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A case of recipient-derived pulmonary Kaposi sarcoma after bilateral lung transplantation

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A 67-year-old, non-Hispanic white male underwent bilateral lung transplantation in 2016 for idiopathic pulmonary fibrosis. The patient had good graft function after transplantation, with a peak forced expiratory volume in 1 second of 2.27 liters. Surveillance bronchoscopy and biopsies done at 1, 3, and 6 months post-transplantation did not show any evidence of acute rejection or infection. He was lost to follow-up until 15 months after transplantation, when he presented with 1 week of dyspnea, cough, and fever. He was started on broad-spectrum antibiotics and intubated for hypoxemic respiratory failure.

A high-resolution computed tomography chest scan demonstrated new and ground-glass opacities in all lobes of the right lung with narrowing of the bronchi distal to the anastomosis in the right hilum (Figure 1A). Bronchoscopy revealed endobronchial nodules extending from the native airways at the right anastomosis to the segmental airways of the transplanted lung (Figure 1B). Endobronchial and transbronchial biopsy specimens demonstrated numerous spindle-shaped cells with positive stains for ETS-related gene (ERG) and human herpesvirus 8 (HHV-8), consistent with a diagnosis of Kaposi sarcoma (Figure 2A–C).

The patient ultimately elected for palliative care and expired. On autopsy, diffuse peribronchial and perivascular infiltration of tumor was noted involving the distal trachea and bilateral bronchi, with more extensive involvement on the right (Figure 2D–E). Fluorescence in situ hybridization analysis for sex chromosomes was performed on a section of tumor given donor–recipient mismatch. Results showed a chimeric XX/XY hybridization pattern with 74% XY cells (recipient) and 26% XX cells (donor), suggesting that the tumor likely arose within the patient's native tissue before spreading into the transplanted lungs.

Kaposi sarcoma (KS) is an angioproliferative disorder requiring infection with HHV-8 for its development.¹ Four epidemiologic–clinical forms of KS are recognized: classical; endemic–African; AIDS-associated and iatrogenic; or transplant-associated.¹ Primary infection by HHV-8 is followed by immune evasion and latent infection in a reservoir of B-cells.^{2,3}

Disclosure statement

The authors have no conflicts of interest to disclose.

Immune dysregulation results in reactivation and promotion of aberrant angiogenesis and a rich inflammatory infiltrate characteristic of KS lesions.³ Skin involvement is the most common manifestation of transplant-associated KS, whereas visceral involvement occurs in 25% to 50% of cases.⁴

The United States is considered a low-prevalence country, with an HHV-8 seroprevalence of 7.3% among U.S. blood donors⁵ and a cumulative incidence of KS of 0.06% at 8 years after solid-organ transplantation.⁶ Risk factors for the development of transplant-associated KS include male gender, Hispanic ethnicity, lung transplantation, travel or residence in Africa or the Mediterranean region, and use of calcineurin inhibitors. The cornerstone of therapy for KS in solid-organ transplantation remains reduction in immunosuppression.⁷ Switching from a calcineurin inhibitor to sirolimus and anthracycline-based chemotherapy are alternative therapies in refractory cases.⁷

Herein we have presented a patient who developed recipient-derived KS after lung transplantation. Most transplant recipients in the United States are HHV-8 seronegative before transplantation, so primary infection with HHV-8 is more likely than acquisition from infected donor organs.⁴ Despite its low prevalence, KS is a major oncologic consideration in solid-organ transplant recipients, particularly after lung transplantation.

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Figure 1.

(A) Representative cross-section of the patient's initial chest computed tomography scan showing extensive opacities and narrowing of the bronchi distal to the anastomosis in the right hilum (red arrow). (B) Endoscopic view of the bronchus intermedius showing nodular lesions throughout the airways (black arrows).

Figure 2.

(A-C) Histologic images of endobronchial biopsy showing a spindle-cell neoplasm with positive immunohistochemical staining for ERG (B) and HHV-8 (C), consistent with Kaposi sarcoma (original magnification: 200×). (D, E) Transplanted lung at autopsy showing prominent pulmonary vasculature (D) (arrowheads). (E) Histologic image of the lung showing the same spindle-cell neoplasm growing along the pulmonary vasculature (original magnification: 200×).