A 71-Year-Old HIV-Negative Male Patient with Kaposi Sarcoma: A Case Report and Literature Review

Hamdi Al Shenawi, ACS, FRCSI* Suhair AlSaad, ACS, FRCSI* Fatema Abdulla, MD** Howra Al-Hashim, MD** Noor Al Shenawi***

ABSTRACT

Introduction: Kaposi's sarcoma (KS) is an opportunistic angioproliferative neoplasm associated with Human Herpesvirus 8 (HHV-8). It has four clinical forms: classic, endemic, iatrogenic, and epidemic HIV-associated. In this case report, we present a case of a 71-year-old Egyptian male who presented with a complaint of a pigmented skin lesion involving his finger. Biopsy confirmed the diagnosis of Kaposi sarcoma.

Discussion: A literature review was performed to analyse the already available data. We intended to expand the amplitude of knowledge about Classic Kaposi Sarcoma (CKS) and add to the literature, as we found that the cases reported in the Arabian gulf countries are limited.

Conclusion: To the best of our knowledge this is the first reported case of a non-HIV patient with CKS in Bahrain. This case report highlights the importance of early detection and management of CKS.

Key words: Kaposi sarcoma, non-HIV, Cutaneous, Classic, Mediterranean, Bahrain

INTRODUCTION

Kaposi's sarcoma (KS) is an opportunistic angioproliferative neoplasm associated with Human Herpesvirus 8 (HHV-8). Moritz Kaposi originally described it in the late 1800s. It gained widespread notoriety during the peak of the AIDS epidemic, where it was frequently found co-occurring with opportunistic infections[1]. KS is classified into four clinical forms: The first is the classic form, typically observed in older men aged 60 years or more of Mediterranean or eastern European origin, presenting as lesions confined to distal lower extremities[1]. It is indolent as patients reported to live more than ten years after diagnosis[1]. The second is the endemic form reported in central Africa, affecting men, women, and children[2]. The third is the iatrogenic form observed in patients with long-term immunosuppression, especially after a solid organ transplant[3-5]. Finally, the epidemic HIV-associated form is found to be more common in homosexual men and individuals with multiple sexual partners. Nowadays, the incidence of this form is declining with the use of antiretroviral therapy[6].

Microscopically, KS typically shows vasoproliferative spindle cells with vascular channels or slits[1]. Herein, we present a case of KS in a 71-year-old HIV-negative patient that involved both the upper and lower limbs.

CASE PRESENTATION

A 71-year-old Egyptian male known case of diabetes mellitus and chronic hepatitis C infection, non-smoker, and non-alcoholic. He is married and has six children. The patient presented with a painless, pigmented skin lesion involving the left index finger of three months duration. Excision biopsy was done. The histopathology report showed atypical spindle cell vascular neoplasm suggestive of Kaposi Sarcoma with absent free margins.

The patient then visited our hospital, where another hyperpigmented lesion was found on the left leg. The left index finger showed a hard scar at the site of previous surgery. The rest of the physical examination, including lymph nodes, neurological, abdominal, chest, and heart examination, were all normal. (Figure 1, 2)

Blood labs including complete blood count (CBC), liver function test, renal function test, and coagulation profile were within the normal range. HIV serology was negative and hepatitis profile was positive for hepatitis C.

* Department of Surgery
College of Medicine and Medical Sciences
Arabian Gulf University
Bahrain.
E-mail: hamdims@agu.edu.bh
** Medical Intern
Salmaniya Medical Complex
Bahrain.
*** Undergraduate Medical Student
College of Medicine and Medical Sciences
Arabian Gulf University
Bahrain.

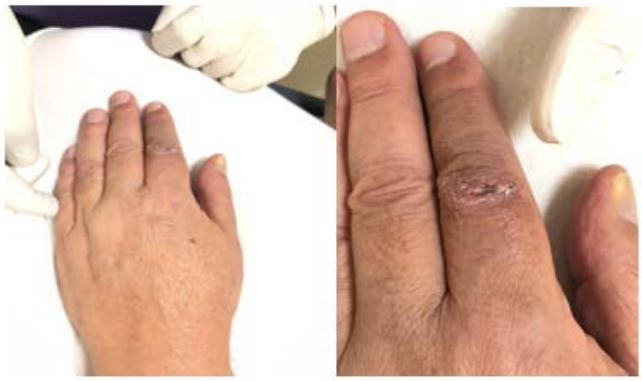

Figure 1. Left index skin lesion after the first surgery in Egypt

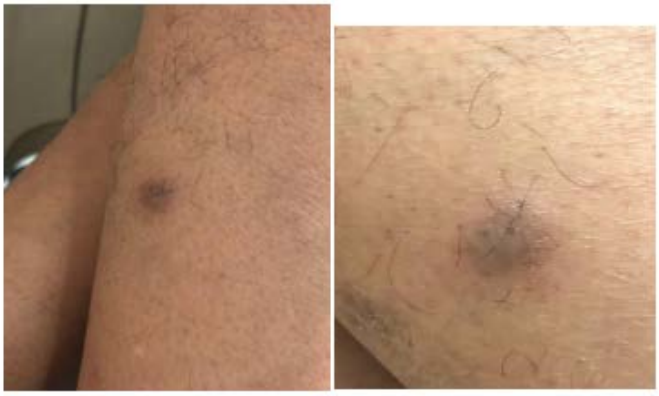

Figure 2. Left leg hyperpigmented skin plaque found on examination

Wide local excision of both lesions was performed. The left index finger was re-excised to ensure safe margins and the left leg hyperpigmented lesion. The biopsies revealed fibroblastic proliferation with a focus of mild atypia in the left index finger and a spindle cell lesion in the left leg. Immunohistochemistry of the specimens was positive for the endothelial cell marker CD31.

Based on the findings above, confirmatory immunohistochemistry for (HHV-8) was positive. A whole-body PET/CT study was done, which relieved no visceral involvement. An oncologist was consulted and decided that no further intervention was needed. Follow-up for the last 3 years with yearly chest and abdomen CT scan, showed no evidence of disease recurrence.

DISCUSSION

Kaposi sarcoma, first described by the Hungarian dermatologist Moritz Kaposi. It is classified into four different clinical patterns: Classic KS (CKS), endemic African KS, iatrogenic KS (immunosuppression-/transplant-associated), and epidemic HIV-associated KS[1]. KS varies widely across the four clinical patterns in incidence across different age groups and its presentation, ranging from small cutaneous lesions to disseminated visceral involvement[7].

The classic type reported in our case was the type described by Moritz Kaposi as idiopathic multiple-pigmented sarcomas of the skin. It has an indolent course with cutaneous involvement, particularly affecting the lower extremities, typically presenting as dark-to-purple macules which progress later to plaques and nodules[8]. CKS mainly affects older men with a mean age of 67 years of Mediterranean or eastern European origin[9].

The definitive diagnosis of CKS is achieved by biopsy and histopathology. Typical histological features include the presence of a vasoproliferative spindle cells which form vascular channels or slits[10]. In addition, the detection of HHV-8 by DNA sequences or immunohistochemical staining can be performed to confirm the diagnosis[11].

The treatment of CKS depends on the clinical presentation and may not be indicated in all cases. For instance, observation without intervention may be an option if the patient has limited asymptomatic cutaneous lesions. However, symptomatic lesions may be treated with either local or systemic tumor-directed therapy, depending on the disease's extent, location, and progression. One of the options for local control is surgical excision which was the chosen method to manage our patient[11]. Generally, the choice of treatment is often based on the experience of the treating physician considering the patient's preferences and comorbidities[11].

In our case, the patient presented with a typical presentation of CKS, a 71-year-old man of Mediterranean ethnic background complaining of darkly pigmented skin lesions involving his upper and lower extremities. According to our knowledge, after extensive research, we did not find any cases reported in Bahrain, making us assume that the cases of CKS are underreported.

Each one of the KS subtypes has a different predominant risk group. In CKS, the main affected population is older males, more than 60 years old, of Mediterranean or eastern European origin. As for the endemic (African) variant of Kaposi sarcoma, the demographics of those commonly affected differ according to age. In adulthood, males are predominantly more affected than endemic Kaposi sarcoma in childhood. Children of both sexes are affected equally. As the name implies, patients are usually of African origin, specifically from equatorial Africa. In the iatrogenic subtype, people at risk are mainly those who are exposed to exogenous immunosuppressants, especially after organ transplant. The risk will increase more in older patients with cyclosporin A[11]. Lastly, the AIDS-associated subtype mainly occurs in homosexual men[11].

The clinical features of Kaposi sarcoma are highly variable across different subtypes. In CKS, the main clinical presentation is purplish, reddish blue macules, plaques, and nodules on the skin, particularly on the lower extremities. The size of these lesions ranges from very small to several centimetres in diameter, and they can remain unchanged for months to years or grow rapidly within a few weeks[11].

The dermatology literature reports 10 different morphologic findings, which are referred to as patch, plaque, nodular, lymphadenopathic (usually in African children), exophytic, infiltrative (the previous two in African adults with endemic KS), ecchymotic, telangiectatic, keloidal, and cavernous or lymphangioma-like variants[11].

CKS is not always limited to the skin. In certain rare cases, it might have extracutaneous manifestations in which the disease might disseminate in the mucous membranes of the mouth and gastrointestinal tract, and regional lymph nodes. Gastrointestinal tract involvement is usually asymptomatic[11]. The disease's progression can also be variable, ranging from a chronic indolent course that barely influences survival to a more aggressive form, which might lead to disability or even death. The indolent course of the disease is the most commonly encountered[11].

Patients in the endemic subtype present similarly to those with CKS, but lesions can be more locally aggressive and involve the lower limbs in lymphedema. Children with the endemic variant of Kaposi sarcoma

Table 1. A literature review of Clinicopathological features of Kaposi sarcoma in non-HIV patients

References	Review of demographic data, clinical and histopathologic features, management, and follow-up
Our case	A 71-year-old Egyptian male, known to have diabetes mellitus and hepatitis C Presented with asymptomatic pigmented skin lesions in his left index finger and left leg Kaposi sarcoma was diagnosed (excision biopsy) Treatment: Wide surgical excision with free margins Follow-up: yearly regular CT scan of chest and abdomen for the last 3 years, showed no recurrence
Jalbert et al. (3)	A 57-year-old American male known to have rheumatoid arthritis on etanercept Presented with multiple telangiectatic lesions on his face and shoulder area and dysphagia of 6 months duration Skin and gastric biopsy diagnosed Kaposi's sarcoma (HHV-8 and CD31 positivity) 4 cycles of systemic chemotherapy due to disseminated disease 5 month follow up showed no recurrence
Yoo et al. (5)	A 70-year-old male on steroids post adrenalectomy due to Cushing syndrome Presented with numerous raised non-blanching purple plaques on both lower extremities Kaposi sarcoma was diagnosed (CD34, CD31, and HHV-8 positivity) Withdrawal of steroid therapy, led to complete regression of skin lesions yet hyperpigmentation remained.
Jan et al. (7)	A 45-year-old Indian female Presented with erythematous violaceous macules over both lower limbs and left index finger. First skin biopsy showed vasculitis. Treated with oral prednisolone, cyclophosphamide and IV methylprednisolone, no response Repeat skin biopsy was suggestive of Kaposi sarcoma Developed hospital-acquired pneumonia and died.
Gupta et al. (9)	A 55-year-old Dominican male Presented with pigmented nodular lesions on lower extremities and soft palate of two year duration Kaposi sarcoma was diagnosed Systemic chemotherapy due to presence of disseminated disease. At 5-month follow-up, skin lesions were noted to be markedly smaller in size.
Meriem et al. (18)	A 78-year-old Moroccan female, known case of diabetes mellitus Presented with pain and discomfort at mastication, ecchymoses and petechiae scattered on the right cheekbone, nodule on the right leg, and pedunculated nodule on the dorsal side of the tongue Kaposi sarcoma was diagnosed (HHV-8 positivity) Dermatologist decided to monitor
Sethia et al. (19)	A 28-year-old male known to have hepatitis B Presented with 3-month history of swelling of the right upper and lower gingiva and right palate. Kaposi sarcoma was diagnosed (CD31, CD34, and HHV-8 positivity) Primary radiotherapy (RT) with 25 fractions over 5 weeks with complete response No recurrence with a follow-up period of 16 months
Nemani et al. (20)	A 21-year-old male Presented with wound in the left ankle and foot for the past 5 months after trauma which progressed to an ulcer. Kaposi Sarcoma was diagnosed Below knee amputation with prosthesis No recurrence after follow up for one and half years.
Grigoriou et al. (21)	A 57-year-old Greek female Presented with extensive discolored skin lesion on both legs. Kaposi sarcoma was diagnosed Oncology Department for further management. Died of acute myocardial infarction 2 months after diagnosis
Tutaeva et al. (22)	35-year-old Caucasian man known to have Hodgkin's lymphoma and is seropositive for EBV, HHV-8 and HHV-6 Presented with upper abdominal pain and episodes of black tarry stools associated with weight loss. Pigmented nodules on the right palm, fourth finger of the right hand and left foot, and multiple small (2–3 mm) round nodules in the subcutaneous fatty tissue of the upper and lower extremities. Kaposi sarcoma was diagnosed (CD34, PDPN, and Anti-HHV-8 antibodies positivity) He received a total of five cycles of ABVD (chemotherapy regimen) Complete remission 19 months after the last ABVD course.
Kim et al. (23)	A 79-year-old female known to have diabetes mellitus and hypertension Presented with pigmented like masses on both upper and lower extremities of 2 years duration Kaposi sarcoma was diagnosed (HHV-8, CD34, CD32, and D2–40 positivity) Treated with palliative radiotherapy with good response Follow up no recurrence

Bayoumi et al. (24)	A 60-year- old Yamani male known to have hypertension and ischemic heart disease Presented with painful scrotal lump with dysuria of 3 weeks duration Kaposi sarcoma was diagnosed 3 cycles of systemic chemotherapy every 3 weeks Follow-up 3 months with CT abdomen and pelvis, no recurrence
Kusenbach et al. (25)	A 3-year-old German boy Presented with small reddish macule behind his right ear Kaposi sarcoma was diagnosed Chemotherapy resulted in temporary improvement, but later tumor progressed rapidly despite chemotherapy Immunological evaluation revealed severe immunocompromise Bone marrow transplant was done leading to complete remission Follow up for 18 months showed no recurrence
Que et al. (26)	An 83-year-old Hispanic female known to have hypertension, atrial fibrillation, and chronic venous insufficiency Presented with painful violaceous eruption on the lower legs of 3-year duration Kaposi sarcoma was diagnosed (HHV-8 positivity)
	67-year-old Hispanic male known to have diabetes mellitus and chronic venous insufficiency Presented with multiple hyperpigmented plaques on distal aspect of legs of 1 year duration Kaposi sarcoma was diagnosed
Guevara et al. (27)	A 48-year-old male Presented with purple plaque over the glans penis near the urethral meatus measuring around 1cm. Associated with painful erection Kaposi sarcoma was diagnosed (CD31, CD34, and HHV-8 positivity) Conservative treatment
Swali et al. (28)	A 66-year-old Hispanic male Presented with pigmented skin lesions in the right foot of 3 months duration Kaposi sarcoma was diagnosed (HHV-8 positivity) Dermatologist prescribed imiquimod 5% and tretinoin 0.05% cream to be applied twice daily to the affected areas
Etesami et al. (29)	A 43-year-old male Presented with multiple erythematous dome-shaped papules on his right auricle of 6 months duration Kaposi sarcoma was diagnosed Total surgical excision with recurrence after 4 years, followed by a second recurrence after 2 years.
Bedier et al. (30)	An 88-year-old Canadian male Presented with scaly erythematous plaques, violaceous subcutaneous nodules on his palms and lower extremities. Patient had weight loss, night sweats, and fever for 4 months. Kaposi sarcoma was diagnosed (HHV-8 positivity) Oral retinoid with topical hydrocortisone and ultraviolet B (UVB) phototherapy, good response
Bortoluzzi et al. (31)	63-year-old Brazilian female Presented with asymptomatic increase of soft palate volume with occasional bleeding on examination there was an ulcerated soft nodular red- purplish lesion Kaposi sarcoma was diagnosed (HHV8, CD31 and CD34 positivity) The intraoral lesion was removed by conservative oral surgery Regular follow up revealed no recurrence 17 months after surgery
Mohanna et al. (32)	71-year-old healthy female Presented with progressive, painless, red papule on the hard palate Kaposi sarcoma was diagnosed Chemotherapy was given Follow up for 5 years, no recurrence
	46-year-old healthy male Presented with growing mass in the groin of 3 month duration Kaposi sarcoma was diagnosed (biopsy of the mass) Surgical excision of the tumor 1 year recurrence with cutaneous nodules on the scalp, lip, and leg Treated with chemotherapy and anti-TB medication for newly diagnosed pulmonary TB but died in few months
Kodra et al. (33)	77-year-old Hispanic male Presented with 6-weeks of dyspnea on exertion, cough with sputum, hemoptysis, fatigue, and weight loss Examination showed multiple pigmented lesions in the lower extremities Skin and bronchoscopic biopsy diagnosed Kaposi sarcoma (HHV-8 positivity) Patient refused chemotherapy and was discharged with hospice
Rodríguez et al. (34)	77-year-old healthy female Presented with firm, reddish slow growing nodules on the anterior helix of the right ear pinna. Kaposi sarcoma was diagnosed (CD31, CD34 and HHV-8 positivity) Surgical excision and no recurrence in 2 years of follow up

Al-Kzayer et al. (35)	A 6-year-old Iraqi male, with cerebral palsy, mental retardation and epilepsy, on valproic acid Presented with painless, pigmented plaque on his left foot, developed post respiratory infection. After 1 year, lesions appeared at his groin, right axilla, and face and cervical lymphedema Kaposi sarcoma was diagnosed (HHV-8, CD34, and CD 31 positivity) Family refused hospitalization After 1 year of his first presentation, patient died due to sudden respiratory distress
Crosetti et al. (36)	A 48-year-old Caucasian male Presented with sensation of foreign body in throat with stomatolalia. Examination revealed large, bulky, mobile red-purple lesion at the base of his tongue Kaposi sarcoma was diagnosed Direct micro laryngoscopy and carbon dioxide laser excision was done Regular follow-up, no recurrence
Zhou et al. (37)	A 63-year-old Miao (Southwest China) male Presented with longstanding pigmented nodules in his upper left eyelid, bilateral auricle for 9 year duration, and upper and lower limbs for 37 year duration Kaposi sarcoma was diagnosed Not given any medical treatment Regular follow-up showed spontaneous partial regression of KS
Touzani et al. (38)	A 72-year-old healthy male Presented with gradual onset of tumor-like granulation tissue at the level of glans penis of 3 year duration Initial biopsy was negative for Kaposi sarcoma, the second diagnosed Kaposi sarcoma Treated with chemotherapy
Makharoblidze et al. (39)	A 45-year-old female Presented with symptoms of pericardial effusion and cardiac tamponade. CT coronary angiography and Transesophageal echocardiography showed a tumor in right atrium auricle extending toward the superior vena cava Kaposi sarcoma was diagnosed Palliative surgical excision done due to the anatomic position of the tumor Patient remained asymptomatic on 10 months follow up
Naimi et al. (40)	A 77-year-old Tunisian man, known to have hypertension, diabetes, and smoker Presented with dysphagia to solids and hoarseness of voice for 4 months and multiple purple macules on the left upper arm. Laryngoscopy showed massive purplish nodule arising from the upper laryngeal wall and infiltrating the vocal cords. Laryngeal kaposi sarcoma was diagnosed (HHV-8, CD34, CD31 positivity) 3D conformal radiotherapy was done Patient died from pulmonary embolism 3 months after radiotherapy.
Seleit et al. (41)	A 42-year-old male Presented with gradually enlarging, pigmented firm facial nodule, surrounded by erythematous halo in the right cheek, of 4 week duration Kaposi sarcoma was diagnosed (HHV-8 positivity) Treated with surgical excision Follow-up for 6 months, no recurrence
	A 32-year-old circumcised male Presented with pigmented penile nodule of 6 month duration Kaposi sarcoma was diagnosed (CD34, and HHV-8 positivity) Treated with surgical excision Follow-up for 6 months, no recurrence
Ozmen et al. (42)	A 71-year-old male, known to have lung carcinoma Presented with 3 pigmented lesions on the scrotum of 5 years duration Kaposi sarcoma was diagnosed Surgical excision was done
Keleş et al. (43)	A 72-year-old female Presented with swelling on her right tonsil of 3 months duration with cervical lymphadenopathy and extension to hypopharynx Kaposi sarcoma was diagnosed (CD34, CD31 and HHV-8 positivity) Right tonsillectomy was preformed, followed by chemotherapy No recurrence after one-year of follow-up

Bojko et al. (44)	A 79-year-old African American male, known to have prostate cancer, Presented for follow up of previously diagnosed KS on his left big toe. Pitting edema over dorsal surface of both feet, with multiple hyperpigmented patches Kaposi sarcoma was diagnosed (CD31, CD34 and HHV-8 positivity) Treated with imiquimod cream, cryotherapy with liquid nitrogen and topical hydrocortisone cream over a course of 1 year but without resolution. Given a trial of alitretinoin gel for 14 weeks, showed improvement Complete resolution three years after his first presentation
	A 47-year-old African American male, bisexual, obese, known to have genital herpes Presented with painful lesion on his feet of 10 months duration Kaposi sarcoma was diagnosed (CD31, CD34, vimentin and HHV-8 positivity) Failed trial of cryotherapy & topical alitretinoin for several weeks. Residual lesion was excised by plastic surgery, with no recurrence on follow up
Faden et al. (45)	An 80-year-old Saudi male, known to have hypertension and diabetes, and previous history of Coronary Artery Bypass Graft 8 years ago Presented with multiple pigmented lesions at the junction of hard and soft palate, buccal mucosa and alveolar ridge Kaposi sarcoma was diagnosed (CD31, D2-40, and HHV-8 positivity) Treated with radiotherapy 30 doses Follow up for 4 years, showed no recurrence
Alamri et al. (46)	A 63-year-old male Presented with small, pruritic, slightly thickened brown lesion on glans penis on the ventral aspect of 8 months duration. Kaposi sarcoma was diagnosed (CD34, CD31 and HHV-8 positivity) Treated with excisional biopsy Follow up for 3 years, no recurrence
Alghanim et al. (47)	A 73-year-old Saudi male, known to have dermatomyositis on azathioprine and prednisolone Presented with multiple pigmented lesions on his face, neck, chest, upper back and upper limbs after one month of immunosuppression treatment Kaposi sarcoma was diagnosed Azathioprine and prednisolone was discontinued, but he continued to develop more skin lesions. Treated with chemotherapy At one year follow-up, no recurrence
Baazeem et al. (48)	A 59-year-old Yemeni male, known to have hypertension and dyslipidemia Presented with a firm, non-tender, spherical nodule in the glans penis Kaposi sarcoma was diagnosed Surgical excision of the lesion 18-month follow up showed no recurrence
Gunawardena et al. (49)	24-year-old Saudi male, on prednisolone and cyclosporine post renal transplant Presented with low grade fever, small right-sided pleural effusion with fine reticulonodular shadowing in both lungs, abdominal pain, and melena Transbronchial and gastric biopsy confirmed Kaposi sarcoma Stopped immunosuppression therapy Complete resolution with no recurrence but kidney function deteriorated and started on hemodialysis
	27-year-old Sudani male who is on prednisolone and cyclosporine Presented with general body ache, dry cough, epigastric pain, and melena CT scan showed a mass in the right lung lobe Lung biopsy showed Kaposi sarcoma Stopped immunosuppression therapy Complete resolution of Kaposi sarcoma with no recurrence, but on hemodialysis

are more likely to have systemic involvement, including lymph nodes and visceral involvement. Clinically the course of the disease wildly varies from indolent to aggressive visceral involvement. The aggressive forms are more commonly observed in children[11].

Iatrogenic KS usually has a similar presentation to the classic type, but its course may be more disseminated and aggressive if immunosuppressive therapy is not tapered or discontinued[12].

In HIV-associated KS, the clinical features varied among patients ranging from a localized minimal mucocutaneous disease to a disseminated visceral involvement. Skin lesions occur in nearly all patients, usually multiple, mainly affecting the lower extremities, head, and neck. It can be associated with lymphedema, typically involving the face and lower extremities. The visceral involvement is relatively common in this subtype. According to autopsy reports of these patients, every organ has been affected except the brain[13].

Biopsy is the definitive method to diagnose KS, and the microscopic features of all four different types of KS do not differ. All forms show evidence of angiogenesis, inflammation, and spindle cell proliferation[11].

The mononuclear cell infiltrate is no longer prominent, and few extravasated erythrocytes and macrophages are present between spindle

cells[11]. The lining cells of the clearly developed vascular structures are positive for vascular markers (such as factor VIII). In contrast, the spindle cells consistently stain for CD34 and commonly CD31 but are factor VIII negative[11]. In addition to observing the typical histologic features on standard microscopy, a polymerase chain reaction can be performed on the skin lesions to detect amplified human herpesvirus 8 (HHV-8) DNA sequences within the spindle cells, thus confirming the diagnosis[11].

Radiographic evaluation is not usually warranted in cases of asymptomatic CKS limited to the skin and should not be done based on the severity of the skin involvement. Even radiological imaging has low yield if the gastrointestinal system is suspected to be involved[11]. An endoscopic evaluation of the gastrointestinal mucosa is the golden standard if the gastrointestinal system is suspected to be involved[11].

Considering the absence of treatment to eradicate HHV-8 infection, usually, the goal of managing KS is not curative but rather to palliate the symptoms, shrinking the size of cutaneous or visceral lesions and slowing the progression of the disease. Choosing the best applicable treatment option for each patient depends on several factors: the subtype of KS, the extent of the disease, the patient's age, and comorbidities.

In CKS, in asymptomatic patients, observation only may be sufficient. If the lesion is resectable in symptomatic patients, simple excision can be the choice. Otherwise, if not resectable, radiotherapy may be considered. Systemic chemotherapy is the last resort, usually preserved for extensive or refractory disease. Brenner et al. retrospectively analysed the data of 123 patients, observation was the main approach in 39 patients, and they remained progression-free for a median of 4 months. 29 patients underwent surgical resection and remained recurrence-free for a median of 15 months[14].

In Iatrogenic KS, immunosuppression progressive tapering or withdrawal is considered the cornerstone of the management and, if possible, switch to the mammalian target of rapamycin (mTOR) inhibitors, such as sirolimus and everolimus in post-transplant KS[13]. mTOR acts as an antiproliferative with an immunosuppressive effect. Krengel and his colleagues reported an 80-year-old male who developed iatrogenic KS of the skin after receiving immunosuppressive medications to treat his myasthenia gravis. Switching to everolimus resulted in almost complete disappearance of the skin lesions, with a remission lasting for 9 months[15].

The options used in the management of HIV-associated Kaposi sarcoma include HAART alone, local therapy, immunotherapy, cytotoxic therapy, and molecular targeted therapy. All HIV-associated Kaposi sarcoma patients must receive HAART as it is the mainstay of treatment in HIV and has been reported to aid in the regression of an existing Kaposi sarcoma and even the possibility of a complete cure. Local therapeutic options are mainly indicated for local disease, including surgery, cryotherapy, and radiation, among other methods. As for patients with visceral involvement, the combination of HAART and single or multiple chemotherapeutic agents may be used with discontinuation of chemotherapy after disease regression[16].

In the endemic subtype, the treatment also is similar to the CKS. In a retrospective study by Stein M.E. et al. of 47 African patients diagnosed with the endemic variant. 29 patients who presented with a local disease were treated with local radiotherapy, and 17 patients were treated with chemotherapy. The response rate in these patients was >80%, regardless of the chosen treatment modality. Therefore, they concluded that the endemic subtype is chemo- and radiosensitive[17].

Table 1 summarises 42 case reports (including the present case), non-HIV KS, including demographic, clinical, histopathological features, management, and follow-up. The literature review was performed by searching electronic databases and using the search terms "Kaposi sarcoma, non-HIV, HIV-negative."

CONCLUSION

Kaposi sarcoma is a rare disease as this is the first reported case of CKS in non-HIV patient in Bahrain. The diagnosis of Kaposi sarcoma is established by unique histopathological features. Treatment widely varies from conservative treatment, surgical excision, and systemic tumor-directed therapy. This case report highlights the importance of early detection and management of CKS.

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Availability of Data and Materials: The original contributions presented in this study are included in the article/supplementary material, further inquiries can be directed to the corresponding authors.

Ethical Considerations: The studies involving human participants were reviewed and approved by the Research and Ethics Committee (REC) College of Medicine and Medical Sciences, Arabian Gulf University. The patient/participant provided his written informed consent to participate in this study. Written informed consent was obtained from the individual for the publication of any potentially identifiable images or data included in this article.
Potential Conflict of Interest: None

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