesis rather than analysis is what is now required, lest we lose our way in the terminologic forest. For ordinary purposes I recognize three varieties of sarcoma of the skin. The first is that slightly malignant, superficial growth called sarcoid by Boeck. The second is the localized sarcoma, either primary in the skin or secondary to sarcoma of other organs. The third includes the idiopathic, diffuse sarcomas, pigmented or white. In the present state of our knowledge of the subject I do not believe that variations in color, course, etc., justify us in subdividing maladies which are microscopically almost identical.

DR. M. B. HARTZELL, Philadelphia—I have just one word to say of the character of the so-called sarcoma cutis. Personally I have doubted whether all of these cases have a right to be called sarcoma; if so, then they differ from the surgeons' sarcoma. What the surgeons class as sarcoma is rapidly destructive and rapidly terminates in the death of the patient, but the so-called sarcoma of the skin frequently lasts for years. As has already been said by the previous speaker, the confusion in the use of terms is already great enough without adding new names. We ought to be slow to believe that all of the so-called sarcoma cutis is actually sarcoma. I recall a case that was exhibited at a clinical meeting of the American Dermatological Association as sarcoma in which there was simply extensive slate-colored discoloration of the skin extending to the sclera. We call many of these cases sarcoma because microscopically they look like sarcomatous tissue, but they are probably often something else.

DR. LIEBERTHAL—In reply to Dr. Gottheil I wish to say that in my paper there was neither an attempt made to multiply terms nor to introduce new ones. The long Latin name did not originate with me, but with Kaposi, and has since been accepted by the dermatologic world. Therefore, I have no apology to offer for repeating it before a body of dermatologists most of whom have a national as well as an international reputation and are familiar with it and its interpretation. As long as there is no better name suggested and accepted we do well to let the old one stand in order to avoid confusion.

Regarding the remark made by Dr. Hartzell, I would refer to the essay mentioned in my paper in which Kaposi defends his standpoint concerning the name applied by him to the first form. I did not attempt to solve the question of the place to be assigned to it in pathology, but only entered into the consideration of the similarities between the two forms.

Clinical Report.

SEVERE BRAIN INJURY WITHOUT FOCAL SYMPTOMS.

WILLIAM A. DAVISON, M.D.
BRIDGEPORT, CAL.

The Injury.—July 8, 1902, at 7 p. m., E. L., male, aged 27, had an iron bolt, with a one-inch square top on its end, driven by direct violence through the vault of the cranium. The bolt cut through his hat and scalp, crushing in at least one square inch of the frontal and nearly as much of the left parietal bones, carrying the fragments deep into the brain substance. A triangular piece of left parietal bone, one inch wide at its base and one and one-half inches long, was broken down and rested on the brain. The front edge of the wound was one and one-half inches in front of the coronal suture, a little to the left of the median line of the frontal bone, extending across the median line probably one-half inch, thence backward across the coronal suture into the left parietal about one inch. It thus made an oblong opening about one and one-half by two inches, with jagged edges.

The brain injury involved the superior portion of the first, second and fourth frontal convolutions, the ascending parietal, the superior parietal and probably the superior sylvian convolutions of the left side, the superior portions of the first frontal on both right and left sides, the superior portion of the ascending, and the anterior portion of the superior parietal lobes of both sides.

Treatment.—The course of the wound was slightly to the left. How deep the bolt penetrated I did not have the necessary curiosity to ascertain, but was satisfied to calculate by the size of the opening that there must be buried in the cerebrum the fragments of a section of bone measuring one and one-half by two inches. The conclusion was verified later. I irrigated and removed all visible bone fragments (save that of the left parietal) and such invisible pieces as were easily detected by gentle application of the probe. On removal of the pieces of bone, brain matter and blood clots came away. The triangular piece of the left parietal was elevated and left *in situ*. The bolt had not remained in the wound.

Receipt of the Injury.—No one witnessed the accident and the patient could not tell how it happened further than that he was leading a saddled horse under a wire clothesline attached at each end to a post—at one end by this iron bolt, which inflicted the blow. The clothesline caught under the pommel of the saddle. The post with the protruding bolt was broken off at the ground and hurled some feet away. The man was thrown to the ground, from which he was seen arising by his wife, who at this juncture happened on the scene. With her assistance he walked fifty feet to his bed.

Symptoms.—Hemorrhage was free but not excessive. There was some shock and for a time semi-consciousness; no loss of

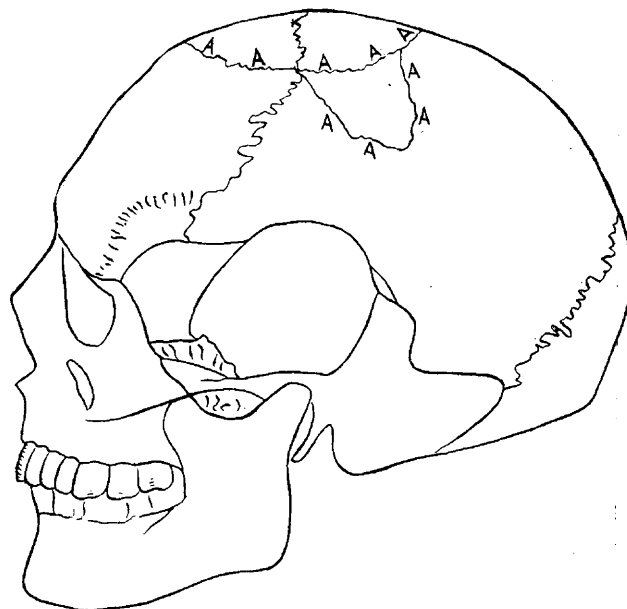


Diagram of injury. The lines marked A—A represent the skull fractures.

any of the senses. There was an indisposition to talk, but at all times he would speak if urged. There was also acute, lancinating pain in the front of the head, scanty secretion of urine and little desire to pass water for a day or two. I could not say there was paralysis, for the fluid being scanty and high-colored, I did not allow him to retain it longer than eight hours without use of catheter. The bowels were constipated, but yielded readily to purgatives. I did not consider the bladder or rectal torpor more than one often finds with active men confined suddenly to bed. There was no motor nor sensory paralysis at any time, in spite of the fact that he lost at least a tablespoonful of brain substance from the motor area. From beginning to recovery the temperature never exceeded 99.6. The pulse varied from 40 to 70 for the first ten days and was very weak. This was combated with 1/50 gr. sulph. strychnia hypodermically.

After about ten days the pulse rallied and gradually resumed its normal force and activity. There was no change in the pulse beyond its slow and weak action. The pupils were not markedly affected at any time, but for a day or two were probably not so sensitive to light as in the normal state.

After-Treatment.—For several days small pieces of bone presented themselves and were removed. Irrigation of hot borated water, borated gauze dressings and sulph. strychnia as indi-