

## HISTORY OF ONCOLOGY

---

# The eminent dermatologist Moriz Kaposi (1837-1902) and the first description of idiopathic multiple pigmented sarcoma of the skin

M. Karamanou<sup>1</sup>, C. Antoniou<sup>2</sup>, A.J. Stratigos<sup>2</sup>, Z. Saridaki<sup>3</sup>, G. Androutsos<sup>1</sup>

<sup>1</sup>Department of History of Medicine, Medical School, University of Athens, Athens; <sup>2</sup>First Department of Dermatology, "Andreas Sygros" University Hospital, Medical School, University of Athens, Athens; <sup>3</sup>Laboratory of Tumor Cell Biology, Medical School, University of Crete, Heraklion, Greece

### Summary

In 1872, the Hungarian born dermatologist Moriz Kaposi that was practicing in Vienna first described a rare endemic disease that bears his name, among elderly persons of Central European or Mediterranean origin named "idiopathic multiple pigmented sarcoma of the skin". Ten years later the Italian

dermatologist Tommaso de Amicis confirms Kaposi's findings. For more than a century the disease was known as a rare low-grade malignancy till the 1980s AIDS epidemic.

**Key words:** HIV disease, idiopathic multiple pigmented sarcoma of the skin, history of dermatology, Moriz Kaposi

### Introduction

Kaposi sarcoma is a systemic multifocal angiomatous tumor, characterized by multiple red to purple macular or papular skin lesions slowly evolving to nodules or plaques. Lesions may coalesce or become ulcerated or may form nodular or fusiform tumors. Most commonly lesions first appear in the distal part of the extremities and spreading later in a more disseminated multifocal pattern. Clinically Kaposi sarcoma is classified into 4 distinct forms: classic that predominately affects elderly males of Jewish, Eastern Europe or Mediterranean origin and represents the form originally described by Moriz Kaposi; African (endemic) that affects children and adults in the eastern half of equatorial Africa; iatrogenic, most commonly encountered in organ transplant recipients undergoing immunosuppressive chemotherapy, cancer patients under chemotherapy or radiation therapy; and epidemic or AIDS-related [1].

Initially described in 1872 by Moriz Kaposi as *idiopathic multiple pigmented sarcoma of the skin*

[2], Kaposi sarcoma was practically unheard, before the 1980s AIDS epidemic, comprising only 0.3% of all cancers in men and 0.1% in women in US and Europe. Afterwards, the incidence of Kaposi sarcoma increased more than a thousand fold in homosexual/bisexual men, intravenous drug abusers and sexually promiscuous men and women at high risk for HIV infection, peaking in 1989 in US the annual incidence at 9.6 per 100,000. Since then, rates have declined to pre-AIDS level thanks to HAART (Highly active anti-retroviral therapy), the effective combined therapy for HIV disease [3].

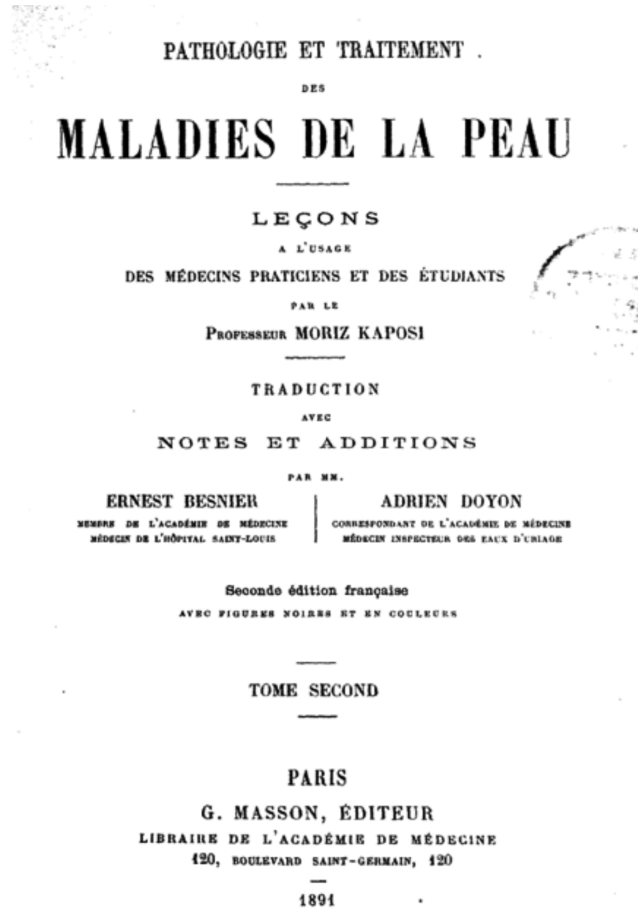
### Moriz Kaposi's life and work

Moriz Kohn was born in Kaposvar, Hungary, on October 23, 1837 in a poor Jewish family. He completed his early education at Pressburg, Bratislava, and in 1856 he enrolled in Vienna's Medical School (Photo 1). On December 13, 1861 he was graduated and soon afterwards he became assistant at the University clinic of Carl-Ludwig



**Photo 1.** The renowned dermatologist Moriz Kaposi.

Sigmund von Ilanor (1810-1883), first Professor of Syphilology at the University of Vienna [4]. In 1866 he completed his thesis entitled: "Syphilis of the mucous membranes of the mouth, throat, nose and larynx" (Syphilis der Schleimhaut der Mund, Rachen, Nasen und Kehlkopfhöhe) and joined the prestigious dermatology department of Vienna's General Hospital directed by the so called "father of German dermatology" Professor Ferdinand Ritter von Hebra (1816-1880) [5,6]. It seems that his choice to devote himself in dermatology would be decisive not only for his professional life but for his personal too. Von Hebra appreciated Kohn's brilliant mind and excellent qualities of clinical researcher and teacher and together formed a close working relationship that culminated in 1869 with the marriage of von Hebra's daughter Martha (1851-1946) to Kohn [6]. Just before the marriage, he decided to embrace Roman Catholicism and changed his name from Kohn to Kaposi, with "Kaposi" being a "play" on the name of his birth place Kaposvar. In his application, he explained that many physicians in Vienna were named Kohn and he wanted to avoid confusion. It



**Photo 2.** Frontispiece of Kaposi's-Besnier's-Doyon's leading textbook of dermatology (French edition).

was said that when Kaposi asked his father-in-law, von Hebra, about his dowry, the later transferred six of his wealthiest patients with severe psoriasis to his care [4].

In 1876, Kaposi was appointed associate Professor of Dermatology and after the death of von Hebra in 1881, he became chairman of Dermatology at the University of Vienna, competing successfully with several distinguished dermatologists, among them his brother-in-law Hans von Hebra (1847-1902) [7]. His election provoked the malicious comments of his colleagues which are reflected in the writings of Professor William-Auguste Dubreuilh (1857-1935), Professor of Dermatology in Bordeaux's School of Medicine: "It is said that he (Kaposi) took Hebra's daughter, house, chair and clientele, leaving the rest to his brother-in-law, Hans von Hebra" [8].

But what remains undisputable is Kaposi's leading contribution to the development of Vienna School of Dermatology. He was acknowledged as a charismatic teacher, a polyglot speaker fluent in German, Hungarian, French and English and his lectures attracted students and physicians from



**Photo 3.** Kaposi's sarcoma after a moulage of the renowned artist Jules Baretta (1834-1923). Hôpital Saint-Louis, Paris.

many countries [7]. Among his several achievements in dermatology, he described first the cutaneous and systemic manifestations of lupus erythematosus (1869/1872); the idiopathic multiple pigmented sarcoma of the skin (1872) that bears his name; xeroderma pigmentosum (1882); lichen ruber moniliformis (1886); and varicelliform eruption (1887) [9]. Also, he was the first to use the pathologic examination in the diagnosis of dermatologic diseases. In 1872, Kaposi completed the von Hebra notable book on dermatology *Textbook of Skin Diseases* (Lehrbuch der Hautkrankheiten) [10] and in 1880, he published his independent work on pathology and treatment of skin diseases [11]. The manuscript was edited and translated in French by the French dermatologists Ernest Besnier (1831-1909) and Andrien Doyon (1827-1907) who did not just translate the text but added the view of the French School of Dermatology creating thus the first syncretic book of dermatology, away of nationalistic fetishes, errors and eccentricities that characterized dermatology and syphilology for more than a century [12] (Pho-

to 2). The Kaposi-Besnier-Doyon opus was considered the most influential textbook of dermatology dominating the field for more than 20 years.

For his significant contribution in dermatology, Kaposi became member of the Imperial Order of Emperor Leopold, he was appointed officer of the Légion d'honneur and he was elected member of many national and international scientific societies. Suffering from two episodes of cerebral stroke, the first in 1900 and the second a year later, Kaposi died in Vienna on March 6, 1902 at the age of 65 years [7].

### The idiopathic multiple pigmented sarcoma of the skin

In 1872, Kaposi described a new entity, observed in 5 men aged 40-68 years with skin lesions primarily involving the feet, that he initially named "idiopathic multiple pigmented sarcoma of the skin". 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 quarter coin to that of the palm of the hand. The lesions then appear on the ankles and forearms and in 2-3 years progress to the face and trunk ....later, they ulcerate and become gangrenous and necrotic" [2]. At autopsy, similar lesions were found in the trachea, esophagus, stomach, liver and bowel. Kaposi mentions that patients were experiencing pain in hands and feet and as the disease was progressing, they were developing fever, hemoptysis and bloody diarrhea. On histological examination of 2 nodules, Kaposi noted: "they presented a picture of small cell sarcoma, with cells appearing in masses and clumps. In some areas there were small hemorrhages in the corium or in the papillae. In addition, there was much yellow-brown to black pigment lying separate from the cells" [2]. Therapeutically, according to Kaposi, no treatment was of value and it was also observed that if the lesions were surgically removed, others appeared. Arsenic administered orally or by injection was ineffective in most cases while x-ray treatment reported later, in 1911, was of some benefit. However, the majority of patients died within 5 years [13].

In 1882, 10 years after Kaposi's first description, the Italian dermatologist Tommaso de Amicis (1838-1924) delineated another group of 12 patients with Kaposi sarcoma. Except for one small child, all were Neapolitan men aged 39-44 years [14]. In 1894 and after further histological stud-

ies, Kaposi suggested to replace the term *pigmented* with *hemorrhagic* in order to distinguish *sarcoma* from *melanosarcoma*, the then term for *malignant melanoma* [7,15]. Over time and after the suggestion of the dermatologist Heinrich Koebner (1838-1904) the disease shifted to *Kaposi's sarcoma* (Photo 3).

## Conclusion

Almost unheard for more than a century, Ka-

posi sarcoma comes to the forefront in 1980s after Alvin Friedman-Kein's report proving its association to HIV disease [16]. In 1994 Yuan Chang, Patrick Moore, and Ethel Cesarman, scientists in Columbia University, linked the etiology of Kaposi sarcoma to human herpesvirus 8 (HHV-8) [17,18]. However, it was Moriz Kaposi whose first description suggested the relationship of skin changes to an associated systemic disease [2,7].

## References

1. Stratigos JD, Katoulis AC, Stratigos AJ. Kaposi's sarcoma. In:Katsabas AD, Lotti TM (Eds):European Textbook of Dermatological Treatment. Berlin, Springer, 2003, pp 239-247.
2. Kaposi M. Idiopathisches multiples Pigmentsarkomen der Haut. Archiv Dermatol Syph 1872;4:265-273.
3. Harris RE. Epidemiology of chronic disease. Burlington (MA), Jones and Bartlett, 2011, pp 262-265.
4. Oriel JD. Moritz Kaposi (1837-1902). Int J Std AIDS 1997;8 :715-717.
5. Kohn M. Die Syphilis der Schleimhaut der Mund, Rachen, Nasen und Kehlkopfhöhle. Erlangen, Enke, 1866.
6. Cribier B, Halioua B, Revuz J, Tilles G. Quelques cas historiques en dermatologie. Paris, Springer, 2011, pp 115-121.
7. Löser C, Plewig G, Burgdorf WHC. Pantheon of Dermatology:Outstanding Historical Figures. London, Springer, 2013, pp 571-581.
8. Dubreuilh W. Souvenirs dermatologiques. In:Deliberationes Congressus Internationalis IX-I, Budapestini 13-21 Septembre 1935, vol IV, pp 53-57.
9. Holubar K., Ferencic-Fatovic S. 1902-2002:A hundred years later, Moriz Kaposi 1837-1902:a historical reappraisal. Wien Klin Wochenschr 2001;113:885-893.
10. Hebra F, Kaposi M. Lehrbuch der Hautkrankheiten. Stuttgart, Enke, 1872.
11. Kaposi M. Pathologie und Therapie der Hautkrankheiten und Vorlesungen. Wien, Urban & Schwarzenberg, 1880.
12. Besnier E, Doyon A, Kaposi M. Pathologie et traitement des maladies de la peau :Leçons à l'usage des médecins praticiens et des étudiants. Paris, Masson, 1891.
13. Allbutt TC. A system of medicine, vol 9. New York, MacMillan, 1911, p 846.
14. de Amicis T. Studio clinic ed anatomo-patologico di dodici nuove osservazioni di Dermo-polimelano-sarcoma idiopatico. Napoli, Trani, 1882.
15. Kaposi M . Zur Nomenclatur des idiopathischen Pigmentsarcoms Kaposi. Arch Dermatol Syph 1894 ;29:164.
16. Friedman-Kien AE. Disseminated Kaposi's sarcoma syndrome in young homosexual men. J Am Acad Dermatol 1981;5:468-471.
17. Chang Y, Cesarman E, Pessin MS et al. Identification of herpesvirus-like DNA sequences in AIDS-associated Kaposi's sarcoma. Science 1994;266:1865-1869.
18. Moore PS, Chang Y. Detection of herpesvirus-like DNA sequences in Kaposi's sarcoma in patients with and without HIV infection. N Engl J Med 1995;332:1181-1185.

**Correction**

In vol.18 (1): 124-130,2013 issue (c-MYC and h-TERT co-expression in colon adenocarcinoma: a tissue microarray digitized image analysis) there was a mistake in the authors' names sequence. The correct sequence is: Georgakopoulos G., Tsiambas E., Korkolopoulos P., Kavantzas N., Karameris A., Ragkos V., Rigopoulos D.N., Vilaras G., Athanasiou A.E., Tatiou D., Patsouris E. The authors apologise for their mistake.