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KAPOSI'S SARCOMA

MULTIPLE IDIOPATHIC
HEMORRHAGIC
SARCOMA

his monograph is the initipublication in a series of tudies regarding the Cucaneous Manifestations of the Diseases of the Reticuloendothelial System.

American Lecture Series®



KAPOSI'S SARCOMA

Multiple Idiopathic Hemorrhagic Sarcoma

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INTRODUCTION

In the eighty-six years since Kaposi first described Multiple Idiopathic Hemorrhagic Sarcoma, the disease has continuously stimulated interest. In the course of time every conceivable facet of the interesting disease has been recorded and discussed: racial incidence, age and sex predisposition, terminology and nosology, possible causative factors, pathogenesis, symptomatology, hematologic aspects and treatments. There is, as a result, a voluminous literature on the subject, so large that to keep abreast of it is almost impossible. Dr. Bluefarb, in undertaking to review and to assess critically this vast fund of accumulated knowledge, has rendered an invaluable service.

Gathering the material for this monograph was obviously a laborious task. It is not important that he is unable to ascribe the disease to a single cause; it is not important that his discussion of treatment ends on a depressing note. It is important that all available facts are here recorded in one small monograph. In this easily accessible book is a critical analysis of the various views as to the cause of the disease, a sober appraisal of the various therapies that have been suggested; indeed, a full consideration of every phase of the disease—and also, many little-known facts are herein recorded.

Dr. Bluefarb is uniquely fitted for the task for he has long been a serious student of the cutaneous manifestations of lymphomas and of so-called reticuloendothelial diseases. He has contributed significantly to the literature of this subject. He has presented the material in this monograph on Kaposi's sarcoma simply and in logical sequence. It should long serve as an authoritative source of reference.

Herbert Rattner, M.D.

PREFACE

This monograph is one of a contemplated series of studies on the cutaneous manifestations of reticuloendothelial system diseases. The recent increased interest in this group of diseases, particularly the cutaneous aspects, seems to justify this series of publications, since no text in the English language is available on this subject. The cutaneous manifestations of this group of "blood" dyscrasias may exhibit such a striking and diverse morphological picture that an extensive portion of the dermatological literature is devoted to this subject. We have consulted, and quoted freely from, all publications on this subject which are included in the bibliography and wish to acknowledge indebtedness to these contributors.

We are indebted to Francesco Ronchese, M.D., and Samuel J. Zakon, M.D., for the use of their illustrations. We are also indebted to Herbert Rattner, M.D., Chairman of the Department of Dermatology, Northwestern University Medical School, for reading this manuscript and writing the introduction; to Miss Madge Walsh, Chief Medical Photographer, Northwestern University, for the photomicrographs, and to Miss Sophie Price, of the Medical Library Staff, Northwestern University, for verification of the bibliography.

S.M.B.

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KAPOSI'S SARCOMA



1. Moritz Kaposi (1837-1902). A previously unpublished photograph (through the courtesy of Paul E. Bechet, M.D., and Arthur C. Curtis, M.D.).

I

HISTORICAL

In 1872 Kaposi (132a) first described a condition which he named "Idiopathic Multiple Pigment Sarcoma of the Skin." The present designation of "Kaposi's Sarcoma" was suggested by Köebner (137b) in 1891. Kaposi agreed to this designation, but twenty years later (132b) changed it to "sarcoma idiopathicum multiplex hemorrhagicum," a name he believed was more accurately descriptive of the condition, and would also indicate the source of pigment (hemosiderin) in the lesions. In his classical description of the disease, Kaposi noted the symmetry and multiplicity of the lesions from the onset as a characteristic of the disease. This report was based on five cases, but he later added 20 cases which he had observed. He regarded the disease as distinctive and believed it to be a special and peculiar form of round cell sarcoma, which occasionally presented characteristic spindle cells.

II

SYNONYMS

The variety of descriptive terms suggested since Kaposi's original report give evidence of the varied conceptions of this disease. There are differences regarding the histologic changes of the various lesions in one individual, as well as in portions of the same lesion, which accounts, in part, for the various theories about this disease.

Year	Writer	Synonyms
1868	Kaposi	Primaren idiopathischen hautsar- coma
1872	Kaposi	Sarcoma idiopathicum multiplex pig- mentosum
1878	Tanturri	Sarcoma idiopathicum telangiec- todes
1883	Hardaway	Sarcoma cutis
1884	Babes	Angiosarcoma peritheliale fusocel- lulare
1889	Funk	Sarcomatosis gummatoides
1891	Köebner	Sarcoma idiopathicum multiplex hemorrhagicum der extremitaden
1894	Kaposi	Sarcoma idiopathicum multiplex hemorrhagicum
1894	Unna	Acrosarcoma multiplex cutaneum telangiectodes
1898	Tommasoli	Primitives hemorrhagisches acrosar- coid
1899	Bernhardt	Sarcomata idiopathica multiplicia pigmentosa cutis

Synonyms

Year	Writer	Synonyms
1899	Gilchrist	Angiosarcoma
1899	Pospelow	Acroangioma hemorrhagicum
1899	Sellei	Granuloma multiplex hemorrhagi-
		cum
1901	Radaeli	Angioendothelioma cutaneum
1902	Lieberthal	Multiple idiopathic hemorrhagic sar- coma
1902	Pelagatti	Acrosarcoma
1910	Sequeira	Granuloma angiomatodes
1912	Rasch	Sarcoma cutaneum telangiectaticum
		multiplex
1912	Sternberg	Kaposi's sarcoma
1913	Martinotti	Sarcoma hemangioendotheliale in-
		travasculare
1915	Sibley	Granuloma multiplex hemorrhagi- cum
1917	Gaucher	Sarcomatosis primitiva telangectosica
1922	Bertaccini	Angioendothelioma cutaneum
1925	Guiffre	Sarcomatosis telangiectatics cutanea
		idiopathica generalisata
1926	Guarini	Multiple angiosarcoma
1927	Hamdi and Halil	Perithelioma multiplex nodulosum
		cavernosum lymphangiectodes cu- taneum
1928	Hudelo, Cailliau	Pseudosarcomatosis telangiectatica
	& Chene	
1928	Pautrier and	Pseudosarcoma (Kaposi) myoneuro-
	Diss	vascular dysgenesis
1928	Nicholas and	Sarcomatose telangiectatique pig-
	Favre	mentaire
1931	Tramontano &	Hemangioendothelioma cutaneum
	Fittipaldi	
1932	Hamdi & Resat	Acroperithelioma idiopathicum mul-
		tiplex cavernosum lymphangiec-
		todes cutaneum

Year	Writer	Synonyms
1934	Kusnezow	Endoperithelioma sarcomatodes multiplex idiopathicum (typus Kaposi)
1935	Lang & Haslhofer	Systematized angiomatosis
1939	Choisser & Ramsey	Angioreticuloendothelioma
1953	Ronchese and Kern	Angioreticulomatosis