Case Report

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A rare coexistence of Kaposi's sarcoma and myelodysplastic syndrome in an immunocompetent patient: A fortuitous association?

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Abstract

Kaposi's sarcoma (KS) is an angiogenic tumor related to the Human Herpes Virus 8 (HHV8). Its association with myelodysplastic syndrome (MDS) had been rarely reported in the literature. Herein, we relate a case of coexistence of KS and MDS in a human immunodeficiency virus (HIV) negative female patient, which is to our knowledge the fourth reported in the literature. Some data suggest that viral agents might be involved in the pathogenesis of MDS. The HHV8 is a virus with tropism for hematopoietic cells (monocyte, B lymphocyte) and expression of transforming viral genes that may affect cell proliferation and thus cause the trigger of myelodysplastic disorders. However, other studies on larger populations are necessary to determine the causal correlation between these two disorders.

Keywords: Kaposi's sarcoma; myelodysplastic syndrome; HHV-8.

Introduction

Kaposi's sarcoma (KS) is an angioproliferative, multi-centric and indolent tumor caused by the Human Herpes Virus 8 (HHV8). It comes in 4 subtypes: Endemic in equatorial Africa, classic non-endemic, epidemic in patients living with HIV and iatrogenic in organ transplant patients [1]. Myelodysplastic syndrome (MDS) is a heterogeneous group of hematopoietic stem cell diseases characterized by abnormal development of one or more myeloid lines with inefficient hematopoiesis and risk of transformation into acute leukemia.

The simultaneous occurrence of multiple primary neoplasms in the same individual is not rare and suggests the involvement of multiple common etiopathogenetic factors [2]. Herein, we report a rare case of coexistence of KS and MDS in a human immunodeficiency virus (HIV) negative female patient, which is to our knowledge the fourth reported in the literature.

Case Report

A 75-year-old woman with a history of hypertension and a chronic end-stage renal failure, presented 2 months before her consultation nodular angiomatous lesions on the 2 lower limbs (**Figure 1**), palmoplantar sides and on the oral palate (**Figure 2**), all evolving in a context of weight loss and asthenia. The clinical examination had found conjunctival pallor, inguinal lymphadenopathies and a large painful right lower limb. A venous echo doppler was performedand has revealed a venous thrombosis of the superficial femoral vein. The skin biopsyhas objectified a dermal fusocellular proliferation with the presence of medium to large cells with elongated monomorphic fine chromatin nuclei and extracellular hemosiderin deposition with HHV-8 positivity. The HIV serology was negative. A blood count was performed and showed a macrocytic normochromic anemia with hemoglobin at 5 g/dl; leukocytes at4000/mm3; platelets at 184,000/mm3 and normal rates of B12 vitamin and folic acid. The patient's bone marrow aspiration has objectified the presence of signs of dyshemopoesis and multilinear dysplasia thus concluding to a myelodysplastic syndrome without an excess of blasts.

In view of this, our patient was diagnosed with Kaposi's sarcoma with myelodysplastic syndrome without signs of transformation. A chest X-ray was performed without any detectable abnormalities. However, the gastroduodenal fibroscopy and the proctologic examination could not be performed due to the patient's refusal. Our patient benefited from blood transfusions with treatment of her venous thrombosis with heparin therapy and received a hemodialysis session for her end-stage renal failure. Bleomycin was proposedto her but due to its unavailability, she was planned for radiotherapy but unfortunately, she died before her first session due to a pulmonary embolism.

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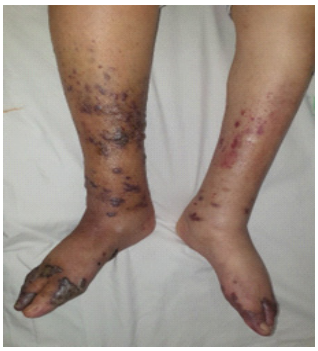

Figure 1: *Nodular angiomatous lesions on the 2 lower limbs.*

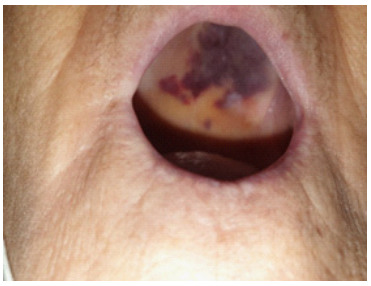

Figure 2: *Nodular angiomatous lesions of the oral palate.*

Discussion

Kaposi's sarcoma is a low-grade tumor originating from vascular and lymphatic endothelium. It is causedby the HHV-8 also known as KS – associated herpesvirus, which isa gamma herpes virus with double-stranded DNA that infects a variety of cells including endothelial cells, B cells and monocytes [1]. DNA sequences of this novel herpesvirus have been isolated from lesions of all KS variants and from peripheral blood mononuclear cells of affected patients [1]. In our work, we report a simultaneous diagnosis of Kaposi'ssarcoma and myelodysplastic syndrome which is, to our knowledge, the fourth case reported in the literature [3,4,5]. This association had been rarely described, therefore its etiogenesis remains not completely clarified and difficult to elucidate. Myelodysplastic syndrome is a heterogeneous group of clonal disorders that may be related to many etiologies [7-10]. Genetic factors could contribute to the disease; the acquisition of somatic mutations promote clonal expansion and dominance of a particular hematopoietic stem cell [8]. Also, some literature data suggest that viral agents might be involved in the pathogenesis of hematologic malignancies [9]. Some viruses can infect the bone marrow stem cell leading to deregulated cytokine production and changes in cell clones resulting in the onset of myeloproliferative disorders, such as MDS [9]. Yamamoto et al. reported the case of a patient who developed a myelodysplastic syndrome 2 years after an Epstein-Barr virus (EBV) infection associated with the hemophagocytic syndrome [11]. In a study about the incidence of human T-cell leukemia virus (HTLV) in hematological diseases in a non-endemic region of Central Europe, Karlic et al. had detected HTLV-1 only in myelodysplastic syndromes with an incidence of 17% [12]. Recently, it has been suggested that the SARS-CoV-2 virus, by promoting the reactivation of other viruses such as Cytomegalovirus (CMV) and EBV, could also induce the development of MDS [13].

Conclusion

The HHV-8 is a virus with tropism for hematopoeitic cells (monocyte, B lymphocyte) and expresses some transforming viral genes that can affect cell proliferation and thus cause the trigger of myelodysplastic disorders. However, further studies are needed to determine whether the association of these 2 pathologies has a well-established etiopathogenic causal correlation or whether it is a fortuitous association.

Conflict of interest: None.

References

1. Dupin, Nicolas. Update on oncogenesis and therapy for Kaposi sarcoma. Current Opinion in Oncology.2020; 32: 122-128.

2. Safai B, Miké V, Giraldo G, Beth E, Good RA. Association of Kaposi's sarcoma with second primary malignancies: possible etiopathogenic implications. Cancer. 1980; 45(6): 1472-9.

3. Tombuloğlu M, Keskin A, Töbü M, Cağirgan S, Büyükkeçeci F, Soydan S. Kaposi's sarcoma in the course of juvenile myelodysplastic syndrome. Acta Oncol. 1995; 34(2): 263-4.

4. Pamuk GE, Aydogdu E, Turgut B, Demir M. The simultaneous diagnosis of Kaposi sarcoma and MDS RAEB-II in a human immunodeficiency virus-negative patient. A rare occurrence. Ann Hematol. 2009; 88(4): 389-91.

5. Gürbüz M, Özyurt N, Utkan N. Patient with Kaposi Sarcoma and Myelodysplastic Syndrome Ankara Üniversitesi Tıp Fakültesi Mecmuası. 2018; 71(3): 265-267.

6. Fossati S, Boneschi V, Ferrucci S, Brambilla L. Human immunodeficiency virus negative Kaposi sarcoma and lymphoproliferative disorders. Cancer. 1999; 85(7): 1611-5.

7. Shallis RM, Ahmad R, Zeidan AM. The genetic and molecular pathogenesis of myelodysplastic syndromes. Eur J Haematol. 2018; 101(3): 260-271.

8. Schratz KE, DeZern AE. Genetic Predisposition to Myelodysplastic Syndrome in Clinical Practice. Hematol Oncol Clin North Am. 2020; 34(2): 333-356.

9. Raza A. Hypothesis: myelodysplastic syndromes may have a viral etiology. Int J Hematol. 1998; 68(3): 245-56.

10. Hasserjian RP. Myelodysplastic Syndrome Updated. Pathobiology. 2019; 86(1): 7-13.

11. Yamamoto H, Hattori H, Takagi E, et al. MonoMAC syndrome patient developing myelodysplastic syndrome following persistent EBV infection. The Japanese Journal of Clinical Hematology. 2018; 59(3): 315-322.

12. Karlic H, Möstl M, Mucke H, Pavlova B, Pfeilstöcker M, Heinz R. Association of human T-cell leukemia virus and myelodysplastic syndrome in a central European population. Cancer Res. 1997; 57(21): 4718-21.

13. Juric I, Katalinic L, Furic-Cunko V, Basic-Jukic N. Myelodysplastic syndrome in a kidney transplant recipient after SARS-CoV-2 infection: can SARS-CoV-2 induce myelodysplastic syndrome? Int Urol Nephrol. 2022; 54(7): 1775-1776.