# KAPOSI SARCOMA IN THE HEAD AND NECK REGION FOLLOWING HEMATOPOIETIC STEM CELL TRANSPLANTATION: ANALYSIS OF 13 CASES

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## Introduction

Kaposi sarcoma (KS) occurs mainly in patients who are severely immunocompromised especially following solid organ transplantation where the risk to develop KS is 400–500 folds higher than that of the general population. However, the development of KS following Hematopoietic Stem Cell Transplantation (HSCT) is rare. Human herpesvirus 8 (HHV-8) is causally associated with pathogenesis of KS, encoding putative oncogenes and genes that stimulate angiogenesis and cell proliferation. The aim of this study was to review the clinical characteristics of KS in the head and neck region following HSCT.

## Methods

We conducted an English literature search (years 2000-2022) for cases of KS involving the head and neck region following HSCT. Key words included in the search: Kaposi sarcoma, hematopoietic stem cell transplantation, bone marrow transplantation.
In addition, a two new cases from our medical center were include.
Cases with no sufficient clinical data or definite diagnosis were excluded.

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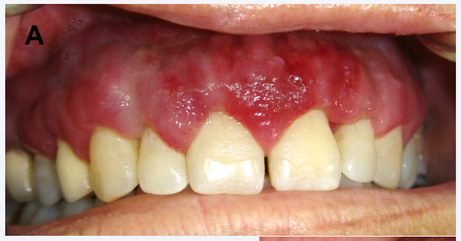
|---|---|---|
| **Gender** | Male | 76.92% |
| | Female | 23.08% |
| **Mean age** (range) | | 37.1 (7-69 years) |
| **Type of BMT** | allo-HSCT | 84.62% |
| | auto-HSCT | 15.38% |
| **Immunosuppressive therapy** (N) | Cyclosporine | 10 |
| | Prednisone | 9 |
| | Methotrexate | 2 |
| | Rituximab | 1 |
| | Tacrolimus | 1 |
| **Mean time from HSCT to KS diagnosis** | | 229 days (100-463) |
| **HHV-8 positive** | | 91.67% |

Table 1. Data summary from the review of literature



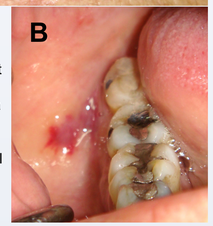




Figure 1: A 56 years-old-female presented with gingival swelling of two weeks duration. She underwent allo-HSCT due to MDS 7 months earlier. Oral examination revealed a generalized swelling and erythema of the gingiva (A) and bilateral hematoma-like lesions of the buccal mucosa (B). Biopsy confirmed the diagnosis of KS.

## Results

A total of 13 cases met the inclusion criteria. Eleven patients developed KS following allogeneic and 2 following autologous HSCT. The mean age at diagnosis was 37.1 years (range: 7-69 years) with predilection for males (male to female ratio 3:1). The mean time from HSCT to the presentation of KS was 229 days (range: 100-463). The oral mucosa (9 cases) was the most prevalent site for KS in the head and neck region, followed by skin lesions (6 cases). Only three cases demonstrated lymph nodes involvement. The status of HHV-8 was reported in 12 cases and was positive in 11 (91.67%). Ten patients (76.92%) had immunosuppression withdrawn, following diagnoses of KS, with complete response. The other three patients were treated with radiotherapy, cryotherapy and rapamycin. Nine patients were free of disease at the last follow-up (mean 44,7 months; range 2 months-15 years). Two patients died of KS, one and fifteen months following the diagnosis. One patient died due to relapse of the underlying disease.

## Conclusion:

Kaposi sarcoma is a rare complication of HSCT characterized by a rapidly progressive course and might be life-threatening.
HHV-8 has been shown to cause KS in significant percentage of immunosuppressed patients, and its presence should be sought in blood and in biopsy specimens.
In many cases, withdrawal of immunosuppressive therapy leads to resolution of KS. Other treatment modalities include radiotherapy, surgery, cryotherapy and chemotherapy. Early diagnosis is necessary for best prognosis.

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