A CASE OF MULTIPLE IDIOPATHIC HEMORRHAGIC SARCOMA (KAPOSI)

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REPORT OF CASE

History.—The subject of this report first came under observation in January, 1919. He was an Austrian Jew, 48 years of age. He complained of pain and swelling in the legs and arms and a cutaneous disorder of about fifteen months' duration. The lesions appeared first on the hands, later on the legs. The toes, ears and body finally became affected. The lesions were accompanied by swelling of the hands and feet. At the time of the first examination the lesions were large and numerous on both hands and feet. Scattered lesions were present on the arms, legs and trunk. There was one lesion on the right eyebrow and one on each ear. The patient stated that the lesions began as hard, flat, elevated red areas about the size of peas. They gradually grew larger and those on the hands and feet in some instances coalesced to form plaques. Previous to the onset of the skin manifestation he noticed itching of the ears and back. This was not present at the time of examination, there being no subjective sensations except pain in the swollen lower extremities. For about six months he had noticed an enlargement on the left side of the chest which had slowly increased in size to about that of a hen's egg. It was not painful nor in any way disturbing except on account of its size and location. The patient had always been well except for an attack of pneumonia some years ago. He had had no previous skin disorder nor was there anything in his family history bearing on his present condition.

Physical Examination.—The patient was a well developed, rather stout, middle aged man. His general condition was good. Scattered diffusely over the body, being more profuse, however, on the hands, feet and lower portion of the back, were dull red to brownish, infiltrated, flat nodules from the size of a pea to that of a quarter dollar. The smaller lesions were more erythematous than the larger ones and those on the extremities were distinctly purplish. Many of the larger lesions were less infiltrated, and there was less discoloration and infiltration in the center of the plaques. On the hands and feet the lesions had become confluent, forming large, purplish, infiltrated plaques; on each ear, involving chiefly the central portion, was a purplish, infiltrated plaque gradually fading at the border into the normal skin. The lesions on the body elsewhere were sharply demarcated from the surrounding normal integument. The nodules were all fixed to the skin and apparently extended into the deeper layers of the corium. In the left anterior axillary line under the lateral border of the pectoralis major muscle was an egg-sized, hard mass which was freely movable and apparently not fixed to the surrounding structures. In the supraclavicular space on each side was a similar but smaller nodule. There was considerable brawny edema of the legs, extending almost as high as the knees, and of the hands and lower forearms. There were no lesions in the mouth or pharynx. No changes in the viscera were discernible by ordinary examination. The urine was negative, and there were no changes in the blood. There was 80 per cent. of hemoglobin. Leukocytes and erythrocytes

Fig. 1.—Full length view of the patient showing general distribution of the lesions.

Fig. 2.—Types of lesions on the legs when the patient first came under observation.

were present in normal numbers, and a differential count of the leukocytes revealed no disturbance in the proportions of the various elements. A biopsy was performed August, 1919, a lesion on the inside of the right thigh being removed at that time. The second biopsy was performed August, 1920, a lesion over the sacrum being removed.

The treatment consisted of arsenic used internally and roentgen-ray therapy. There was considerable improvement, especially after radiotherapy was begun. The lesions, especially those on the hands, became much less infiltrated. The edema and pain in the legs decreased greatly. The lesions on the ankles and on the calves and sides of the legs became cystic. This change began to occur

Fig. 3.—Lower portion of the leg showing beginning cystic change.

in July, 1920, when a few cysts the size of a split pea appeared in the regressing lesions. These cysts increased in number and size until at the time of writing they were scattered over the lower two thirds of the leg below the knee and ranged in size from that of a pea to that of a small cherry. This change was probably due to static conditions. There was no pain except in the right great toe on which there was a vascular sessile tumor, the size of a pea, which was surrounded and covered by a warty keratosis. This lesion improved considerably under radiotherapy.

Histopathology.—The tissues removed at biopsy were fixed in Zenker's fluid and stained in several different ways. In general, there was a slight atrophy of the stratum corneum, and the papillary processes were slightly shortened. There was slight edema. In the corium groups of cells appeared. In most of the sections this cell infiltration lay deep, but in a few places there was a rather diffuse cellular growth in the superficial portions. The cellular infiltrations in some places were around hair follicles and coil glands, and in practically all instances there was an increase of the vascular elements in the cell mass. Except in the portions where the growth was in the superficial layers, it was well demarcated from the surrounding connective tissue, but there was no definite capsule. Collagen fibers did not penetrate the cell group.

Fig. 4.—Photomicrograph showing the general distribution of the cell infiltration; $\times 60$.

Along the course of the blood vessels elastic fibers were seen to pass into the cell mass. The cellular elements were of several types; round and spindle shaped cells predominated. In some parts, areas of a single type of cell were seen. In most places there was a mixture of round and spindle cells and cells with irregularly shaped nuclei. There were a few scattered mast and plasma cells. In the peripheral portions of the cell masses there were large deposits of yellowish brown pigment which was largely intracellular and granular. The granules were fairly uniform in size and were usually round. There was some extracellular pigment but not a large amount.

The vascular elements were proliferated, there being apparently many more vessels and capillaries than were seen in a normal tissue. The capillaries and lymph spaces were dilated. In practically all parts of the sections the vessels were empty. In a few places there was a slight proliferation of the endothelium, but in most parts there was apparently no change. No hemorrhages were to be seen in any part of the sections.

In sections from a second biopsy the general structure of the tissue was the same. The cellular elements were scant, however, and there was no pigment. The vascular proliferation and dilatation persisted.

Fig. 5.—Photomicrograph showing types of cells and vascular proliferation; $\times 325$.

This case corresponds clinically to the cases described first by Kaposi under the title of "Multiple Idiopathic Hemorrhagic Sarcoma." Since the publication of his report, numerous cases have been reported and various interpretations have been given to the histology. Some authors hold that it is a true sarcoma, while others incline to the view that it is primarily a granuloma and may later become neoplastic. Sellei¹ strongly advocates the view that it is a granuloma. Pollitzer² con-

1. Sellei: *Monatsh. f. prakt. Dermat.* **34**:497, 1902.

2. Pollitzer: *J. Cutan. Dis.* **27**:143, 1909.

siders it a true sarcoma. Sequeira believes it to be an inflammatory condition. Gilchrist and Ketron³ report two cases in which the histologic picture varies from that of an angioma to that of a type of tissue resembling a connective tissue new growth. Ewing⁴ states that it is "an infection granuloma of unknown origin which in its later stages in predisposed subjects and under suitable conditions may take on genuine neoplastic properties. . . . The entire vascular system seems to be overtaken by degeneration with overgrowth of vaso-for-

Fig. 6.—Photomicrograph showing types of cells and vascular proliferation, with a group of pigment cells in the center of the field; $\times 325$.

mative cells, leading in many instances to true tumor formation." Justus⁵ reports inoculating white mice with tissue from a case of multiple idiopathic hemorrhagic sarcoma (Kaposi) with production of systemic lesions. He carried the condition through five generations of mice.

It is difficult to draw conclusions from a single case, yet the diffuse

3. Gilchrist and Ketron: *J. Cutan. Dis.* **34**:429, 1916.

4. Ewing: *Neoplastic Diseases*, Philadelphia, W. B. Saunders Company, 1919.

5. Justus: *Arch. f. Dermat. u. Syph.* **99**:446, 1910.

nature of the growth with the multiplicity of cell types, and close relation of the cell masses to epithelial structures and blood vessels, tends to a view that the lesions in this case have been produced on an inflammatory basis. The cells themselves are, however, largely connective tissue types, and there is a practical absence of the cells usually found in active inflammatory processes.⁶

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6. In addition to the references already given, the following articles may be of interest:

Macleod: *Brit. J. Dermat.* **17**:173, 1905.

Mariani: *Arch. f. Dermat. u. Syph.* **98**:267, 1909.