Non-HIV Associated Kaposi's Sarcoma: A case report

Sarcoma de Kaposi no asociado a VIH: un caso clínico

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ABSTRACT

Kaposi sarcoma (KS), first described in 1872, is an angioproliferative neoplasm that often presents with red-purple macules in the skin. This report is of a case of classic/iatrogenic form of KS in a 79-year-old male, that had a prolonged hospitalization due to surgical complications. After discharge, he presented a red-purple macule. A biopsy was made and KS was confirmed. He was HIV negative. The patient did not require any other treatment asides from the total removal of the lesion. Kaposi's sarcoma is an uncommon disease, still very associated with HIV. This case demonstrates the importance of recognition of KS in non-HIV patients. There are four types of Kaposi's sarcoma and the importance of its recognition in non-HIV patients.

Keywords: Kaposi sarcoma; HIV-negative; classic; macule; biopsy.

INTRODUCTION

Kaposi's sarcoma (KS) was first described by a Hungarian dermatologist, Moritz Kaposi, in 1872[1,2]. KS is an angioproliferative neoplasm that often presents with red-purple macules in the skin. There are four types of categories that these patients can be divided into Classic KS, epidemic KS, iatrogenic KS and endemic KS[3]. The classic KS is often associated with elderly men, normally with a Mediterranean and Eastern European heritage. The epidemic KS is related to HIV-positive patients. The endemic is also called de African endemic form and usually occurs in children and adults of sub-Saharan Africa[4]. Iatrogenic KS is seen in immunosuppressive drug therapy, including transplant patients. Since 1981 the majority of KS has been seen in HIV-positive patients[4], but with the evolution of medical therapies, like immunosuppression drugs, physicians must be mindful that this rare neoplasm can appear more often in non-HIV patients. The classic variant normally has an indolent course and presents with skin lesions that affect more often the lower limbs and feet (85-98%)[5]. We present a case of a 79-year-old man with a classic/iatrogenic KS.

CASE DESCRIPTION

We report the case of a 79-year-old man, that was referred to an Internal Medicine consult by his surgeon, for treatment of oesophagal candidiasis. The patient had a medical history of partial gastrectomy over 25 years. He wasn't medicated for any other conditions. When asked, he admitted that he had extra-conjugal relations until 1999, when he was abroad in France and Mozambique. Four months before the consult, the patient was submitted to a laparoscopy cholecystectomy that was complicated by a sub-hepatic and peri-hepatic hemoperitoneum. He had a prolonged hospitalization and received multiple courses of antibiotics. A month after being discharged, he went to his surgeon complaining of odynophagia. At the time, he was submitted to an upper digestive endoscopy that showed white mucosal plaques adherent to the mucosa of the oesophagus. A biopsy was performed and confirmed the diagnosis of candidiasis. Around this time, the patient was already being followed in the Internal Medicine consult and was medicated with fluconazole 400 mg per day for 14 days. At the physical examination, he mentioned a small purple macula in his right knee. There were no lymphadenopathies or other changes in the physical exam.

A biopsy of the knee lesion was made and confirmed the diagnosis of Kaposi's sarcoma. The neoplasm was fully removed at the time of the biopsy. For the investigation of Kaposi's sarcoma and oesophagal candidiasis, an HIV test was ordered and came back negative. Considering these clinical features another test was made, at the same time other entities that could cause an immunosuppression state were sought.

The second serologic test for HIV also came negative. All other laboratory findings were in the normal range. The CD4 count was within the normal range with a CD4/CD8 ratio of 1.08. A pan-tomography was performed and showed no enlarged lymph nodes or other images suggestive of cancer. The patient underwent another upper and total colonoscopy, to obtain biopsies of the intestinal mucosa, looking specifically for Kaposi sarcoma. No signs of KS were found either through direct visualization or in the biopsies made. The oesophagal candidiasis was much better.

The patient didn't need any other treatment, apart from the removal of the single lesion of the KS.

At 3-month and 6-month follow up, no more skin lesions were noted. He remains asymptomatic up to today, with his scans and analytical studies within normal ranges.

DISCUSSION

Kaposi's sarcoma can be classified into four categories: classic, endemic, iatrogenic and AIDS-associated. Non-AIDS KS is considered a very rare disease[3]. The classic form is more frequent in an elderly male with Mediterranean heritage. A male-to-female ratio of 15:1 has been reported[3].

KS classically presents with red-purple macules in the skin, which affects more the lower limbs with the classic form and rarely presents a visceral involvement[2]. The iatrogenic form presentation is very similar to the classic form[8]. The definitive diagnosis is made with a skin biopsy and histopathology.

The course of the classic form is indolent[3,6] and has a good prognosis with no significant impact on the survival rate. In case of recurrence, local cryotherapy usually resolves the isolated lesions.

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Other therapies are available, when the KS manifest with multiple lesions, like intralesional chemotherapy with vincristine[7].

The iatrogenic form normally resolves when the immunosuppressive medications are stopped or decreased. In this case, these risks/benefits have to be balanced.

Management of KS depends on primary aetiology. Nowadays with active antiretroviral therapy (HAART), HIV patients have decrease rates of KS[6].

In our case, the patient had two simultaneous diseases that are often considered opportunistic. Extensive etiologic exams were performed, including two HIV tests and CD-cell count. The patient remained asymptomatic and didn't need any other treatment, apart from the anti-fungic and the removal of the KS. The local recurrence is rare after a complete excision[2].

Considering that the patient had a long-term hospitalization due to surgical complications with multiple courses of antibiotics, this was considered to be the cause for his immunocompromised state and the cause for both esophagi candidiasis and KS. He also fitted the classic form, according to age, heritage and site of the lesion.

Kaposi's sarcoma is an uncommon disease, still very associated with HIV. It is a very important differential diagnosis in skin lesions of HIV patients. With this case, we reinforced the idea, that there are four types of KS and the importance of its recognition in non-HIV patients. The skin biopsy is very important for the diagnosis and accurate treatment.

CONFLICT OF INTEREST

The authors declare that there is no potential conflict of interest relevant to this article.

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AUTHORS' CONTRIBUTION

Costa, Raquel and Mendes, Tiago wrote de paper; Fontes, Joana, Sousa, Barbara and Silva, Joana reviewed the paper.

ETHICAL ASPECTS

All participants submitted a consent form to be included in this study.

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