Anaplastic Kaposi is a rare form of vascular tumor, clinically notable for its high local aggressiveness, propensity for deep invasion and increased metastatic capacity. This disease arises in patients with a history of Kaposi and because of its rarity, few clinical data are available.

We report the case of an 84-year-old patient admitted for the management of Kaposi's disease. The dermatological examination revealed the presence of a burgeoning ulcerocutaneous tumor affecting the entire dorsal surface of the left foot associated with necrosis of the four right toes and erythematous patches in the legs, thighs and trunk and involvement of the oral and genital mucosa. A skin biopsy confirmed the diagnosis of Kaposi and the assessment did not reveal the presence of other visceral locations. Chemotherapy treatment and amputation were proposed, but the evolution was marked by the death of the patient.

Keywords: Anaplastic; Chemotherapy; Treatment.

Introduction

Classic Kaposi is a human herpes virus 8 (HHV8) - associated skin vascular tumor that is rarely considered an actual cause of death, because of its low-grade malignant potential. Only two studies, comprising respectively 5 [1] and 8 [2] patients concerned anaplastic KS seen in areas endemic for classic KS.

We report a case of HIV-negative anaplastic kaposi in a 84-year-old.

Case Report

This is an 84-year-old patient admitted to our training in 2016 for the management of erythematous angiomatous plaques in the left foot. On the dermatological examination, it was noted the presence of infiltrated erythematous plaques sitting on the dorsal surface of the left foot. The rest of the somatic examination was normal. A cutaneous biopsy was requested but the patient was lost to follow-up until the month of 12/2017 when the patient presented to the emergency department for a tumefaction involving the dorsal surface of the left foot and the diagnosis of necrotizing fasciitis was made, he underwent incision with good progress until the month 09/2018 when we were solicited for the appearance of a hyperbourgeonnante lesion on the dorso lateral side of the left foot with presence of angiomatous erythematous plaques at the level of the contralateral foot, left thigh and left flank. X-ray of the limb showed osteolysis. A skin biopsy was performed on the hyperbourgeonous lesion and one on the erythematous plaque which were in favor of Kaposi’s disease. Note that in 2 months the patient replaced the hyperbourgeonnant lesion in a ulcerocutaneous tumor taking the entire left foot (Figure 1) associated with a necrosis involving the 4 right toes in addition to erythematous plaques at the feet, thighs and trunk associated with an involvement of the oral and genital mucosa (Figure 2).

A balance sheet in search of other locations was realized and was normal and CT angiography showed stenosis of the posterior tibial artery.

The patient was a candidate for chemotherapy associated with amputation but the evolution was marked by his death before the start of treatment.
Discussion

Kaposi’s disease (MK) is a multicentric cell proliferation whose neoplastic nature remains controversial. Originally described in 1872 by the Viennese dermatologist Moritz Kaposi under the name of "idiopathic multiple pigmented sarcomas of the skin", it occurs in different epidemiological contexts allowing to individualize four main forms: classical or Mediterranean MK [3, 4]; the endemic MK described in sub-Saharan Africa before the HIV epidemic; MK occurring after organ transplants or iatrogenic immunosuppression; MK related to HIV or epidemic infection. These different forms of MK share basic clinical lesions and a similar histological picture characterized by a proliferation of spindle cells of endothelial origin, and a single viral etiological agent, the human herpesvirus 8 (HHV-8) [5-8].

Anaplastic transformation was first described in 1959 by Cox and Helwig [9] and, subsequently, other cases have been reported in the context of classic, African, and AIDS [10, 11].

The anaplastic forms are rare and there are few in the literature; they are described mainly in endemic countries. The few published series have proved that anaplastic Kaposi can occur in non-endemic countries and that all forms of the latter can evolve into anaplastic form, with a strong local aggressiveness. Some factors promoting this progression have been described, including the duration of SK, chronic lymphedema, various treatments received, including radiotherapy and chemotherapy, immunosuppression and HIV co-infection [12-14].

If the diagnosis of anaplastic KS is to be made clinically before the rapid onset of a tumor, histopathological examination is essential to allow diagnosis and to rule out other forms of malignancy, including angiosarcoma. In histopathological terms, anaplastic KS is in the form of relatively undifferentiated, medium to large, dense masses of epithelioid cells with a high number of mitoses and areas of necrosis. The expression of the HHV-8 antigen means that such proliferation can be attributed to SK. Unlike epithelioid angiosarcomas, in which the expression of CD34 and D2-40 often disappear, the anaplastic KS retains these two antigens [15].

There is no therapeutic consensus concerning anaplastic KS. While paclitaxel and liposomal doxorubicin are efficacious in the view of certain authors [2, 14].

Because of the absence of response of KS to standard therapies, coupled with associated infectious complications, amputation is occasionally required like the case of our patient.

Conclusion

Our case allows us to take stock of this entity, still poorly known, Kaposi’s disease which proves to be a very aggressive form can even be fatal. approach, more specifically a non-conservative surgical treatment with systemic chemotherapy, seems to be the most appropriate option for patients with anaplastic KS. Unfortunately, amputation due to deep tissue invasion and/or metastases is often required.

References

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