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CLASSIC KAPOSI SARCOMA: A 20-YEAR RETROSPECTIVE STUDY OF 85 PATIENTS FROM A MOROCCAN DERMATOLOGY DEPARTMENT

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ABSTRACT

Classic Kaposi sarcoma (KS) is a rare, HHV-8-associated angioproliferative disorder that predominantly affects elderly men of Mediterranean descent. Despite its overall indolent course, its epidemiological, clinical and therapeutic profile in North Africa remains incompletely characterized. We conducted a retrospective descriptive study of all patients hospitalized for Kaposi sarcoma in the Dermatology Department of Ibn Sina University Hospital, Rabat, over a 20-year period (2002-2022). Of 85 patients identified, 78 (91.8%) had the classic form, 4 (4.7%) an iatrogenic form and 1 (1.2%) an HIV-related epidemic form. Among classic KS patients, male predominance was marked (sex ratio 5.5), mean age was 69 years, and the median diagnostic delay reached 18 months. Papulo-nodular lesions of the lower limbs were the most frequent presentation (71.8%), and extra-cutaneous involvement was documented in 17.9% of cases at diagnosis, most often gastrointestinal. Using the Krieger staging system, over half of the patients presented with locally aggressive or disseminated disease (stage II-IV). Systemic chemotherapy, chiefly intramuscular bleomycin, achieved stabilization or objective response in the majority of treated patients, and disease-specific mortality was low (3.8%). These findings confirm the generally indolent, treatment-responsive nature of classic Kaposi sarcoma in a Moroccan cohort, while underscoring the value of systematic staging and extended work-up to detect clinically silent visceral involvement.

KEYWORDS

Kaposi sarcoma, HHV-8, classic form, epidemiology, retrospective study, Krieger staging, bleomycin

MAIN ARTICLE

INTRODUCTION:

Kaposi sarcoma (KS) is an angioproliferative disorder of mesenchymal origin that involves both the skin and internal organs, and is causally linked to infection with human herpesvirus 8 (HHV-8) [2]. First described in 1872 by Moritz Kaposi as a multiple pigmented sarcoma of the skin [1], the disease is now recognized under four distinct clinico-epidemiological forms: the classic (Mediterranean) form, affecting elderly men with an indolent course; the endemic form, seen in younger sub-Saharan African patients with a more aggressive behavior; the iatrogenic form, occurring under long-term immunosuppression; and the epidemic form, associated with HIV infection. Although these forms differ in presentation and course, they share common histological features and a constant association with HHV-8 [2]. Morocco is one of the few countries where the classic form remains, by far, the most frequently encountered variant. Through this retrospective study conducted in the Dermatology Department of Ibn Sina University Hospital in Rabat, we aimed to describe the epidemiological, clinical, therapeutic and evolutive characteristics of Kaposi sarcoma in our setting and to compare our findings with data from the literature.

MATERIALS AND METHODS:

We performed a descriptive, retrospective, single-center study spanning 20 years (January 2002-December 2022), including all archived records of patients hospitalized for Kaposi sarcoma in the Dermatology Department of Ibn Sina University Hospital. Inclusion criteria required clinical lesions consistent with KS, a compatible skin biopsy, and adequate work-up to classify the epidemiological form and assess disease extent; HIV-positive (epidemic) cases were analyzed separately. Eighty-five records met these criteria. Data were collected using a standardized case report form covering demographic, clinical, histological, therapeutic and follow-up variables, and were processed using Microsoft Excel. Clinical staging of classic KS followed the Krieger classification, which defines four stages (I to IV) according to lesion type, localized or diffuse distribution, aggressiveness and clinical course [5].

RESULTS:

Of the 85 patients collected, 78 (91.8%) had classic KS, 4 (4.7%) had iatrogenic KS related to prolonged immunosuppressive therapy, and 1 patient (1.2%) had HIV-associated epidemic KS with a CD4 count of 100/µL, who died three years later from severe pulmonary sepsis despite antiretroviral therapy (Figure 1). Given the predominance of the classic form, the following results focus on the 78 classic KS patients.

Epidemiology: Male predominance was marked, with 66 men (85%) and 12 women (15%), giving a sex ratio of 5.5. The mean age was 69 years (range 25-89), with 79% of patients aged between 50 and 80 years. The number of hospitalized cases increased over time, with a maximum of 25 cases recorded between 2018 and 2022. The median diagnostic delay was long, at 18 months (range 1 month to 14 years).

Clinical presentation: Pain and pruritus were the main functional symptoms, reported in 74.4% and 28.2% of patients respectively. Papulo-nodular lesions were the most common presentation (71.8%), followed by angiomatous plaques (64%) and macules (50%); keratotic lesions were seen in 43.6% and ulcerative lesions in 11.5%. Lymphedema was present in 56.4% of patients. Lesions predominantly affected the lower limbs (97.5%, with plantar involvement in 62.8%), followed by the upper limbs (60.2%); a disseminated pattern was seen in 12.8% of patients. Mucosal involvement was documented in 35.9% of patients (oral 27%, genital 16.7%, anal 1.3%), and dermoscopy, performed in 28 patients, most frequently revealed a pinkish-violaceous background (100%), a multicolored pattern (92.9%) and a whitish collarette (57.1%) (Figure 3).

Histology and work-up: All biopsies confirmed the classic triad of vascular proliferation, spindle-cell component and hemosiderin deposits. HHV-8 PCR, performed in 22 patients, was positive in all. Extended work-up revealed extra-cutaneous involvement in 14 patients (17.9%) at diagnosis, most frequently gastrointestinal (n=6), followed by bone (n=5), pulmonary (n=5) and lymph node involvement (n=4).

Staging and treatment: According to the Krieger classification, 27 patients (34.6%) were stage I, 12 (15.4%) stage II, 25 (32.1%) stage III and 14 (17.9%) stage IV (Figure 2). Therapeutic abstention was chosen for 17 stage-I patients; local treatment (surgery, cryotherapy or radiotherapy) was used in 22 patients; and systemic chemotherapy, mainly intramuscular bleomycin, was administered in 49 patients (60.3%), yielding complete or partial clinical response in the majority of cases, with hematologic toxicity as the main

adverse event. Paclitaxel and combination regimens (ABV, vinblastine plus farmorubicin) were reserved for bleomycin failures or advanced stages.

Outcome: At last follow-up (median 18 months), 34.6% of patients had stable disease, 25.6% a partial response and 12.8% complete remission, while 5.1% progressed and 3.8% relapsed. Six patients died (7.7%), only three of which were directly attributable to Kaposi sarcoma (3.8%), mostly related to chemotherapy toxicity or infectious complications; overall survival did not appear to be significantly influenced by classic KS given its generally indolent course.

DISCUSSION:

Our cohort confirms the marked male predominance and elderly onset classically described for Mediterranean KS, with a sex ratio and mean age closely comparable to other large series such as that of Iscovich et al. [3]. The long diagnostic delay observed in our patients likely reflects both the indolent natural history of the disease and limited awareness among patients and primary care providers, reinforcing the need for early dermatological referral. The predominance of lower-limb lesions and the high rate of lymphedema are consistent with the vascular and lymphatic tropism of KS, while the substantial proportion of patients with mucosal or extra-cutaneous involvement at diagnosis (17.9%) argues for systematic screening even in the absence of specific symptoms, since visceral disease can remain clinically silent until advanced stages. The Krieger classification proved practical in stratifying our patients and guiding therapeutic choices, in line with its established role in classic KS management [4]. Systemic bleomycin remained our first-line treatment for advanced or symptomatic disease, achieving response rates comparable to those reported in the literature, while local treatments and taxane- or anthracycline-based regimens were reserved for refractory or advanced presentations, consistent with current therapeutic recommendations for classic KS [5]. The relatively low disease-specific mortality observed in our series supports the overall favorable prognosis of classic KS, although the retrospective design, the high rate of loss to follow-up, and the single-center setting limit the generalizability of our findings and highlight the need for prospective, multicentric studies to better define the epidemiology and optimal management of the disease in Morocco.

CONCLUSION:

This 20-year retrospective study confirms that classic Kaposi sarcoma remains the predominant form of the disease in our setting, typically affecting elderly men with an indolent, HHV-8-associated cutaneous process. Beyond confirming known epidemiological and clinical patterns, our findings emphasize the substantial rate of mucosal and extra-cutaneous involvement at diagnosis, which justifies systematic extended work-up regardless of clinical symptoms. Staging using the Krieger classification remains a practical tool to guide treatment, and systemic bleomycin continues to offer satisfactory disease control with an acceptable safety profile. Prospective, multicentric studies are warranted to refine the epidemiological profile of Kaposi sarcoma across Morocco and to help establish consensus therapeutic protocols.

FIGURES:

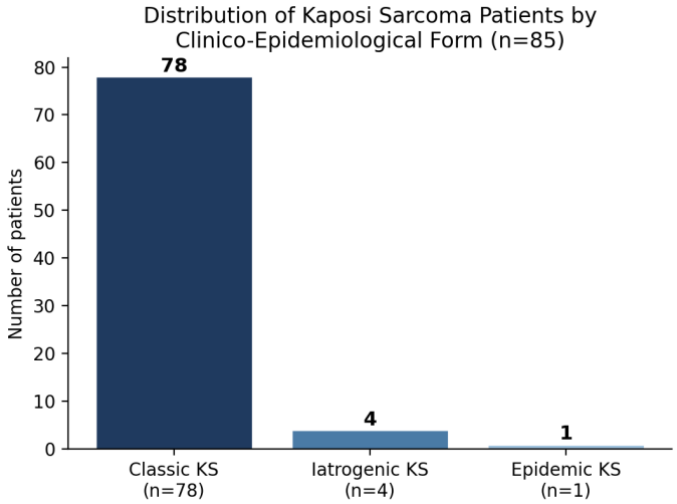

Figure 1: Distribution of Kaposi sarcoma patients according to clinico-epidemiological form (n=85).

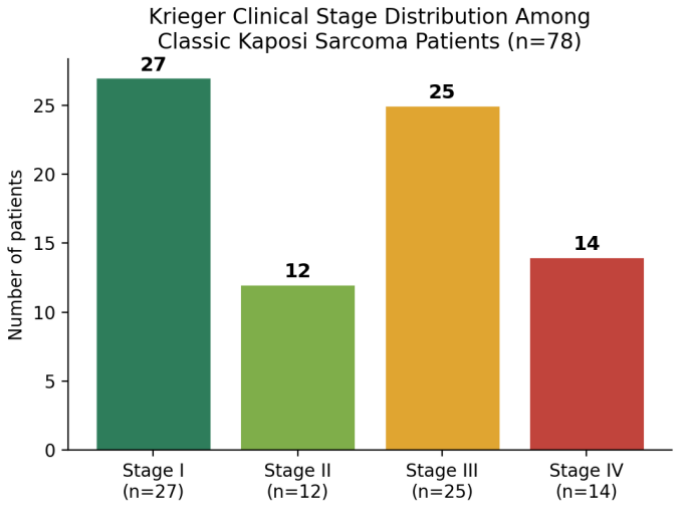

Figure 2: Krieger clinical stage distribution among classic Kaposi sarcoma patients (n=78).

TABLES :

Table 1: Main epidemiological and clinical characteristics of classic Kaposi sarcoma patients (n=78).

Characteristic	Value
Sex ratio (M/F)	5.5
Mean age, years (range)	69 (25-89)
Median diagnostic delay	18 months (1 month-14 years)
Papulo-nodular lesions	71.8%
Lower-limb involvement	97.5%
Mucosal involvement	35.9%
Extra-cutaneous involvement at diagnosis	17.9%
Systemic chemotherapy administered	60.3%
Disease-specific mortality	3.8%

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