Scholars Journal of Medical Case Reports

Sch J Med Case Rep 2015; 3(6)540-542 ©Scholars Academic and Scientific Publishers (SAS Publishers) (An International Publisher for Academic and Scientific Resources) ISSN 2347-6559 (Online) ISSN 2347-9507 (Print)

DOI: 10.36347/sjmcr.2015.v03i06.028

Classic Kaposi sarcoma with massive bone destruction: rare form (a case report)

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Abstract: We report a case of a classic Kaposi sarcoma of the right foot with rapid and massive bone destruction leading to a transtibial amputation in a 70-year-old patient. The aggressive character of this indolent form, known by its slow Evolution makes the particularity of this presentation.

Keywords: Kaposi sarcoma; classic form; bone destruction.

INTRODUCTION

Kaposi sarcoma (KS) is an angioproliferative neoplasm that is probably a lymphatic origin and is associated with human herpes virus 8 (HHV8) infection. There are four variants of Kaposi sarcoma whose the classical form. This entity is usually considered indolent and characterized by slow evolution. The visceral involvement and aggressive character are very rare. We report the case of an aggressive classic KS with massive bone destruction.

OBSERVATION

A 70 years old patient of Mediterranean origin with no notable medical history, consulted for pigmented lesions of the feet evolving for over 20

years, complicated 1 month ago by bleeding. This symptomatology evolves in a general conservation. Dermatological examination revealed in the right foot a keratotic patch overcome of some angiomatous nodules with bilateral lymphedema without other skin or mucosal lesions. The HIV serology was negative and the skin biopsy returned in favor of KS. The decision to radiotherapy was laid but the patient refused. After one month, he was presented to the emergency with excruciating pain, ulceration and partial necrosis of the toes of the right foot (figure 1). Radiography had objectified massive bone destruction (figure 2). Extension assessment has not objectified other disease site. The patient was treated at this stage by a transtibial amputation.



Fig-1: Clinical presentation of cutaneous involvement

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Fig-2: massive bone destruction

DISCUSSION

Kaposi sarcoma is a multifocal angiogenic process characterized by multicentric vascular and cellular proliferation whose neoplastic Nature Remains discussed [1]. There are 4 different clinical-epidemiologic variants of KS [2,3], including African KS (endemic), acquired immune deficiency syndrome (AIDS) related KS (epidemic), transplantation (or immunosuppression) associated KS, and classic KS. This latter is a rare form, mainly affecting HIV negative men aged from Central and Eastern Europe, Mediterranean and Jews people, at the lower limb which is the case of our patient.

Clinically, KS affected mostly cutaneous sites European African and populations. individuals immunosuppressed (like human immunodeficiency virus infected patients), KS tends to be more aggressive and widespread. Patients present multifocal skin lesions, mucosal surfaces involvement, and lymph nodes with viscera dissemination. Skin lesions may be purple, brown, or red and frequently are accompanied by localized lymphedema. KS progresses from an early patch stage, to a thicker plaque stage and later, toward a nodular stage as the spindle cells proliferates. These large tumors may ulcerate, become deeply invasive, and create disfiguring lesions [4]. In our patient who had a classic KS, the lesions were nodular and ulcerated with distal necrosis suggesting rather angiosarcoma and possible degeneracy that has eliminated histological by aggressive immunohistochemical study. This appearance is very rare [5,6] and it is mainly the preserve of epidemic and endemic forms.

Bone and skeletal muscle KS involvement are unusual. It was described for the first time by Moriz Kaposi [7] and estimated at 4.5% [8] mainly in extended and multi visceral forms [9]. In the classic

form, it usually sits beside skin lesions and in several published reports, osseous KS lesions were asymptomatic. In our patient, bone pain in the foot was excruciating and lesions were localized without visceral involvement.

In the classic KS, bone involvement is peripheral beside skin lesions by direct invasion [2]. It evolves gradually over several years. Typically, radiographs show alteration of the soft tissues and of the skeleton. At the level of soft tissue, the X rays disclose solitary or multiple tumors which are located subcutaneously or more deeply. Skeletal changes that may occur anywhere include erosion, bone rarefaction and cyst formation. Different lesions were identified more easily with MRI than with CT scans. MRI also appeared to be more sensitive for detecting the involvement of spongy bone. Scintigraphy with sequential thallium and gallium scanning was helpful, to distinguish between KS, infections and lymphomas of the bone [10,11]. In our patient, bone destruction was rapid and massive which makes the rarity of this case of classic form.

Definitive diagnosis of KS is made through a biopsy. The clinical differential diagnoses of KS include bacillary angiomatosis, pyogenic granuloma, lymphoma, hemangiomas or other benign vascular proliferations. However, these entities are easily ruled out on histologic examination [12].

Several different treatment modalities have been used for classic KS. For localized lesions, treatment with surgical excision, or electrocauterization and curettage are generally effective. An alternative method of treatment is intralesional chemotherapy. In more advanced cases with disseminated cutaneous involvement, radiation and single agent or combination systemic chemotherapy has

achieved relatively good control of the lesions and effective short-term palliation. Radical surgical procedures, such as amputation, are used rarely today [2].

In our case and front of this aggressive and unusual behavior of the disease in a short period, amputation was necessary. The patient has benefited secondarily of a lower limb prosthesis enabling him to find his autonomy.

CONCLUSION

Our case describes a rare evolution of the classic Kaposi sarcoma and takes us to wonder whether this pathology is changing evolutionary profile.

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