Case Report

Simultaneous presentation of two noninflammatory lung diseases in an HIV-infected patient

Rafael Martínez-Girón, Santiago Martínez-Torre[1]

Departament of Anatomic Pathology, INCLINICA Foundation for Clinical, Pneumological and Carcinogenic Research, Calvo Sotelo, 16, 33007, Oviedo, [1]Department of Family and Community Medicine, Hospital Universitario La Paz, Paseo de la Castellana, 261, Madrid 28046, Spain

Address for correspondence:
Dr. Rafael Martínez-Girón, INCLÍNICA Foundation, Calvo Sotelo, 16, 33007 Oviedo, Spain.
E-mail: rmartinezgiron@hotmail.com

Submission: 10-10-2016
Accepted: 10-11-2016

Access this article online

Quick Response Code:

Website: www.thoracicmedicine.org

DOI: 10.4103/atm.ATM_335_16

Abstract:

The simultaneous presentation of two noninflammatory pulmonary diseases, pulmonary alveolar proteinosis and Kaposi's sarcoma (Ks), in an HIV-infected patient, is described. A 29-year-old black race patient was admitted to the hospital because of general malaise, weight loss, dyspnea, chest pain, and cough with hemoptoic expectoration. Chest X-rays revealed a patchy bilateral alveolar pattern with a tendency toward the formation of condensations. The serological test revealed HIV positivity (CD4 counts of 393 cells/mm^3). Because there was no response to the treatment course, a thoracic CT was performed, showing interlobular thickening with intralobular septal lines and ground glass opacities ("crazy-paving" pattern). An open lung biopsy was performed. Histopathological diagnosis of pulmonary alveolar proteinosis and pulmonary Ks was made.

Key words:

HIV-infection, pulmonary alveolar proteinosis, pulmonary Kaposi's sarcoma

Pulmonary alveolar proteinosis (PAP) is a rare disease characterized by an accumulation of pulmonary alveolar spaces containing a proteinaceous material that is rich in phospholipids due to defective surfactant clearance by alveolar macrophages.[1] Kaposi's sarcoma (Ks) is a low-grade mesenchymal tumor that involves blood and lymphatic vessels. Moreover, Ks may involve the lung parenchyma, bronchial tree, and pleural surfaces.[2]

This case presents the unique simultaneous presentation of two noninflammatory pulmonary diseases, PAP and Ks, in an HIV-infected patient.

Case Report

A 29-year-old black man, an immigrant from Equatorial Guinea, was admitted to the hospital because of general malaise, weight loss, dyspnea, chest pain, and cough with hemoptoic expectoration. On physical examination, the patient was found to be febrile (38.2°C), tachycardic and tachypneic. A diminished respiratory murmur in both lungs was noticed during auscultation. Cervical and axillary lymphadenopathies were also noted. Noncutaneous and oropharyngeal lesions were observed.

Chest X-rays revealed a patchy bilateral alveolar pattern with a tendency toward the formation of condensations [Figure 1a]. The serological test revealed HIV positivity (CD4 counts of 393 cells/mm^3). Initially, the patient was treated with co-trimoxazole. Because there was no response to the treatment course, a thoracic computed tomography (CT) was performed, showing interlobular thickening with intralobular septal lines and ground glass opacities ("crazy-paving" pattern). Moreover, marked peribronchovascular thickening and parenchymal nodularity were also observed [Figure 1b].

Based on these findings, an open lung biopsy was performed. In the first histological sections [Figure 1c], the alveolar spaces showed occupation by a dense and eosinophilic material containing cells corresponding to alveolar macrophages that were loaded with lipid-containing vacuoles. On the upper right, infiltration by long, atypical mesenchymal cells can be observed. In other histological sections [Figure 1d], fusocelular-type elements are observed, forming interwoven bundles and occasionally leaving cracks covered

How to cite this article: Martínez-Girón R, Martínez-Torre S. Simultaneous presentation of two noninflammatory lung diseases in an HIV-infected patient. Ann Thorac Med 2017;12:125-6.

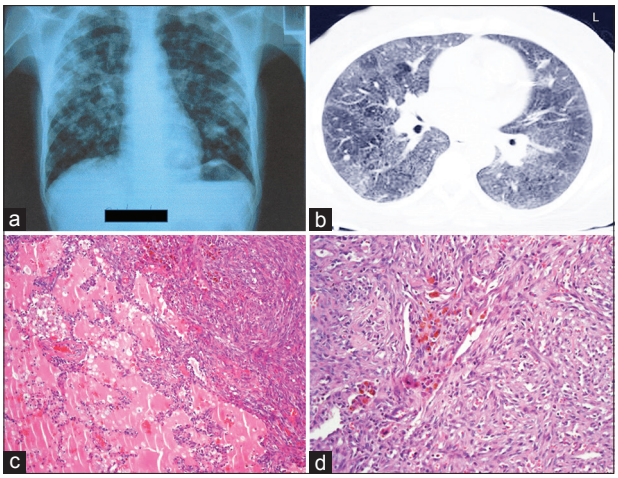

Figure 1: (a) Chest X-ray showing a patchy bilateral alveolar pattern with a tendency toward the formation of condensations. (b) Thoracic computed tomography showing interlobular thickening with intralobular septal lines and ground glass opacities ("crazy-paving" pattern). Marked peribronchovascular thickening and parenchymal nodularity is also observed. (c) Lung biopsy. The alveolar spaces are occupied by a dense and eosinophilic material containing cells corresponding to alveolar macrophages that are loaded with lipid vacuoles. On the upper right, infiltration by long, atypical mesenchymal cells can be observed (H and E, ×100). (d) Lung biopsy. Fusocellular-type elements are present, forming interwoven bundles and occasionally leaving cracks covered by the endothelium and proliferation of fibroblasts. Areas of interstitial hemorrhage are also frequently observed (center of image) (H and E, ×200)

by endothelium and proliferating fibroblasts. Areas of interstitial hemorrhage were also frequently observed (center of the image). The histopathological diagnosis was PAP and pulmonary Ks.

Initial management included antiretroviral therapy with zidovudine and didanosine. After optimizing the antiretroviral therapy and before beginning the antitumoral treatment, the patient died.

Discussion

This case presents the unique simultaneous presentation of two noninflammatory pulmonary diseases, PAP and Ks, in an HIV-infected patient.

It is becoming increasingly recognized that HIV infection promotes the risk of noninfectious pulmonary diseases.[3] Because pulmonary infections during HIV infection are generally well-known, clinicians must be aware of HIV patients presenting noninfectious pulmonary diseases.

PAP is a rare disease characterized by an accumulation of pulmonary alveolar spaces containing a proteinaceous material that is rich in phospholipids due to defective surfactant clearance by alveolar macrophages. Three clinical forms of PAP have been defined depending on the etiology: Genetic, auto-immune, and secondary. The forms of this disease are known to be secondary to other pathologies such as neoplasias, immunodeficiencies, and environmental exposure to inorganic agents. Despite these risk factors, there have been few reports of PAP in HIV-infected patients.[4]

Ks is a low-grade mesenchymal tumor that involves blood and lymphatic vessels. In HIV-infected patients, Ks is diagnostic of AIDS. Ks may involve the lung parenchyma, bronchial tree, and pleural surfaces. In general, patients with pulmonary Ks have advanced HIV disease. Evidence from epidemiologic, serologic, and molecular studies indicate that Ks is associated with human herpes virus type 8, which is also known as Ks-associated herpes virus.[5]

Before the advent of highly active antiretroviral therapy (HAART), pulmonary Ks had been reported in approximately 10% of patients with AIDS and 25% of patients with cutaneous Ks[6] (in our case, one of the most striking features was the absence of cutaneous lesions characteristic of Ks, especially in a patient suffering from pulmonary complications). In a cohort of 305 HIV-infected patients, 25 had pulmonary Ks, and the incidence of pulmonary Ks was higher in Africans that in non-Africans.[7]

Some studies have demonstrated that the incidence of Ks decreased from 30/1000 patient-years in the pre-HAART era to 0.03/1000 patient-years in the HAART era.[8]

Because pulmonary Ks in HIV-infected patients is often indistinguishable based on clinical and radiologic criteria derived from opportunistic pneumonia, it is important to highlight that, due to the prolonged survival of such patients, noninfectious pulmonary complications must be taken into account to prevent respiratory compromise and failure.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References

1. Chan AK, Takano A, Hsu AL, Low SY. Pulmonary alveolar proteinosis. J Thorac Dis 2014;6:E77-80.
2. Sigel K, Pitts R, Crothers K. Lung malignancies in HIV infection. Semin Respir Crit Care Med 2016;37:267-76.
3. Staitieh B, Guidot DM. Noninfectious pulmonary complications of human immunodeficiency virus infection. Am J Med Sci 2014;348:502-11.
4. Tejwani D, Delacruz AE, Niazi M, Diaz-Fuentes G. Unsuspected pulmonary alveolar proteinosis in a patient with acquired immunodeficiency syndrome: A case report. J Med Case Rep 2011;5:46.
5. Schulz TF, Cesarman E. Kaposi sarcoma-associated herpesvirus: Mechanisms of oncogenesis. Curr Opin Virol 2015;14:116-28.
6. Aboulafia DM. The epidemiologic, pathologic, and clinical features of AIDS-associated pulmonary Kaposi's sarcoma. Chest 2000;117:1128-45.
7. Palmieri C, Dhillon T, Thirlwell C, Newsom-Davis T, Young AM, Nelson M, *et al.* Pulmonary Kaposi sarcoma in the era of highly active antiretroviral therapy. HIV Med 2006;7:291-3.
8. Gasparetto TD, Marchiori E, Lourenço S, Zanetti G, Vianna AD, Santos AA, *et al.* Pulmonary involvement in Kaposi sarcoma: Correlation between imaging and pathology. Orphanet J Rare Dis 2009;4:18.