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SIMULTANEOUS OCCURRENCE OF KAPOSI'S SARCOMA AND ANAPLASTIC T LYMPHOMA IN NON-AIDS PATIENT: NEW ASSOCIATION OR LIKELY COINCIDENCE

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ABSTRACT

Background: the association of Kaposi's sarcoma and lymphoma is rarely encountered, although the involvement of HHV8 in several lymphoid pathologies is known. **Case presentation:** we report a case of a 78-year-old man with Kaposi's sarcoma confirmed by cutaneous biopsy associated with anaplastic T large cell lymphoma on lymph node. Patient received chemotherapy according to the R-CHOP protocol with a good improvement. **Conclusions:** the concomitant existence of kaposi's sarcoma and lymphoproliferative disorders in Non-AIDS patients remains rare, it has been reported and appears mostly developed on the same lymph node, our case is unique because it's the first case reporting association between kaposi's sarcoma limited on the skin and anaplastic T large cell lymphoma, this association supposed different ethiopathogenic mechanisms of the both neoplasmas suggesting coincidental occurrence.

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INTRODUCTION

Kaposi's sarcoma (KS) is a multifocal angioproliferative disorder of vascular endothelium, primarily affecting mucocutaneous tissues with the potential to involve visceral (Mahnaz Fatahzadeh *et al.,* 2012; Lynen *et al.,* 2005). There are four clinical forms: classic, endemic, iatrogenic and epidemic KS show a distinct natural history and prognosis (Mahnaz Fatahzadeh *et al.,* 2012). The classic variant is rare; evolving in a chronic form and essentially affects patients over the age of 60 years, with male predominance, mostly in the Mediterranean and Eastern Europe. Endemic form of KS observed in young adults and HIV-negative children, and may progress to rapidly fatal fulminant lymph node form. Iatrogenic KS affects subjects who have had organ transplants or long-term immunosuppressive therapy and may regress after stopping this immunosuppression. Epidemic variant of KS is associated with HIV infection (Boulanger, 1999). It's a rare opportunistic tumor. It is also the second-most frequent tumor affecting HIV patients worldwide (Martellotta *et al.,* 2009) and the more aggressive form of this disorder.

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The infectious agent required for the development of KS is herpes virus type 8 (HHV-8), also known as the herpes virus associated with Kaposi's sarcoma (KSHV), remains present in all clinical subtypes of KS (Wood and Feller, 2008). Infection with this virus is necessary condition, but it is not sufficient alone to cause KS, other genetic factors and cytokines production seems be important in pathogenesis of KS (Ensoli *et al.,* 2001). Anaplastic Large Cell Lymphoma (ALCL) is a rare and aggressive peripheral T-cell lymphoma of CD30 positive lymphoproliferative lymphoma and extranodal sites. It comprises two subtypes, according to the expression of a protein called anaplastic lymphoma kinase (ALK): ALCL ALK positive and ALCL ALK negative. The coexistence of these pathology has been rarely associated specially in Non-acquired immunodeficiency syndrome (AIDS) Hodgkin's and then non-Hodgkin's lymphomas represent the most frequent event, observed in 6 to 17% of the classical KS. We report the case of a 78-year-old man with KS confirmed by cutaneous biopsy and anaplastic large cell lymphoma confirmed by the histological and immunohisto chemical study of lymph node.

Case presentation

A 78 year-old man, with no particular pathological history, was admitted in consultation for cutaneous lesions and

superficial right inguinal lymphadenopathy evolving since 6 months complicated 2 weeks before his admission by low grade fever and night sweats sensation, the patient also reports an unencrypted weight loss. Physical examination shows that the patient has good general condition, ECOG at 1, stable hemodynamically and on respiratory terms, afebrile, with right plantar papular lesions. The remainder of the clinical examination shows the presence at the right inguinal level of a lymphadenopathy of 3cm which adheres to the deep plane, not painful and without inflammatory sign. Diagnosis of nodular KS was retained after specific histological study who shows presence of vascular proliferation and non-encapsulated nodular fusocellular nerve cells, hollowed out by tight slits and large cavities in which red blood cells were observed, peripheral presence of a perivascular inflammatory infiltrate lymphocyte and plasma cell type with siderophages and spindle cell positivity with 100% CD34, HHV8 + 80%.

The HIV1 and HIV2 serologies were negative.

Given the presence of the general signs, the existence of adenopathy could not be related to a systemic form of KS and a biopsy of the right inguinal adenopathy was performed and objectified anaplastic large cell lymphoma CD45 +, CD4 +, CD30 + with Ki67 estimated at 70%, partial CD3 expression, No expression of CD8, CD5, CD56, CD10 and CD20, HHV8 was negative. Extension assessment was performed, showing isolated external and inguinal iliac adenopathies (Fig1), the bone marrow biopsy was not infiltrated; the patient was classified as stage IIBb according to ANN ARBOR classification. Chemotherapy was started according to RCHOP protocol (Rituximab, Cyclophosphamid, Vincristine, Doxorubin and steroid), the patient received six cures with complete remission after the 3rd RCHOP treatment.

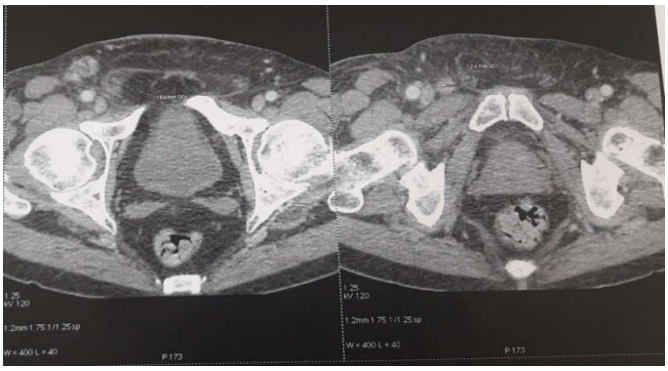

Fig. 1. External and inguinal iliac adenopathies

DISCUSSION

Kaposi's sarcoma was first described in 1872 by the Hungarian dermatologist Moritz Kaposi (Kaposi, 1872). It's a vascular tumor, affecting predominantly male aged over 50 years, except in Africa where the peak frequency is between 25 and 44 years, extracutaneous lesions are the most common implicating the lymph nodes, gastrointestinal tract and lung (Reynolds *et al.,* 1965). This disease is one of tumors induced by viruses most likely due to chronic HHV8 infection. In fact, seroconversion precedes Kaposi lesions by an average of 33 months (Jacob *et al.*). High incidence of KS associated HHV8 in AIDS patient was reported but KS on Non –AIDS patient is rare, incidence varies according to individual factors: origin, age, sex, and immune statut (Licci *et al.,* 2007). On the other hund, the association of KS with other rare lymphoproliferative diseases has been particularly emphasized, mainly affecting immunocompromised individuals, especially those infected with HIV, the main associated lymphoproliferations are: primary serous lymphoma, multicenter castelman disease and plasmaplastic lymphoma. In fact, KS association with lymphoproliferative disorders was observed for the first time in 1920 by Cole and Crump (Safai *et al.,* 1980). Since then, many publications have been published interesting association between KS and lymphoproliferative disorders on AIDS and Non-AIDS patients (Carbone, 2005; Chang *et al.,* 1994). Several authors have reported the coexistence of KS with other malignancies lymphoproliferation, the association has become usual. An Italian team affirms the association of the genome HHV8 in three cases of angioimmunoblastic lymphadenopathy out of fifteen studied, in HIV-negative subjects (Luppi *et al.,* 1996). Others noted the presence of the genome of HHV8 in benign reactional lymphadenopathies: according to the Chang team (Chang *et al.,* 1994) HHV8 was found in the ganglia of three AIDS patients, for Soulier (Soulier *et al.,* 1995) in a ganglion of HIV-negative woman and Chadburn in a ganglion of HIV-negative woman with systemic lupus (Chadburn *et al.,* 1997).

These ganglia have histologically follicular hyperplasia. Numerous hypotheses have been proposed to explain the increase of the frequency of association of KS with lymphomas, the first and that the agent responsible for KS can independently stimulate the reticular tissues towards neoplasia (Reynolds *et al.,* 1965), the proof is the positivity of HHV8 in several lymphoproliferations, the second possibility is that the KS is part of the spectrum of the reticular neoplasms, thus, in two cases previously described the tissue sections have revealed a histological transition of the KS and the lymphoma especially the Hodgkin's lymphoma. Non-AIDS related KS appears mostly limited to the skin and is well responsive to treatment (Rescigno *et al.,* 2013). In fact, in contrast to AIDS associated KS, KS in Non-AIDS patients seems less aggressive, mostly limited to the skin (Licci *et al.,* 2007). Research in the medline database found no reported cases of KS with nodal anaplastic T lymphoma especially in an immunocompetent patient with negative HHV8 serology, it is thus that during the course of Kaposi's sarcoma the appearance of a lymphadenopathy should alert the physician to the possibility of the association with hematological disease especially in the presence of general signs, the negativity of the HHV8 obliges us to look for other risk factors that may explain this association. Our case is unique by several aspects, first it's the first case of KS associated with anaplasic T large cell lymphoma, second KS and lymphoma were not found within the same lymph node like mostly reported in literature, KS was limited to the skin as reported for Non-AIDS patient. The response to treatment was good and complete remission was achived. That supposed different etiopathogenic mechanisms of the two neoplasias suggesting coincidental occurrence.

Conclusion

We have reported, to our knowledge, the first case of Kaposi's sarcoma associated with lymph node anaplastic T lymphoma. It's an exceptional association making probably the role of risk factors other than HHV8 in the association of Kaposi's disease-malignant hemopathy, this concept would become an essential element to progress in understanding the pathophysiology of

these two diseases in order to ensure better therapeutic management.

List of abbreviations

KS: Kaposi's sarcoma
HHV8: Human herpes virus 8
AIDS: Acquired Immune Deficiency Syndrome
HIV: Human Immunodeficiency Virus
ALCL: Anaplastic Large Cell Lymphoma
ALK: Anaplastic lymphoma kinase

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