

Upper tracheal Kaposi's sarcoma: An infrequent cause of upper airway obstruction

ERICK EDUARDO CÉPEDES MENESES*

BENITO ARTURO NIEBLA ÁLVAREZ†

JOSÉ MORALES GÓMEZ‡

JOSÉ LUIS TÉLLEZ BECERRA||

ELIO GERMÁN RECINOS CARRERA¶

* General Surgeon. Third year Thoracic Surgery Resident, Instituto Nacional de Enfermedades Respiratorias Ismael Cosío Villegas (INER). National Institute for Respiratory Diseases. Mexico City, Mexico.

† Thoracic Surgeon. INER.

‡ Thoracic Surgeon. Chief of the Division of Thoracic Surgery, INER.

|| Thoracic Surgeon, INER.

¶ Pulmonary Pathologist. INER.

Trabajo recibido: 18-IX-2007; aceptado: 05-XI-2007

ABSTRACT

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The frequency of Kaposi's sarcoma (KS) in patients with acquired immune deficiency syndrome (AIDS)

has decreased since the introduction of highly active antiretroviral therapy. Its commonest manifestations are cutaneous violaceous papulae or nodules. When KS presents in the tracheobronchial tree, the lesions are usually flat or only slightly elevated, and seldom produce symptoms of airway obstruction.

We report a case of an homosexual man with AIDS and cutaneous lesions, admitted with a 20 day's history of progressive upper airway obstruction without a past history of tracheal intubation, instrumentation or trauma, the responsible lesion being an upper intratracheal KS obstructing about 60% of the tracheal lumen; a vocal cord nodule and the cutaneous lesions were also KS.

Key words: Kaposi's sarcoma, tracheal Kaposi's sarcoma, vocal cord Kaposi's sarcoma, acquired immunodeficiency syndrome (AIDS), cutaneous Kaposi's sarcoma, upper airway obstruction, tracheal tumors, vocal cord tumors.

Palabras clave: Sarcoma de Kaposi de vía aérea, sarcoma de Kaposi traqueal, sarcoma de Kaposi de cuerda vocal, síndrome de inmunodeficiencia adquirida, sarcoma de Kaposi cutáneo, obstrucción de vía aérea superior, tumores traqueales, tumores de cuerda vocal.

RESUMEN

El sarcoma de Kaposi (SK) en pacientes con síndrome de inmunodeficiencia adquirida (SIDA), ha disminuido desde la introducción de la terapia antirretroviral altamente efectiva. Las manifestaciones más comunes son nódulos o pápulas violáceas cutáneas. Cuando se localiza en tráquea o bronquios, es raro que provoque síntomas de oclusión de vía aérea superior debido a que las lesiones habitualmente son aplanadas o apenas elevadas.

Informamos sobre un paciente homosexual con SIDA y lesiones cutáneas que se presentó por síntomas de obstrucción progresiva de vías aéreas superiores de 20 días de evolución, sin antecedentes de intubación, instrumentación o trauma traqueal, causado por SK intratraqueal; un nódulo de cuerda vocal y las lesiones cutáneas también fueron SK.

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CASE REPORT

A 23 year old homosexual male, diagnosed 5 months before with the human immunodeficiency virus (HIV) and receiving treatment with efavirenz (Sustiva, Bristol-Myers Squibb Co, Princeton, NJ) and zidovudine + lamivudine (Combivir, Glaxo-SmithKline, México, DF) presented himself to the Emergency Department of our institution with a twenty day's history of progressive stridor, wheezing, hoarseness, dyspnea, and the appearance of several violaceous, nodular lesions in his left thigh; there was no past medical history of tracheal intubation, instrumentation, or trauma. He had oral thrush, an erythematous pharynx and 1 to 1.5 cm violaceous nodular lesions at the left ear, right supra scapular region and left thigh, and multiple erythematous papules in the thorax; the rest of the physical examination and the chest roentgenogram were unremarkable. Laboratory values documented: 22 TCD4 cells/mm³, leucopenia; tests for hepatitis B and C, toxoplasmosis and cytomegalovirus were negative. Several skin lesions were biopsied. He was admitted with the suspicion of upper airway stenosis and tentatively managed with saline and racemic epinephrine nebulizations, with improvement. The next day, fiberoptic bronchoscopy (FB-18V, PENTAX, Tokio, Japan) showed a violaceous nodular lesion in the right vocal cord and a tracheal, violaceous, bi-lobulated tumor implanted at the level of the 1st and 2nd tracheal rings (Figure 1A, 1B), occluding the diameter of the trachea by about 60%; the

rest of the tracheobronchial tree, although appeared normal, was profusely lavaged with saline solution. Immediately, rigid bronchoscopy (10318, Karl Storz, Germany) was used to core out and resect the tumor with alligator forceps, without bleeding. The tracheal tumor, vocal cord nodule and multiple skin lesions were diagnosed as KS (Figure 1C). Bronchoalveolar lavage culture grew *Mycobacterium avium*. His postbronchoscopy course has been satisfactory.

DISCUSSION

In 1872, Moricz Kaposi, a dermatologist from Hungary, described the disease bearing his name; he characterized the tumors as violaceous, macular cutaneous lesions with irregular borders, affecting predominantly the lower extremities of elderly men. Its origin seems to be the pluripotent angioblast, probably stimulated by cytomegalovirus or other growth factors (oncostatin M, gamma interferon, interleukin (IL)-1, IL-6, IL-8, tumor necrosis factor, vascular endothelial growth factor).¹

This entity, Kaposi's sarcoma (KS), is observed about 20 times more frequently in homosexual men, and is the acquired immunodeficiency syndrome (AIDS)-related most common malignancy and the second most common AIDS associated disease.

Pulmonary KS affecting the parenchyma, pleura or tracheobronchial tree presents in 47-75% of patients with cutaneous disease; the lung involve-

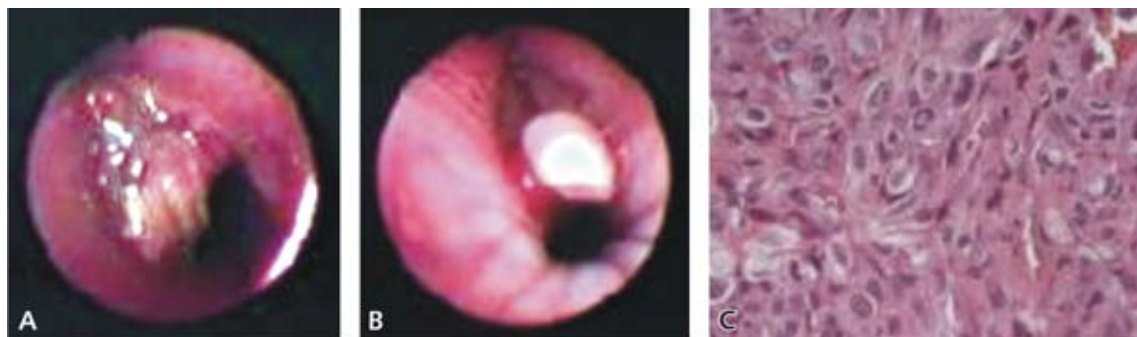


Figure 1. A) Images obtained at bronchoscopy; there is an obstructing lesion, about 60%, at the entrance of the trachea; B) Lesion at the vocal cord, C) Neoplasm characterized by multiple vascular channels, spindle cells with atypical mitosis and extravasated erythrocytes (40XX).

ment occurs late in the course of the disease.² Dyspnea, cough, chest pain, hemoptysis and wheezing can occur as presenting symptoms. Frequently, a concomitant infection due to *Pneumocystis jiroveci* or *Mycobacterium avium* is present.³

Tracheobronchial KS in AIDS patients is not common, and generally occurs in patients with CD4 count below 100 per cubic millimeter.⁴ KS lesions in the tracheobronchial tree may vary from flat, violaceous, vascular appearing purple-red raised nodules, or diffuse wall thickening.² Exceptionally, a growth significant enough to cause airway obstruction is found. In the larynx, the supraglottic location is the most common site causing difficulty of the airway (65%), followed by glottic (47%) and subglottic (18%) disease.^{5,6}

In this paper, we report a case of tracheal and vocal cord SK that obstructed the airway and presented as tracheal stenosis which was treated by resection with immediate improvement of symptoms and no complications. The complications reported for KS of the upper airways are excessive bleeding, the need for urgent tracheostomy to preserve a patent airway, and death which fortunately did not occur in this patient.⁷

Pathologic examination shows KS tumors to be composed of spindle cells, extravasated erythrocytes between the stromal cells, multiple irregularly shaped vascular spaces lined by pleomorphic cells and inflammatory infiltrate.⁸

Biopsy of the airways is not always necessary, unless there are atypical lesions or absence of the cutaneous manifestations of KS,¹ but the accuracy by fiberoptic bronchoscopy may not be high because of the patchy distribution and deep location of the tumor.⁹ In this case, the resected specimen of the severely obstructed airway was sent for examination and the diagnosis made at the same time as that of the skin lesions. The use of alligator forceps to resect tissue may increase the risk of bleeding.³

Although in good condition at the present time, the overall survival after the diagnosis of bronchopulmonary involvement has been reported from 4 to 6 months.¹⁰

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Correspondence:

Dr. Benito Niebla Álvarez and Dr.
Erick Céspedes Meneses.
Postal address: Instituto Nacional de
Enfermedades Respiratorias Ismael
Cosío Villegas. Calzada de Tlalpan
4502, colonia Sección XVI. México,
DF. Postal code: 14080. Telephone:
56664539.
e-mail: ihiyotl@yahoo.com