Kaposi’s Sarcoma Not Related To HIV: Case Report

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Abstract

Introduction: Kaposi’s sarcoma is an angio proliferative disorder related to a malignant neoplasm predominantly of mucocutaneous tissues. There are four clinical presentations of Kaposi’s sarcoma: Classic, Endemic, Epidemic or related to HIV and iatrogenic, all of which are mainly caused by human herpes virus type 8.

Objective: We present a case of a 73-year-old male, type II diabetic, with a family history of cutaneous carcinoma, diagnosed with non-HIV related Kaposi’s sarcoma.

Discussion: Kaposi’s sarcoma is mainly associated with the herpes virus 8, this seems to be necessary for the development of the disease. We present a case of classical Kaposi’s sarcoma, commonly affecting people older than 60 years. It is characterized by violaceous red maculæ in lower limbs progressing to nodules. The health care provider must be able to make the diagnosis based on skin lesions and perform a biopsy only to confirm histological diagnosis. The 5-year survival for low eruptivity 97%, moderate 92.7% and high 75%.

Conclusion: We found a classic Kaposi sarcoma with no relationship to HIV infection, a case not well documented, with palliative management and with a good response to it, since there are not established management protocols available.

Introduction

Kaposi’s sarcoma is an angio proliferativo multi focal disorder related to a malignant neoplasm that affects mostly mucocutaneous tissues of the organism.¹ ² There are three types of dermatological lesions that manifest maculæ at the beginning of the disease, plaques that indicate intermediate progression, and the presence of palpable nodules in more advanced stages.³ ⁴

Epidemiologically, there are four clinical presentations of Kaposi’s sarcoma: Classic, Endemic, Epidemic, and Iatrogenic,¹ all of them have in common the participation of the human herpes virus type 8 (VHH-8), currently recognized as the principal cause of all the clinical presentations.

Kaposi’s sarcoma classic is considered de most common form of this disease, defined as a malignant neoplasm of spindle cells of endothelial origin that affects primarily white men over 60 years old, clinically characterized by painless skin lesions in the form of red-brown or violaceous maculæ located in: head, neck, hands,and feet; on the other hand it can also manifest as painless lesions in the form of papules that are located in upper and lower limbs being this the most common site of appearance.⁵ ⁹

Endemic Kaposi’s sarcoma is characterized by generalized lymphadenopathy, with aggressive clinical behavior, having a survival of two years after diagnosis.¹⁰ Epidemiologically it manifests in two forms, the first one develops in middle-aged adults among the ages of 25-50 years and the second subtype is triggered in children under 10 years old.

Epidemic Kaposi’s sarcoma or associated with the human immunodeficiency virus (HIV), develops in homosexual or bisexual population due to unprotected anal intercourse.¹¹ ¹² This type of sarcoma is characterized clinically by the presentation of small painful cutaneous lesions¹³ without the presence of pruritus, mainly affecting the external genitalia¹⁴ but can also develop on the face, trunk, and extremities. If it manifest itself in the oral cavity it would appear as violet-colored maculæ.¹³ Iatrogenic Kaposi’s sarcoma is related to the use of immunosuppressive therapy in patients who have received some type of organ transplant or who have an immunosuppressive treatment for an autoimmune disease.¹⁵

Two percent of the cases resume spontaneously, most have a slow progression that presents itself as isolated lesions than may disappear and appear in other sites in a painful tumor form that the slightest trauma can ulcerate. When it comes to long evolutions there are edemas secondary to venous stasis and lymphatic obstruction due to the stimuli of endothelial growth factors.¹⁶

The majority of these patients are prone to recurrent opportunistic infections such as esophageal candidiasis, toxoplasmosis, pulmonary tuberculosis and hepatitis B, with the risk of relapsing into a Burkitt lymphoma-type neoplasm. Herpes virus infections associated with Kaposi’s sarcoma (KSHV) are predominantly latent, during this phase the viral genome replicates and secretes nuclear antigens

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associated with latency (LANA) a protein that allows it to persist as a circular multicopy and as extra chromosomal episome in all cellular division in the cell nuclei.17,18 The LANA can be expressed as LANA/N-terminal associated to mitotic chromosomes through the junction of histone H2A/H2B, or as LANA-C-terminal which can be connected simultaneously to the terminal repetition of the DNA, 19–30 as viral genome of the LANA lacks enzymatic functioning it must acquire chromosomes in order to distribute its viral DNA during mitosis.21,31–33

As treatment goes, for those patients with asymptomatic lesions it is considered only vigilance, in those with multiple nodules and maculae of short evolution it is recommended the use of radiotherapy for being radiosensitive and its percentage of efficacy (63-85%), and in cases where there are mucocutaneous disseminated lesions bigger than 1 cm of diameter it is opted for chemotherapy using bleomycin, etoposide and vinblastine, the latter presents a response in the majority of the cases (60-80%).

Case presentation

It is a male patient of 73 years of age, married, mestizo, and farmer, with a family history of cutaneous carcinoma, allergic to shellfish, a former alcohol consumer, non-smoker, and with type 2 diabetes mellitus.

Patient presents itself in August of 2013 due to intermittent edema in his left hand, with inflammatory changes, since October of 2012, the patient reports that the edema became persistent since December of 2012 accompanied with changes in coloration for what he visited a vascular specialist that referred him to an dermatologist. The dermatologist found lesions on the forehead of the face, on the left hand and foot, so a skin biopsy was performed obtaining positive results for Kaposi’s sarcoma on hands and feet with two HIV negative serologies.

Initially the patient was managed on an outpatient basis, presenting a physical limitation in the left hand for the presence of the edema, due to the purple skin lesions suffered a glove-like epidermal thickening extended to the elbow and both lower limbs accompanied of multiple hard 1 cm diameter nodules; presenting other purple macular lesions of 3cm of diameter dispersed in thorax and the left thigh, no metastatic lesions were found in liver, lung or brain.

He was given palliative treatment with chemotherapy based on paclitaxel 300mg/day and cisplatin 100mg/day starting in August and completing six cycles in December of 2013, during this period the patient presented peripheral hypostasis in the palms, so he received treatment with pregabalin 75 mg/m^2. With the therapeutic management received the thoracic lesions, edema, and nodules in the extremities disappeared. The patient presented a recurrence in his left hand becoming infected, a culture was performed isolating Staphylococcus for which he has given levofloxacin 150mg a day for 7 days.

The patient was kept under strict observation, due to manifestation of epidermal thickening and dryness in the lower extremities, but without the presence of purple lesions, the diameter of his left hand was reduced 60%. All of his soft tissue was tensed and with the persistence of the edema, deformities in his finger began to appear, presenting osteomyelitis in the carpi and distal phalanges.

In May of 2015 the edema in the left hand increased, antibiotic coverage was changed to ampicillin/sulbactam to which he did not show improvement, an interconsultation was made with a dermatologist for new treatment and it was decided to perform an infracondyl aramputation in June of 2105 due to lack of response to treatment.

A year later in May of 2016, new sarcomatoid lesions presented in the left arm and leg which were treated with liposomal doxurrubicin 20 mg/m^3. Currently the patient is in good general condition, without cutaneous exacerbations, and has a good response to treatment.

Discussion

Kaposi’s sarcoma is a malignant neoplasm, described by Moritz Kaposi in the years 1872, whose main affection is the mucocutaneous tissues,1,2 it has as a causative, agent the human herpes virus type 8, suggesting the use of the name of herpes virus associated with Kaposi’s sarcoma.1,34 The presence of the virus seems to be necessary but not enough for the development of the disease, since a high seropositivity of VHH-8 has been observed in certain geographical areas that would not necessarily be related to an incidence of Kaposi’s sarcoma.

In this opportunity it is presented a classic Kaposi’s sarcoma, a vascular neoplasm of indolent course that mostly affects men older than 60 years of age, whose clinical features are the presence of red-violaceous macula in the lower limbs with a slow evolution that can lead to the formation of plaques, nodules and/or tumors which can acquire a warty appearance that later become ulcerous formations. Histologically, a moderate nuclear atypia can be observed with the presence of mitosis and vascular necrosis with deposits of hemosiderin phagocyted by sickle cells.

The health provider should be able to correlate a diagnosis based on the cutaneous lesions characteristic of the disease and needs the performance of a biopsy to confirm the histological diagnosis. The staging of the disease (low, moderate or high) can be performed with simply observing the clinical characteristics and the number of isolated lesions per year, all this being important to provide an adequate prognosis of the disease to the patient. According to the staging of the disease, the survival at 5 years is 97%, 92, 7%, and 75% for those with a low, moderate, and high stage.35

Conclusion

It is concluded that the descriptions of different literatures coincide with our reported case, where we found a classic Kaposi’s sarcoma, not related to HIV, this being an unusual case reported; where a previously described palliative care was given since there are no protocols or established management for these patients and as it was observed the patient had a good response to the therapeutic that was provided.
References


