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HISTORY OF SCIENCE

CONTRIBUTIONS OF MORIZ KAPOSÍ TO KNOWLEDGE OF CANCER AND PRECANCEROUS CON- DITIONS: SARCOMA AND XERODERMA PIGMENTOSUM

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Moriz (Moritz, Moric, Moricz) Kaposi, whose original surname was Kohn, was born in Kaposvar, Hungary on October 23, 1837. Though his parents were poor, he completed his early education at Pressburg, Bratislava, and in 1861 received an M.D. degree at the University of Vienna. From 1866 to 1869 he was an assistant to Dr. Ferdinand Hebra, founder and chairman of the Department of Dermatology at the University of Vienna. He proceeded from "Privatdozent" to associate professor, and later assumed Hebra's chair until his own death on March 6, 1902. Kaposi was intimately involved in almost every clinical dermatologic advance of his time. He published widely; students and patients came from many countries to his clinic in Vienna. His trainees later occupied chairs of Dermatology at many other European universities. The first descriptions of several dermatologic conditions are attributed to Kaposi, two of which are pigmented sarcoma of the skin (Kaposi's sarcoma) in 1872 and xeroderma pigmentosum (XDP) in 1882. His descriptive and clinical studies of patients with XDP antedated the discovery of the recessive inheritance pattern and the fact that ultraviolet light sets up the mechanism for malignant melanoma of the skin. He did, indeed, recognize the seriousness of the condition and its precancerous nature.

† † †

INTRODUCTION

Moriz (Moritz, Moric, Moricz) Kaposi, whose original surname was Kohn, was born in Kaposvar, Hungary on October 23, 1837, the son of poor parents. His early education was obtained in Pressburg, Bratislava, and in 1861 he received the M.D. degree at the University of Vienna. It was there that Kaposi's interest in dermatology began as he worked as an assistant to Dr. Ferdinand Hebra who had founded and chaired the Department of Dermatology at that University. Kaposi soon became Privatdozent at the Allgemeines Krankenhaus after completing a dissertation on syphilis of the mucous membranes (Editorial, *JAMA*, 1964).

A short time later Kaposi married Dr. Hebra's daughter. Still later, he became associate professor of Dermatology; and after the death of Dr. Hebra in 1881, he became chairman of Dermatology at the University of Vienna, a post he retained until his own death on March 6, 1902.

Not only did Kaposi show excellence in his descriptions and knowledge of many dermatologic conditions, but he was knowledgeable in the descriptions of their histology and pathology (Braun, 1982). He published many dermatologic papers, and his lectures attracted students, patients, and physicians from many other countries to his clinic. Kaposi was an excellent speaker and was invited to speak at many international dermatologic meetings. Many of his students later became chairmen of Dermatology departments in a number of European countries.

Two of a number of dermatologic conditions first described by Kaposi are: idiopathic multiple pigmented sarcoma (Kaposi's sarcoma) in 1872, and xeroderma pigmentosum in 1882.

KAPOSÍ'S SARCOMA

In 1872, after having seen and attempted to treat several patients with an unusual type of sarcoma, Kaposi's description entitled "Idiopathic Multiple Pigmented Sarcoma of the Skin" (Kaposi, 1872) was published in the *Archives of Dermatology and Syphilology*. It was also published in his book, (Kaposi, 1880).

In about 1892, after further study, Kaposi suggested another name for this disease, idiopathic multiple hemorrhagic sarcoma, which he felt best described it. He described 16 cases of this pigmented sarcoma in men and one boy.

The lesions began on the soles and volar surfaces of the feet. Then the same surfaces of the hands became affected. He described the lesions as "reddish brown, later bluish red, round, moderately firm nodules from the size of a pea to that of a bean, which are in part separate and irregularly situated, in part arranged in groups and diffuse infiltrations varying from the size of a Kreuzer (a quarter) to that of the palm of the hand. The lesions then appear on the ankles and forearms, and in two to three years progress to the face and trunk of the body.

"Later, the lesions ulcerate and become gangrenous and necrotic." The prognosis was poor because no treatment was available. If the lesions were surgically removed, others always appeared. The most difficult symptom was the feeling of tightness and pain in hands and feet for which such palliative prescriptions as emollient ointments, cold compresses, and paregoric were given (Braun, 1982). In addition to the pain, the patients suffered from a high fever, bloody diarrhea, hemoptysis, and general debilitation before death occurred. The patient died within two to three years.

At autopsy, large numbers of lesions were also found in the esophagus, trachea, stomach, heart, liver, and intestinal tract with especially necrotic lesions in the descending colon (Editorial, JAMA, 1964). (Some of the lesions were described as the size of a silver groschen or a thaler.)

Kaposi removed a number of the sarcomatous lesions to study them microscopically. He described them as follows: "A round-cell sarcoma is seen, except that in a few places the characteristic spindle-cell sarcoma is seen. A peculiarity of this type is the presence of capillary hemorrhages, which explain the later bluish-black pigmentation of the originally bluish-red nodules, as well as the excessive hardness of the diffuse infiltrations around the groups of nodules caused by deposits of fibrin (Editorial, JAMA, 1964).

In recent years the following interesting findings concerning Kaposi's sarcoma are: 1) The ratio of male to female occurrence of the disease is 10:1; 2) there are racial, geographic and ethnic factors, with increased occurrences among Northern Italians, Ashkenazi Jews, and the Bantus of South Africa; and 3) there appear to be a number of familial occurrences so that an autosomal dominant gene is postulated (Lynch, 1972).

Treatment in 1986 for Kaposi's sarcoma

One wonders what Dr. Kaposi would think one hundred years after his first description of what is now called "classical Kaposi's sarcoma" (CKS) as distinguished from "epidemic Kaposi's sarcoma" (EKS), a frequent complication in patients with the Auto Immune Deficiency Syndrome (AIDS). Even before the appearance of AIDS, it was found that patients with kidney transplants who were chemically immunologically suppressed had not only impairment of the immune reactivity of the body's lymphocytes, but also many times developed Kaposi's sarcoma (Friedman-Kien and Laubenstein, 1984). This condition then regressed when the suppressive drugs were discontinued.

From learning that the body's immune mechanisms are suppressed in patients with AIDS and that EKS was one of the opportunistic conditions complicating the disease, new therapies for Kaposi's sarcoma became urgent. They now consist of radiation therapy for early lesions, chemotherapy, and biologic response modifiers, such as interferon.

XERODERMA PIGMENTOSUM or XERODERMA, PARCHMENT SKIN

Kaposi was the first dermatologist to report xeroderma pigmentosum (XDP) or xeroderma, parchment skin (Shelley and Crissey, 1953). In one of two young patients (age 18), he noted abnormality of pigmentation while in the second (age 10) there was none.

The first young lady, from one of the northern German states, had developed XDP in early childhood. Dr. Kaposi first examined her in 1863 with the following description: "The skin of the face, ears, throat, neck, shoulders, arms and of the breast to the level of the third rib . . . had a checkered appearance; it appeared to be abundantly dotted over with pigmented spots, the size of a pin-head or bean, and yellowish-brown in color; it was also tightly stretched, as if contracted, was pinched up into a fold with difficulty, and felt very thin. Its surface was smooth in some places, while in others, fine epidermic lamellae peeled off; or there were quite flat, linear furrows marked out on the epidermis, so that the surface appeared as dry as parchment, and wrinkled, while the skin, itself, was tightly stretched. In places, it was of a white color and was without pigment, while there also existed . . . yellowish or dark-brown pigmented spots resembling freckles . . . Beyond a sensation of tightness, the patient had no pain nor

itching. The skin of the whole trunk and of the extremities was quite normal, smooth, pliant, fine. The general state of health was normal” (Shelley and Crissey, 1953). The skin on this patient’s face had contracted so that her eyelids were drawn downward and she could not close her eyes, causing ulceration of the cornea. Her nose was compressed and her ears were indented in places, all due to the thinning and contraction of the skin. The patient died at age 27 from “cancer of the peritoneum” that presumably was not related to XPD, though changes in the patient’s immune system may have been a factor.

Since Kaposi’s time it has been learned, first, that exposure to sunlight (ultraviolet light) causes skin alteration in XDP, melanin deposits (freckles) and, in many cases, malignant melanoma of the skin. Also, it has been learned that XDP is an autosomal recessive disease. More recently, it has been learned that whereas in persons with normal skin, ultraviolet light causes a chain break in the DNA that can be “repaired” (Lynch, 1972), in persons with XPD such a “repair” mechanism is lacking due to an enzyme deficiency (endonuclease). When exposed to sunlight or ultraviolet light, the skin shows pathologic changes that include disturbances of pigmentation and maturation of epidermal cells (Lynch, 1972). Even more recently it has been found that the XPD gene is located on chromosome 1 (McKusick, 1986).

Effective treatment for XPD now consists of using chemical sun screens. At an XPD Clinic at the National Institute of Health, it has been found that accutane, a drug used for treating acne, also slows tumor growth in XPD.

Thus, Moriz Kaposi provided the initial observations of two diseases, one malignant and one premalignant, that later were found to be hereditary.

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