ACTA MÉDICA PORTUGUESA
A Revista Científica da Ordem dos Médicos

Kaposi's Sarcoma: Demographic and Clinical Features, Histopathology, Treatment, and Outcomes in a 10-Year Lisbon Hospital Study

Sarcoma de Kaposi: Características Clínicas e Demográficas, Histopatologia, Tratamento e Prognóstico em 10 Anos num Hospital em Lisboa

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Acta Med Port 2025 Aug;38(8):460-468 ▪ https://doi.org/10.20344/amp.17991

ABSTRACT

Introduction: Kaposi's sarcoma (KS) is a rare angioproliferative neoplasm associated with human herpesvirus 8 infection, presenting as four subtypes: classic, endemic, iatrogenic, and epidemic. While well documented globally, comprehensive data on KS in Portugal remain scarce. The aim of this retrospective study was to provide a detailed analysis of KS cases diagnosed at Hospital Santa Maria, in Lisbon, between 2014 and 2023.

Methods: A total of 113 histopathologically confirmed KS cases were included, focusing on demographic, clinical, and histopathological characteristics, as well as treatment strategies and outcomes.

Results: The mean age at diagnosis was 59.4 years, with a male-to-female ratio of 4.4:1. Most patients (50.4%) were of African origin. Epidemic KS (45.1%) was the most prevalent subtype. Lesions mainly affected the lower limbs (47.8%), and disseminated, mucosal, and extracutaneous involvement were more common in HIV-positive patients. Tumor-stage lesions were frequent (59.3%). Single-modality treatment was used in 53.1% of cases, while 40.7% required combined therapies. Relapse rates were highest in endemic (39.1%) and iatrogenic KS (28.6%) subtypes. The disease-specific mortality rate was 8%.

Conclusion: Our findings suggest that KS remains a significant concern, particularly in immunosuppressed patients. Early diagnosis and multidisciplinary management are essential to improve outcomes. However, limitations such as potential biases from its retrospective design and the single-center scope should be considered.

Keywords: Sarcoma, Kaposi/diagnosis; Sarcoma, Kaposi/epidemiology; Sarcoma, Kaposi/pathology; Sarcoma, Kaposi/therapy

RESUMO

Introdução: O sarcoma de Kaposi (SK) é uma neoplasia angioproliferativa rara associada à infeção pelo herpes vírus humano 8, apresentando-se em quatro subtipos: clássico, endémico, iatrogénico e epidémico. Embora bem documentado globalmente, dados abrangentes sobre o SK em Portugal permanecem escassos. Este estudo retrospetivo tem como objetivo fornecer uma análise detalhada dos casos de SK diagnosticados no Hospital Santa Maria, em Lisboa, de 2014 a 2023.

Métodos: Um total de 113 casos de SK confirmados histologicamente foram incluídos, com foco nas características demográficas, clínicas e histopatológicas, bem como nas estratégias terapêuticas e desfechos.

Resultados: A idade média ao diagnóstico foi de 59,4 anos, com uma razão homem-mulher de 4,4:1. A maioria dos doentes (50,4%) era de origem africana. O SK epidémico (45,1%) foi o subtipo mais prevalente. As lesões afetaram principalmente os membros inferiores (47,8%), e o envolvimento disseminado, das mucosas e extracutâneo foi mais comum em doentes HIV-positivos. As lesões em estádio tumoral foram frequentes (59,3%). O tratamento com modalidades únicas foi utilizado em 53,1% dos casos, enquanto 40,7% exigiram terapêuticas combinadas. As taxas de recidiva foram mais elevadas nos subtipos endémico (39,1%) e iatrogénico (28%). A taxa de mortalidade específica da doença foi de 8%.

Conclusão: Os nossos resultados sugerem que o SK continua a ser motivo de preocupação significativa, particularmente em doentes imunossuprimidos. O diagnóstico precoce e a abordagem multidisciplinar são essenciais para melhorar o prognóstico. No entanto, as limitações inerentes ao desenho retrospetivo e ao caráter unicêntrico do estudo devem ser consideradas na interpretação dos dados.

Palavras-chave: Sarcoma de Kaposi/diagnóstico; Sarcoma de Kaposi/epidemiologia; Sarcoma de Kaposi/patologia; Sarcoma de Kaposi/tratamento

KEY MESSAGES

- Kaposi's sarcoma is a rare neoplasm that remains underexplored in Portugal, with few comprehensive studies available despite its clinical significance.
- In our study, the disease predominantly affected men, with 50.4% of patients being of African origin. The epidemic subtype is the most common, linked to Portugal's HIV prevalence.
- Advanced-stage lesions and extracutaneous involvement were more frequent in immunosuppressed patients, particularly those living with HIV.
- Tailored therapies, including highly active antiretroviral therapy (HAART), are critical for epidemic KS. Early diagnosis and multidisciplinary approaches are essential for better outcomes.

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Recebido/*Received*: 16/01/2025 - **Aceite/*Accepted*:** 29/04/2025 - **Publicado *Online*/*Published Online*:** 20/06/2025 - **Publicado/*Published*:** 01/08/2025

INTRODUCTION

Kaposi's sarcoma (KS) is an uncommon malignant vascular neoplasm that most frequently affects the skin, although it can also involve mucous membranes, lymph nodes, and visceral organs.[1] First described by Moritz Kaposi in the late 1800s as an "idiopathic multiple pigmented sarcoma of the skin",[2] KS remained enigmatic until 1994, when human herpesvirus 8 (HHV-8) was recognized as the major causative agent, particularly in patients with acquired immunodeficiency syndrome (AIDS),[3] leading to its designation as Kaposi sarcoma herpesvirus. Human herpesvirus 8 is an oncogenic virus with variable prevalence worldwide, being more common in men who have sex with men (MSM), and in regions such as sub-Saharan Africa and Mediterranean-bordering countries.[4]

Kaposi's sarcoma exhibits considerable diversity in its epidemiological, clinical, and histopathological manifestations. Four main clinical forms are recognized: classic KS, affecting mainly elderly men of Mediterranean, Eastern Europe, or Middle Eastern origin; endemic KS, prevalent in sub-Saharan Africa; iatrogenic KS, occurring in immunosuppressed individuals, such as organ transplant recipients; and epidemic KS, associated with human immunodeficiency virus (HIV) infection.[5] Although the pathogenesis of KS remains incompletely understood, it is thought to result from a combination of viral infection, immunosuppression, and environmental exposures in genetically predisposed individuals.[6]

Diagnosis is based on clinical assessment, supported by histopathological and immunohistochemical analysis. Kaposi's sarcoma lesions exhibit three primary clinical and pathological stages – macule/patch, plaque, and tumor/nodule –, which often coexist in the same patient, reflecting the disease's progression.[7] Treatment approaches are heterogeneous and tailored, combining local and systemic therapies depending on the individual case.[8]

In Portugal, the demographic and clinical profiles of KS patients remain underexplored, with limited small-scale retrospective studies available.[9-11] Most of these studies lack a detailed breakdown of KS subtypes, making it difficult to assess potential epidemiological shifts or specific clinical challenges in different patient populations. This retrospective study seeks to provide a comprehensive analysis of patients diagnosed with KS at the Dermatology Department of Hospital Santa Maria, in Lisbon, over a 10-year period (2014 - 2023). By examining demographic, clinical, and histopathological characteristics, treatment modalities, and outcomes, this study aims to broaden the understanding of KS in Portugal and contribute to the optimization of patient care.

METHODS

Participant selection

Participants were selected from the database of individuals diagnosed with KS through skin biopsy at the Dermatology Department of Hospital Santa Maria, in Lisbon, Portugal, between January 2014 and December 2023. A total of 113 patients with primary or recurrent KS were included. Inclusion criteria consisted of a confirmed diagnosis of KS and the age of 18 years or older. Histopathological confirmation was conducted using hematoxylin and eosin staining, and immunohistochemistry positive for HHV-8, in cases with equivocal findings. Lesions were classified by histological stage based on their features:

- Macule/patch stage: presence of thin-walled vascular spaces in the upper dermis, accompanied by a sparse mononuclear infiltrate consisting of lymphocytes, plasma cells, and macrophages.
- Plaque stage: enlarged vascular spaces with a denser inflammatory infiltrate and spindle cell bundles surrounding angioproliferative clefts.
- Tumor/nodular stage: cutaneous lesions larger than one centimeter characterized by well-defined nodules composed of large fascicles of spindle-shaped endothelial cells and compact vascular slits.

Patients with incomplete medical records were excluded from the study. Ethics committee approval was obtained from the institutional Review Board of the hospital, and all procedures adhered to the principles outlined in the Declaration of Helsinki.

Data collection

Data were collected from electronic medical records, encompassing demographic details (age at diagnosis, sex, geographic origin, and comorbidities), clinical characteristics (location, appearance, number of lesions, extracutaneous involvement, and KS subtype), histopathological findings, treatment modalities, follow-up information, and patient outcomes. However, as this is a retrospective study, some electronic medical records were occasionally incomplete, which may have led to missing data for certain variables.

Statistical analysis

Statistical analyses were performed using SPSS® software (version 26.0). Descriptive statistical analysis was performed using measures of central tendency and dispersion measures. Inferential analysis was employed with both parametric and non-parametric tests. To evaluate the normality of distribution Kolmogorov-Smirnov and Shapiro-Wilk tests were employed when appropriate. Levene's test was used to verify homogeneity of variances. Qualitative

variables were compared with the χ^2 test (QQ) and Fisher's exact test. Quantitative variables were compared with the Student t-test or an independent samples Mann-Whitney U test (MW). Inter-rater reliability was calculated through Cohen's κ with the strength of agreement being classified according to Landis *et al*.[12] Significance was defined as α = 0.05, with all tests being bilateral.

Table 1 – Demographic, clinical and histopathological features, treatment modalities, and outcomes of KS patients treated at our institution between 2014/2023

		n (%)
Age	Minimum	27
	Maximum	97
	Mean (standard deviation)	59.4 (17.7)
Sex	Male	92 (81.4)
	Female	21 (18.6)
Origin	Africa	57 (50.4)
	Europe	50 (44.3)
	Others	6 (5.3)
Localization	Lower limbs	54 (47.8)
	Disseminated	20 (7.7)
	Head and neck	3 (2.7)
	Mucosal involvement	6 (5.3)
Lesion type (clinical)	Tumors/nodules	74 (65.5)
	Plaques	26 (22.1)
	Macules/patches	14 (12.4)
Number of lesions	< 10	70 (62)
	> 30	17 (15)
KS subtype	Epidemic	51 (45.1)
	Classic	25 (22.1)
	Endemic	23 (20.4)
	Iatrogenic	14 (12.4)
Involvement	Only cutaneous	93 (82.3)
	Cutaneous and extracutaneous	20 (17.7)
HIV	No	62 (54.9)
	Yes	51 (45.1)
Transplanted	No	106 (93.8)
	Yes	7 (6.2)
Iatrogenic immunosuppression	No	110 (97.3)
	Yes	3 (2.7)
HHV-8 status (peripheral blood)	Positive	5 (4.4)
Treatment	Single	60 (53.1)
	Multiple	46 (40.7)
Relapse	No	82 (72.6)
	Yes	25 (22.1)
Deceased	No	80 (70.8)
	Yes	33 (29.2)
Death related to KS	No	104 (92%)
	Yes	9 (8%)

RESULTS

This study included 113 patients, whose demographic, clinical and histopathological features, treatment modalities, and outcomes are summarized in Table 1.

Demographics

Patients' ages were between 27 to 97 years old, with a mean age at diagnosis of 59.4 (± 17.7) years. A statistically significant difference in mean age was observed, with patients with epidemic KS being younger compared to those with other variants, while patients with classic KS were older than the remaining subtypes (Table 2).

The cohort was predominantly male (n = 92, 81.4%), with a male-to-female ratio of 4.4:1. More than half of the patients (n = 57, 50.4%) were of African origin. The diagnosis of KS was made in 59.3% (n = 67) of cases during Dermatology appointments, followed by Dermatology urgent care in 24.8% (n = 28), and the inpatient setting of the Infectious Disease department in 15.9% (n = 18) of cases.

Clinical characteristics

Lesions were most found exclusively in the lower limbs, affecting 47.8% of patients (n = 54), followed by lower limb involvement with other sites in 19.5% (n = 22; predominantly the upper limb in 16 cases). In addition, 17.7% of patients (n = 20) presented with disseminated cutaneous lesions, of which 70% (n = 14) were HIV-positive. Mucosal involvement was observed in six patients, four of whom were HIV-positive. The most common clinical lesion type was tumors or nodules (n = 74, 65.5%), followed by plaques (n = 25, 22.1%).

The number of lesions varied, with 61.9% (n = 70) having less than 10 lesions. Regarding the clinical diagnosis, the hypothesis of KS was considered in 75.2% (n = 85) of cases. The other most frequently considered clinical hypotheses were pyogenic granuloma (n = 11, 9.7%), malignant melanoma (n = 5, 4.4%), and post-inflammatory hyperpigmentation (n = 3, 2.7%).

Regarding the clinical subtypes, the epidemic form of KS was the most common (n = 51, 45.1%), followed by the classic form (n = 25, 22.1%), endemic (n = 23, 20.4%), and iatrogenic (n = 14, 12.4%).

Immunosuppression was present in 57.5% of patients (n = 65), mainly due to HIV infection (n = 51, 45.1%), renal transplantation (n = 7, 6.1%), ongoing chemotherapy for other malignancies (n = 3, 2.7%), or chronic corticosteroid use (n = 3, 2.7%). Extracutaneous involvement, predominantly involving lymph nodes, lungs, and stomach, occurred in 17.7% (n = 20) of cases, with a higher prevalence in immunosuppressed patients (n = 17, 85%), especially in HIV-positive patients (n = 15, 75%).

Among the HIV-positive patients, 56.9% (n = 29) had CD4 counts below 200 cells/mm^3, with a median CD4 count of 53 cells/mm^3 in this group. However, 19 patients had CD4 counts above this threshold and 10 patients had CD4 counts above 400 cells/mm^3 at the time of diagnosis.

The HHV-8 status in peripheral blood samples was assessed in only five patients, all of whom tested positive for the virus.

Histopathological features

Histopathological analysis confirmed KS in all cases, with immunohistochemistry for HHV-8 showing positivity in all cases where performed (n = 51) – Fig. 1. Tumors or nodules (n = 67, 59.3%) were the predominant lesion type, followed by plaques (n = 30, 26.5%), and patches (n = 16, 14.1%). Cohen's kappa coefficient for agreement between the clinical and histopathological classifications of KS

Table 2 – Comparative overview of the main clinical subtypes of KS

	Epidemic KS (n = 51)	Classic KS (n = 25)	Endemic KS (n = 23)	Iatrogenic KS (n = 14)
Mean age (standard deviation), years	49.1 (13) ***p* < 0.001** (MW)	75.7 (11.5) ***p* < 0.001** (MW)	59.5 (18) *p* = 0.975 (t test)	67.6 (15.8) *p* = 0.056 (t test)
Male, n (%)	42 (82.4)	20 (80)	18 (78.3)	12 (85.7)
Extracutaneous involvement, n (%)	15 (29.4)	0	3 (13)	2 (14.3)
Relapse rate, n (%)	8 (15.7)	5 (20)	9 (39.1)	4 (28.6)
Disease related mortality, n (%)	5 (9.8)	1 (4)	2 (8.7)	1 (7.1)
Histological classification				
Tumor/nodule, n (%)	28 (54.9) *p* = 0.389 (QQ)	14 (56) *p* = 0.704 (QQ)	20 (86.9) ***p* = 0.002** (QQ)	5 (35.7) *p* = 0.055 (QQ)
Plaque, n (%)	15 (29.4) *p* = 0.532 (QQ)	8 (32) *p* = 0.484 (QQ)	2 (8.7) ***p* = 0.030** (QQ)	5 (35.7) *p* = 0.518 (Fisher)
Macule/patch, n (%)	8 (15.7) *p* = 0.673 (QQ)	3 (12) *p* = 1 (Fisher)	1 (4.4) *p* = 0.186 (Fisher)	4 (28.6) *p* = 0.111 (Fisher)

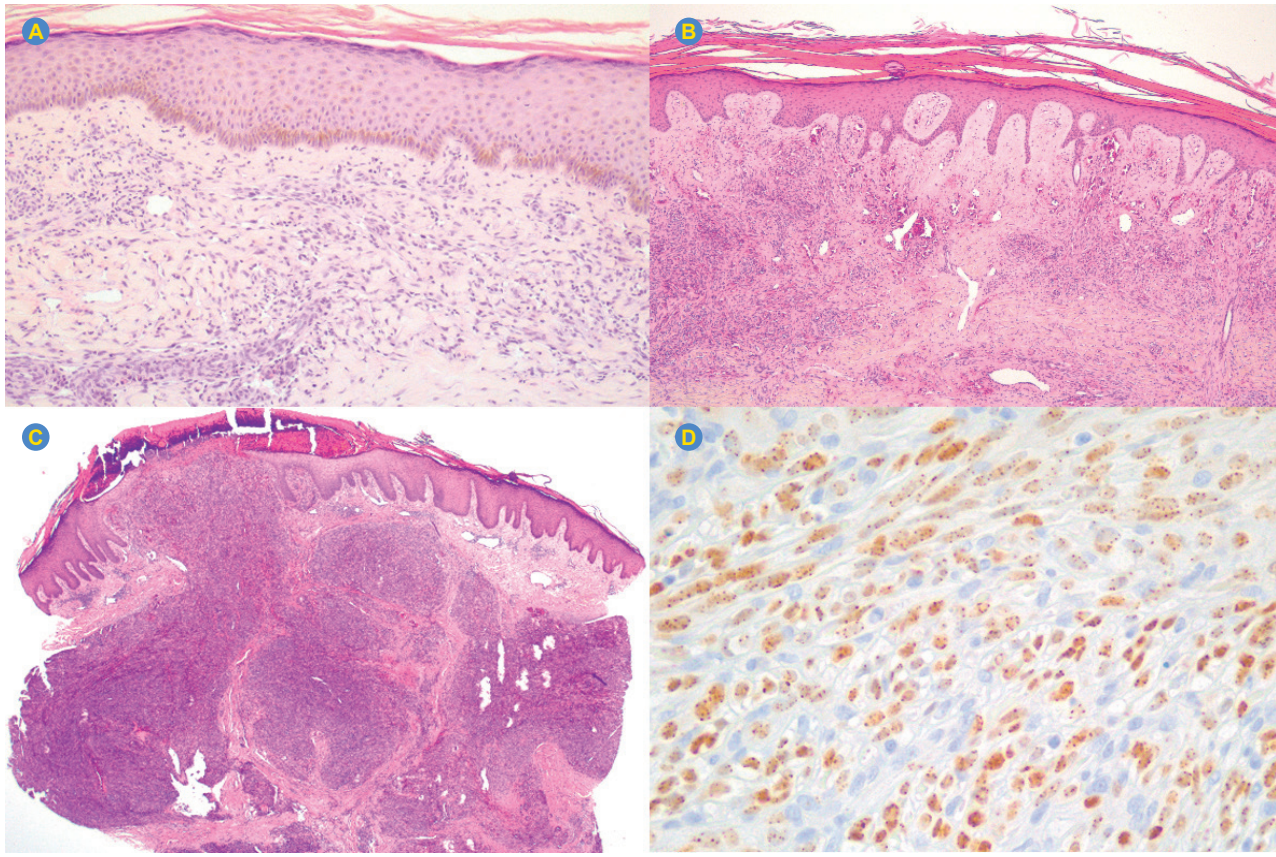

Figure 1 – Histopathological features. (A) Patch stage, H&E x 100; (B) Plaque stage, H&E x 40; (C) Tumor stage, H&E x 16; (D) Immunohistochemistry positive for HHV-8.

showed substantial agreement [κ = 0.60 (95% CI, 0.47 - 0.74), $p < 0.001$] between the two classifications.

Patients with endemic KS showed a significantly higher frequency of tumors/nodules and, to a lesser extent, plaques, compared to other subtypes (Table 2).

Treatment modalities

A single treatment modality was used in 53.1% of patients (n = 60), while 40.7% (n = 46) required multiple approaches (Fig. 2). In three transplant patients, the only treatment provided was the reduction of immunosuppressive

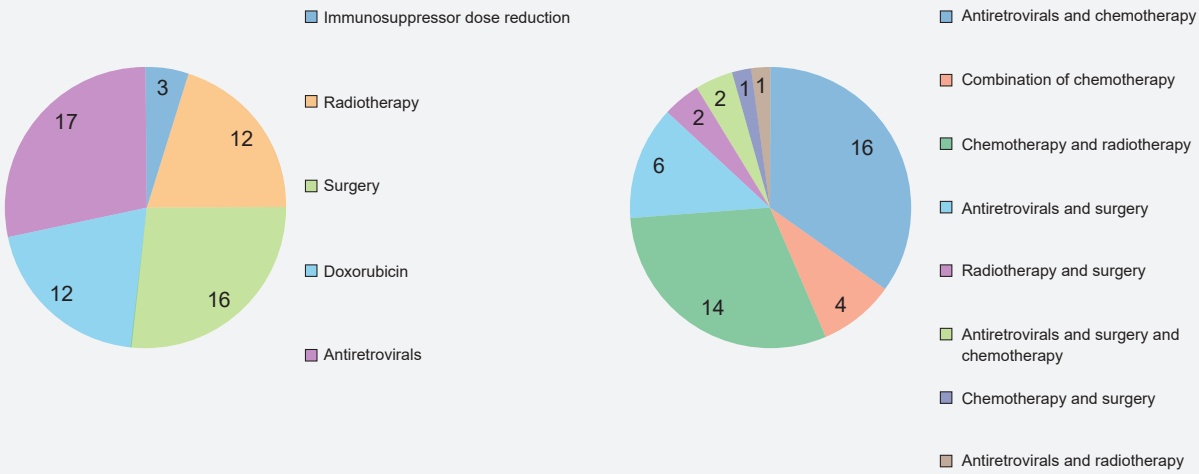

Figure 2 – Distribution of single and combined treatment regimens in patients with KS in our cohort

therapy. Treatment data was unavailable in seven cases.

Local therapies, such as surgical excision or radiotherapy, were used in 58 patients (29 for each modality). Systemic therapies were used in 54.9% (n = 62) of cases, with highly active antiretroviral therapy (HAART) for HIV-positive patients being the most common (n = 42, 37.2%). In 17 patients, HAART was used as the only therapy. Chemotherapy was administered to 44.2% (n = 50) of patients, with pegylated liposomal doxorubicin being the most used chemotherapeutic agent (n = 24, 38.7%). Thirty-four patients (30.1%) required second- or third-line chemotherapy with various systemic combinations such as doxorubicin, paclitaxel, pembrolizumab, and etoposide.

Response to treatment

A complete response maintained for at least two years was achieved in 30.1% (n = 34) of patients, while 65 patients (58.4%) had a partial response. Six patients had no response to treatment (5.3%) – Fig. 3.

Disease progression or relapse during the follow-up period occurred in 26 patients (23%) despite treatment. Notably, relapse rates varied between the different types of KS. Endemic KS had the highest relapse rate, affecting 39.1% (n = 9) of patients, followed by iatrogenic KS with a relapse rate of 28.6% (n = 4). In contrast, classic KS had a relapse rate of 20% (n = 5), while epidemic KS had the lowest rate at 15.7% (n = 8).

Mortality

The overall mortality rate in the cohort was 29.2% (n = 33), and the disease-specific mortality rate for KS was 8% (n = 9). Overall mortality was highest in patients with epidemic KS (n = 15, 45.5%), followed by endemic KS (n = 7, 21.2%), iatrogenic KS (n = 7, 21.2%), and classic KS (n = 4, 12.2%). Mortality due to KS dissemination was highest in patients with epidemic KS (n = 5, 56.6%), followed by endemic KS (n = 2, 22.2%).

DISCUSSION

Kaposi's sarcoma, even though it is a rare condition, remains a significant global health concern, particularly among immunosuppressed individuals and African populations.[13] Our hospital holds a privileged position for studying this neoplasm due to its close collaboration with the Infectious Diseases Department – resulting in a high influx of HIV-positive patients –, its role as a reference center for Portuguese-speaking African countries (PALOP), and its distinctive Dermatology urgent care service in Portugal, which manages a high volume of diverse patient cases.

Demographically, our findings concur with previous studies, confirming Portugal as one of the European countries with a higher age-standardized incidence rate of KS.[13,14] The mean age at diagnosis was 59.4 years, which is consistent with reports from other studies,[10,11,15] highlighting KS's predilection for older adults, especially those in their sixth

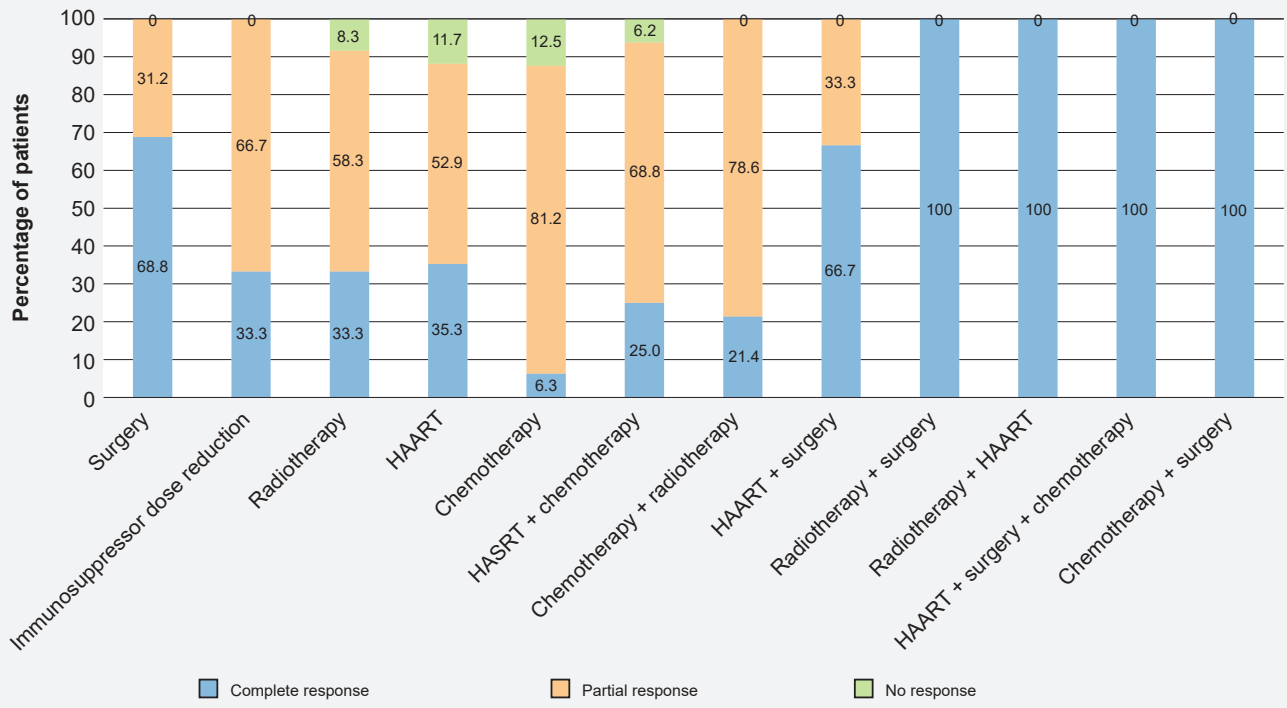

Figure 3 – Distribution of response to treatment according to each treatment regimen in patients with KS in our cohort

decade of life. However, our cohort had a younger mean age than reported in Turkey,[16] where classic KS predominates. This is likely due to the higher prevalence of epidemic KS in our cohort (45.1% *vs* 1.1%), which typically affects younger HIV-positive individuals, contrasting with classic KS that mainly impacts older Mediterranean men.[17]

A comparative overview of published Portuguese series is presented in Table 3, highlighting key epidemiological differences. Notably, our cohort includes a higher proportion of patients of African origin and a broader representation of KS subtypes, particularly the epidemic and endemic forms. These distinctions may reflect both evolving demographic patterns and the specific referral profile of our tertiary hospital.

In terms of sex, our study's male-to-female ratio of 4.4:1 is closely related with published data from European countries, showing KS's higher incidence in men. This may result from biological factors, like the role of sex steroids in immune regulation, and epidemiological factors, including the higher prevalence of HIV among MSM, a high-risk group for epidemic KS.[18,19]

The cohort's geographical diversity is notable, with 50.4% of patients originating from African countries, where endemic and epidemic forms of KS are more prevalent.[20] This diversity reinforces the role of Hospital de Santa Maria as a reference center for this population, contributing to a higher prevalence of these forms of KS in our cohort and providing a unique opportunity to study KS in these

Table 3 – Kaposi's sarcoma cases reported from Portugal

	Junger J[9]	Resende C[10]	Calvão-da-Silva J[11]	Present study
Year	2013	2014	2021	2025
Region	Lisbon	Lisbon	Coimbra	Lisbon
Period, years	19 (1994 - 2012)	13 (2001 - 2013)	10 (2010 - 2019)	10 (2014 - 2023)
n	118	91	38	113
Age, years (mean)	45	59.8	38	59.4
Male/female ratio	3.1	6	6.6	4.4
Origin				
Europe	62 (52.5%)	85 (93.4%)	31 (81.6%)	50 (44.3%)
Africa	36 (30.5%)	6 (6.6%)	-	57 (50.4%)
Others	-	-	-	6 (5.3%)
Location				
Lower extremity	67 (56.8%)	66 (74.2%)	9 (59%)	54 (47.8%)
Upper extremity	6 (5.1%)	-	1 (2.6%)	9 (8%)
Lower/upper extremity	13 (11%)	-	-	18 (16%)
Genitals	2 (1.7%)	-	3 (7.9%)	0
Trunk	12 (10.2%)	-	2 (5.3%)	3 (2.7%)
Head/neck	-	-	1 (2.6%)	3 (2.7%)
Generalized	-	-	12 (31.6%)	20 (17.7%)
Systemic involvement	7 (5.9%)	-	14 (37%)	19 (16.8%)
Immunosuppression	-	-	26 (68.4%)	65 (57.5%)
Variant				
Epidemic	74 (62.7%)	28 (30.8%)	16 (42.1%)	51 (45.1%)
Classic	26 (22%)	61 (67%)	12 (31.6%)	25 (22.1%)
Endemic	5 (4.3%)	1 (1.1%)	0	23 (20.4%)
Iatrogenic	13 (11%)	1 (1.1%)	10 (26.3%)	14 (12.4%)
Histopathology				
Macule/patch	-	14 (15.4%)	-	16 (14.1%)
Plaque	-	29 (35.8%)	-	30 (26.5%)
Tumor/nodule	-	48 (52.7%)	-	67 (59.3%)

-: not available; n: number of patients

high-risk populations.

A key observation is the diversity of KS subtypes, with epidemic KS as the most prevalent (45.1%), aligning with Portugal's relatively high HIV prevalence in Europe.[21] Interestingly, the iatrogenic subtype, accounting for 12.4% of cases, was predominantly seen in solid organ transplant recipients.

Lesions primarily affected the lower limbs (47.8%), consistent with a global pattern of KS.[22] Disseminated cutaneous lesions and mucosal involvement were more common in HIV-positive patients, indicating more advanced disease stages.[23] Most patients (82.3%) had cutaneous-only involvement, while 17.7% had extracutaneous spread at the time of the diagnosis, mainly involving lymph nodes, lung, and the gastrointestinal tract, especially among patients with epidemic KS. This finding lines up with previous studies, showing a higher incidence of extracutaneous involvement in HIV-positive patients, even in the post-HAART era.[24,25] In contrast, classic KS showed no extracutaneous involvement, consistent with its typically indolent nature.

Despite being an AIDS-defining illness commonly associated with CD4 counts below 200 cells/mm^3, KS remained prevalent even in HIV-positive patients with higher CD4 counts and suppressed HIV viral loads. This persistence may be attributed to immune dysfunction, weaker responses to HHV-8, and increased cancer risk.[26]

The histopathological analysis confirmed advanced-stage KS in most cases, with 59.3% of patients presenting with nodular or tumor-stage lesions, facilitating KS as the primary diagnostic consideration during initial consultations. However, early-stage lesions such as macules or patches posed diagnostic challenges, even with histological analysis, complicating early detection.[27,28] In these cases, immunohistochemical analysis for HHV-8 was performed when needed to support the diagnosis.

The high concordance between clinical and histopathological classifications emphasizes the reliability on clinical staging, which is particularly valuable when histopathological resources are limited.

Given the heterogeneity of KS, there are no standard therapeutic guidelines, leading to varied treatment approaches and outcomes.[29] In this cohort, most patients (53.1%) were managed with a single treatment modality, while 40.7% required combined therapies.

Among transplant patients, reducing immunosuppressive therapy was the primary management strategy, demonstrating the delicate balance between controlling KS and preventing organ rejection.[30]

Local treatments, such as surgical excision and radiotherapy, were used in nearly half of the cases, especially in patients with localized disease. Notably, cryosurgery and electrosurgery – often employed in dermatological practice – were not used in this cohort, likely due to the advanced stage of most lesions and the preference for more definitive treatments.

Systemic chemotherapy, particularly pegylated liposomal doxorubicin, was the most used systemic regimen, either alone or in combination, followed by paclitaxel, aligning with international treatment guidelines.[8,29,31]

The majority of patients had complete or partial responses to treatment, particularly when combined with local therapies. Notably, HAART alone was sufficient for achieving partial and complete responses in 88.2% of patients, emphasizing its critical role in managing epidemic KS. Endemic KS and iatrogenic KS had the highest relapse rates, possibly due to more advanced disease at diagnosis, difficulty in achieving long-lasting immune control of HHV-8, and persistent immunosuppression.[32,33]

The overall disease-specific mortality rate of 8% is in line with findings from other studies.[11,34] This rate was consistent across subtypes, except for classic KS, which had the lowest mortality, reflecting its typically indolent course. Although KS is often manageable, it remains a significant cause of mortality in patients with advanced or systemic disease.

This study is one of the largest KS cohorts analyzed in Portugal, providing a robust dataset that encompasses a diverse patient population, including individuals from the Portuguese-speaking African countries and those with HIV, less-studied KS subtypes within a European context. However, its retrospective design introduces potential biases, including incomplete data for some variables, such as HHV-8 status. Additionally, the single-center scope may limit the generalizability of findings to other healthcare settings.

CONCLUSION

This retrospective study offers valuable insights into the demographic, clinical, and histopathological features, as well as treatment and outcomes, with an exploratory statistical analysis of KS cases diagnosed over a decade at a tertiary hospital in Lisbon, Portugal. With 113 patients included, it expands knowledge about KS in Portugal, where data on the disease remain limited. Our findings emphasize the importance of early diagnosis, multidisciplinary management, and tailored therapeutic approaches to reduce relapse rates and improve outcomes, especially in immunosuppressed patients.

AUTHOR CONTRIBUTIONS

All authors contributed equally to this manuscript and approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed

according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

FUNDING SOURCES

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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