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Tonsillar Location of Skin Kaposi's Sarcoma: A Case Report

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Abstract | **Case Report**

We report a case of rare tonsillar location of Kaposi's sarcoma observed in a non infected HIV man, who had a long history of skin kaposi's disease during 28 years. Irradiation of skin lesions on lower limbs was succefully carried out with a dose of 30Gy on 10 fractions. Faced with the late appearance of dysphonia, we discover a locally advanced tonsillar location, and the PET- CT showed other distant supra and subdiaphragmatic lymph nodes involvements. After a first tracheotomy, the patient is treated by radiotherapy, 45 Gy on 25 fractions followed one month later by three cycles of chemotherapy based on Paclitaxel weekly regimen. We noted good locally reponse with no major toxicities after a three month follow up.

Keywords: Kaposi's sarcoma, Tonsillar location, Skin lesions, Radiotherapy.

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INTRODUCTION

Kaposi's sarcoma is an angioproliferative disease first described by Kaposi in 1872, primarily caused by human herpes virus 8 (HHV-8). In its classic form, it commonly affects individuals over 60 years of age and typically presents with cutaneous lesions located on the lower extremities, appearing as maculopapular or angiomatous nodules associated with lymphedema. Multifocal involvement has been documented, including lesions on the face, genital and oropharyngeal mucosa, as well as on the gastrointestinal and bronchial mucosa. Rare locations are often seen in patients with an immunocompromised status. While oropharyngeal involvement is known, laryngeal location is rare. We report a case of tonsillar Kaposi's sarcoma extending into the larynx and pharynx, diagnosed during the extensive stage of skin Kaposi's sarcoma in an HIV-negative patient, treated with radiotherapy followed by chemotherapy.

CASE REPORT

A 74-year-old male patient, with a Karnofsky score of 70, followed for arterial hypertension, presented with violaceous nodular skin lesions on the distal ends of both lower limbs. Some lesions were exophytic, bleeding, and ulcerated, particularly on the soles and toes (Figure 1). Other macular lesions were located on the hands and the left inguinal region. His medical history included a Kaposi hemangiosarcoma diagnosed 28 years earlier in another hospital; HIV serology was negative. He had undergone several local treatments including excisions, laser therapy, and cryotherapy, all followed by multiple relapses.

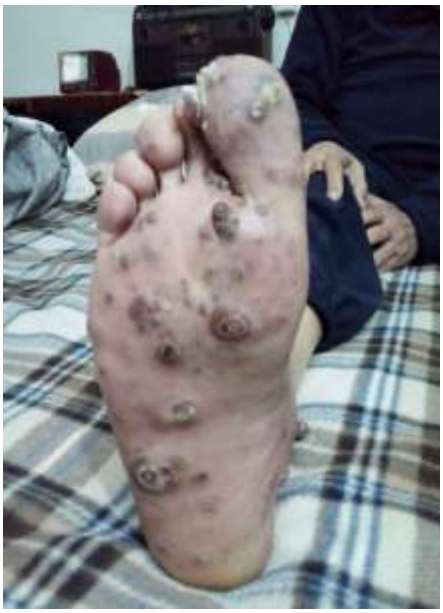

Figure 1: skin lesions before radiation therapy

Current excisional biopsy of a plantar nodule confirmed nodular-stage Kaposi's disease. Immunohistochemistry showed positivity for anti-CD34 and HHV-8 antibodies. An upper gastrointestinal endoscopy showed no additional lesions, and a thoracoabdominopelvic (TAP) CT scan revealed no suspicious images. Viral serologies for hepatitis B, C, and HIV remained negative. The patient was classified as having locally aggressive classic Kaposi's disease. He was treated with external radiotherapy : 30 Gy delivered in 10 fractions of 3 Gy, sequentially on each lower limb. During treatment, the distal end of the irradiated limb (from the knee to the toes) was immersed in a rice-filled tank and the patient seated in front of it on a chair positioned above the treatment table. The dose was prescribed to the isocenter and we used two parallel wide fields of 6 MV photon beams.

Acute side effects included significant and disabling lymphedema in both lower limbs, associated with vascular disturbances. Arterial and venous Doppler ultrasound found no signs of thrombosis. The patient's condition improved with antibiotic therapy, corticosteroids, and local care.

The left inguinal skin nodule was treated separately by complete surgical excision. After eight months, a complete response was observed in the lower limbs, though residual hyperpigmentation persisted (Figure 2). The patient gradually developed dysphonia and respiratory discomfort. Direct laryngoscopy revealed a left tonsillar tumor hypertrophy and a submucosal bulge in the glottic-subglottic area, causing severe narrowing of the laryngeal lumen.

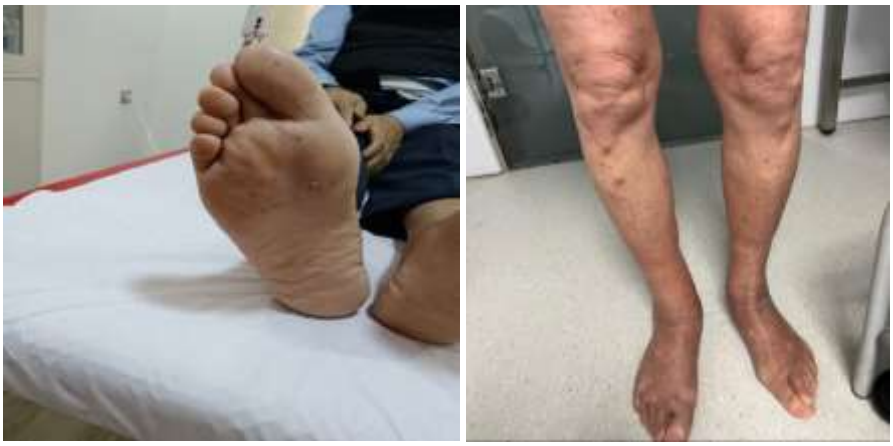

Figure 2 : After radiation therapy, complete response and sequelae of hyperpigmentation

Cervical MRI confirmed the presence of a locally advanced, exophytic tumor in the left tonsillar fossa measuring 57×47×91 mm, crossing the midline toward the right tonsillar fossa, infiltrating the soft palate, retropharyngeal fat, and bulging into the vocal cords, hypopharynx, and larynx, resulting in luminal narrowing. Bilateral laterocervical and supraclavicular lymphadenopathy were also present (Figure 3). Biopsy of the left tonsil confirmed the histologic and immunohistochemical features of nodular Kaposi's sarcoma.

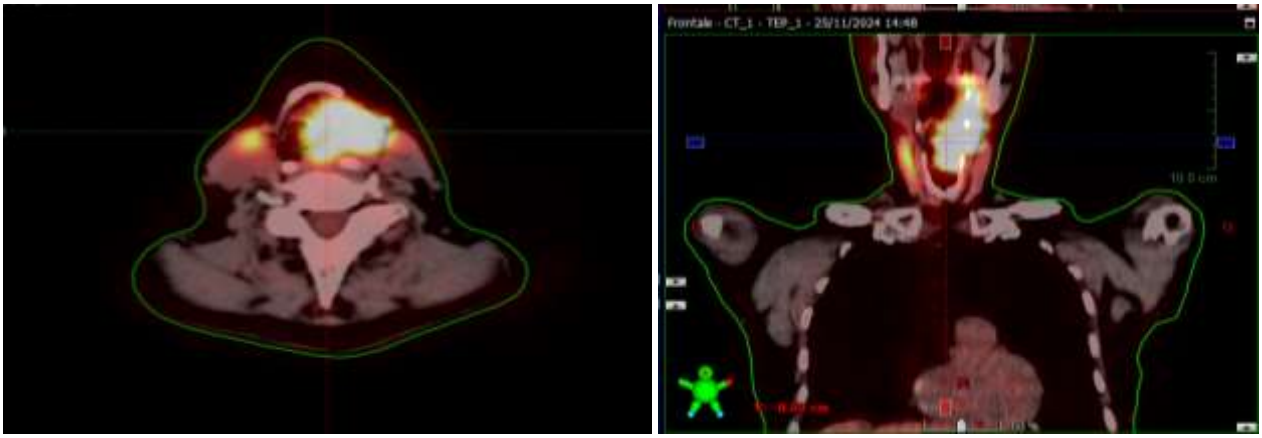

Figure 3 : Locally advanced left tonsillar involvement of kaposi's sarcoma

A PET-CT scan using 18F-FDG revealed, in addition to the left tonsillar mass, active pathological lymph node involvement both above and below the diaphragm, two hypermetabolic subcutaneous lesions, and another muscular lesion in the diaphragmatic pillar.

The patient underwent a tracheotomy prior to the start of radiotherapy. Cervical irradiation was delivered to a target volume including the PTV of the orophyarynx and larynx and the macroscopically involved cervical lymph nodes, using conformal arc therapy at a dose of 45 Gy in 25 fractions of 1.8 Gy. No prophylactic irradiation of adjacent nodal areas was performed (Figure 4). Radiotherapy was well tolerated; a grade 1 radiation dermatitis and mucositis were noted and managed with local treatment. The tracheostomy cannula was removed at the end of radiotherapy, with gradual return of the voice after completion of treatment.

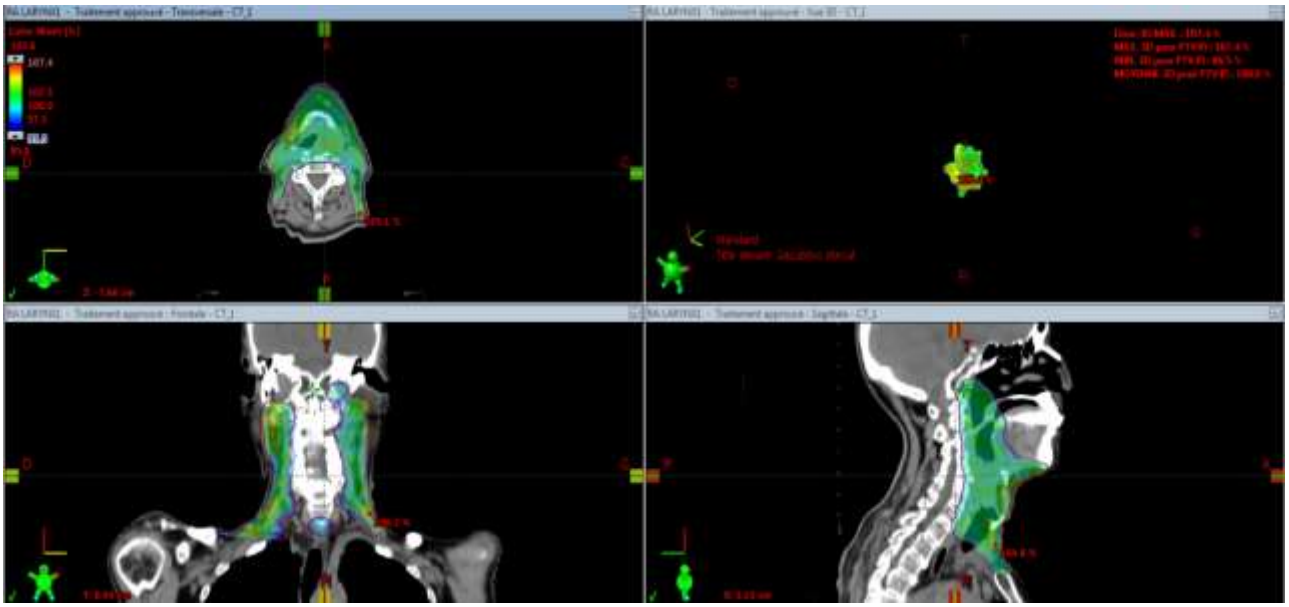

Figure 4: Planning radiation treatment with VMAT : 45Gy - 25 fractions - 1.8Gy/fr PTV include macroscopic tumor and nodes

One month later, as the new pattern of systemic kaposi's sarcoma, the patient received chemotherapy based on 3 cycles of Paclitaxel 60 mg/m² weekly on days 1, 8, and 15, with one week of rest after each cycle. Follow-up by thoraco-abdomino-pelvic CT scan showed a partial response in the left tonsillar mass and supradiaphragmatic and subdiaphragmatic nodes. The patient remained in partial clinical and radiological remission three months after treatment.

DISCUSSION

Kaposi's sarcoma (KS) is classically divided into four epidemiological subtypes:

1. Classic KS seen in elderly individuals of Mediterranean, Central, or Eastern European origin, primarily localized to the extremities, and characterized by a slow progression
2. Epidemic KS, associated with HIV-positive individuals
3. Iatrogenic KS, found in transplant patients under immunosuppressive therapy, which can be extensive and fatal Endemic (African) KS, affecting younger individuals in sub-Saharan Africa characterized by nodular, exophytic, or infiltrative lesions, sometimes associated with lymphoma.

Clinically, Kaposi's sarcoma often presents as violaceous or reddish-blue lesions on the skin or mucous membranes. The cutaneous lesions may appear as macules, plaques or nodules of varying size, which can ulcerate, bleed, or become verrucous and hyperkeratotic. Lymphedema and lymphadenopathy may also be present. Visceral Kaposi's disease can affect the respiratory and gastrointestinal tracts. Oropharyngeal lesions may lead to potentially fatal laryngeal obstruction. Dermoscopy can be useful in identifying the classic colors of vascular tumors.

Despite clinical and evolutionary differences among the four epidemiological subtypes of Kaposi's sarcoma, the histological appearance remains the same with a progressive evolution from the macular stage to the plaque stage and finally to the nodular stage. Characteristic findings include the simultaneous presence of spindle cells, vascular structures, and lymphoplasmacytic elements. Our patient was first descipted with skin Kaposi's angiosarcoma in 1996. The current excisional biopsy of the skin lesion and the tonsillar specimen revealed the same fundamental histopathological features consistent with nodular-stage Kaposi's disease. Immunohistochemistry confirmed the diagnosis in both recent biopsies using the ABC-peroxidase method on deparaffinized sections, showing strong expression of the CD34 marker (an endothelial marker of spindle cells) and for the monoclonal antibody against HHV-8 (LANA or Latent Nuclear Antigen), whose sensitivity and specificity have been demonstrated in several studies [1,2].

Only one classification is validated for KS associated with HIV, including the extent of the disease, localized or extensive, the severity of immunodeficiency assessed by CD4 cell count, and the presence or absence of clinical symptoms. Three situations are distinguished: non-aggressive localized, locally aggressive, and extensive KS [3].

Another classification was developed by an Italian group in 2003 involving 300 patients with classic KS. Four stages are identified, from stage I to IV, based on whether the lesions are maculo-nodular, infiltrative, florid, or disseminated. Local or systemic treatment recommendations are decided according to the clinical stage and radiological findings (total body CT scan) in disseminated stages [4].

The assessment of disease extent in non-HIV-associated Kaposi's Sarcoma should be discussed individually according to clinical symptoms and lesion extent. A table has been proposed by the EDF, EADO, and EORTC groups, which includes, depending on the KS subtype, whether to perform a total body CT scan, bronchoscopy, or gastrointestinal endoscopy : a physical examination, histological study, HIV serology, and standard laboratory tests are routine regardless of the subgroup; HHV-8 viral load and CD4 count are requested in cases of KS with HIV or KS after transplantation. A total body CT scan, bronchoscopy, and digestive fibroscopy are requested depending on symptomatology in patients with classic or endemic KS [5].

In our case, given the long clinical history of KS and the extent of skin lesions, we deemed it useful to request a thoraco-abdomino-pelvic (TAP) CT scan combined with digestive edoscopy before initiating specialized treatment.

FDG PET scan should be part of the staging workup for KS at risk of visceral or musculoskeletal dissemination; performed before or after routine staging, it can help guide radiological explorations or detect other occult lesions. The sensitivity for detecting Kaposi's lesions can reach 71% with 98% specificity, except in cases of digestive involvement [6].

In the classical variant of KS, mucosal involvement is rare (≤5%), and visceral or lymph node involvement occurs in less than 10% of cases. Tonsillar involvement is reported in 8.3% of head and neck cases, while laryngeal involvement occurs in 5% of cases [7,8,9,10,11]. Our patient developed mucosal involvement of the tonsils as well as supradiaphragmatic and subdiaphragmatic lymph node involvement during disease dissemination.

The goal of KS treatment is not to cure but rather to control the disease and its symptoms, provide patient relief, and preserve quality of life. The choice of treatment type depends on the clinical context, the extent and location of lesions, as well as the epidemiological subtype.

Local treatments using laser or cryotherapy are indicated for superficial lesions, with a response rate of 80 to 90% [12] and a risk of residual hypopigmentation. Our patient experienced a long period of local response with dermatological therapies (28 years) before progressing to a more aggressive but localized stage.

Surgical excision of a few superficial and well-defined Kaposi's nodules is possible but should not be recommended for extensive lesions due to a high risk of recurrence and severe functional sequelae.

Local or intralesional chemotherapy in localized stages has the advantage of allowing a high concentration of drug (Vinblastine or Bleomycin) at the lesion site without causing systemic side effects. A local response rate of 70% has been reported for Vinblastine and 65 to 89% for Bleomycin [5].

Radiotherapy is a major treatment modality for Kaposi's Sarcoma, indicated for skin and/or mucosal lesions, either in solitary form to address sometimes unacceptable cosmetic concerns, or in extensive form with palliative intent in cases of painful, weeping or bleeding lesions [13].

The optimal dose is not clearly defined, but KS is sensitive to moderate doses of radiation. This sensitivity is consistent across the four epidemiological forms: classical, endemic, HIV-associated, or post-transplantation [14].

Recommended doses range from 30 to 36 Gy, delivered in fractions of 2 to 3 Gy according to guidelines from three associations: The European Dermatology Forum (EDF), The European Association of Dermato-Oncology (EADO), and The European Organization for Research and Treatment of Cancer (EORTC), using low-energy photons or electrons. High fraction doses (>3 Gy) and concurrent chemotherapy during irradiation are not recommended. Response rates vary from 47% to 99% depending on the series [5].

Other radiation therapy regimen's have been described: 8 Gy in a single fraction and 16 Gy in 4 fractions over 4 days, with no statistically significant difference in overall response in a prospective study treating 596 lesions [15]. Doses greater than 20 Gy are associated with higher response rates, longer local control duration, and fewer side effects, particularly hyperpigmentation [16].

Two randomized trials compared different fractionation schemes in HIV-positive patients with KS : 24 Gy in 12 fractions versus 20 Gy in 5 fractions over 64 lesions, with no difference in relapse-free survival, local

response, or toxicity [17]. However, caution is advised in cases of very extensive lesions and severe lymphedema due to a high risk of necrosis and ulceration.

The second trial [13] compared three arms: 40 Gy in 20 fractions, 20 Gy in 10 fractions, and 8 Gy in a single fraction. There were more complete responses in the 40 Gy and 20 Gy arms compared to the 8 Gy arm: 83%, 79%, and 50% respectively ($p = 0.04$), and fewer local relapses with the first two protocols: 52% and 67% respectively versus 88% with the single 8 Gy fraction protocol ($p = 0.003$).

Radiotherapy is generally well tolerated. Acute side effects are more pronounced when large radiation fields and high fraction doses are used. Acute mucositis is more common in oropharyngeal involvement. Late side effects may include hyperpigmentation in 55% of cases [13], as well as telangiectasia, skin atrophy, or skin fibrosis.

The choice of radiotherapy regimen for skin lesions should be individualized based on the patient's general condition, the severity and extent of the lesions. A short-course regimen may be preferred to relieve pronounced symptoms such as pain, oozing, or bleeding in patients with advanced KS. Conversely, a longer regimen with lower fractionation may be appropriate for patients in good general health, treated with cosmetic intent or when large radiation fields are involved [18].

In the management of KS with pharyngo-laryngeal involment, radiotherapy is valuable for reduction of clinical symptoms and reducing tumor size. A dose of 40 Gy in 20 fractions has been used in a primary nasopharyngeal lesion [19], while other authors suggest 45 Gy (1.8 Gy per fraction) for laryngeal involvement [20]. A conformal radiotherapy technique using IMRT (Intensity-Modulated Radiation Therapy) is recommended [21]. The radiation fields should include the tumor volume (T) and only the involved lymph nodes; there is no benefit in prophylactically treating uninvolved lymph nodes.

Laryngeal involvement of KS can cause airway obstruction, potentially requiring emergency intervention via tracheostomy or even cricothyrotomy [20]. For our patient, we considered it essential to perform an initial tracheostomy before starting radiotherapy to avoid obstruction during treatment.

HDR brachytherapy is a good alternative for treating localized and superficial forms of KS, whether cutaneous or mucosal, offering complete responses and low toxicity. A regimen of three sessions of 8 Gy has been proposed [18].

Paclitaxel-based chemotherapy in disseminated forms of KS has been tested in HIV-negative patients using various protocols : 100 mg as a weekly dose for 12 weeks, or 175 mg/m² every 3 weeks. The protocol of 80 mg/m² either continuously or 3 weeks out of 4 is commonly used and well tolerated. Response rates of up to 80% have been reported. Liposomal anthracyclines remain the first-line treatment for disseminated KS if cardiac function permits, at a dose of 20 mg/m² on days 1 and 21, with response durations of up to 25 months and acceptable toxicity : Grade 4 neutropenia in 5% and hand-foot syndrome in 5% (5). In our KS case's, the weekly paclitaxel protocol was chosen due to the unavailability of liposomal doxorubicin. The patient remains in partial clinical and radiological remission three months after treatment.

A 2024 review article [22], initially including 574 articles focused exclusively on classic KS, developed a therapeutic algorithm based on disease stage with clinical and imaging assessment and incorporating in addition to radiotherapy and chemotherapy, new agents such as immunotherapy, anti-angiogenic agents, CDK inhibitors, and other targeted therapies currently under investigation.

Conclusion

Tonsillar involvement in classic Kaposi's sarcoma is rare but may occur as an initial presentation or during disease dissemination. Moderate-dose radiotherapy can significantly improve symptoms and yield good local control. Radiotherapy should be integrated into the treatment plan even during the disseminated phase, separate from chemotherapy.

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