Journal Homepage: - www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/15739
DOI URL: http://dx.doi.org/10.21474/IJAR01/15739

RESEARCH ARTICLE

LOCALLY ADVANCED ORAL KAPOSI'S SARCOMA IN AN HIV NEGATIVE WOMAN TREATED WITH EXCLUSIVE RADIOTHERAPY: CASE REPORT

B. Assenhaji Louizi[1], M. T. Saoudi[2], A. Uakkas[1], M. Ait Erraisse[1], W. Hassani[1], FZ. Farhane[1], Z. Alami[1] and T. Bouhafa[1]

1. Department of Radiotherapy and Brachytherapy, Hospital of Oncology, CHU Hassan II, Fez, Morocco.
2. Medical Oncology Department, Oncology Hospital, CHU Hassan II, Fez, Morocco.

Manuscript Info

Manuscript History
Received: 25 September 2022
Final Accepted: 27 October 2022
Published: November 2022

Key words:-
Oral Kaposi 's sarcoma, Radiotherapy In Classic Kaposi's Sarcoma, Case Report

Abstract

The occurrence of oral Kaposi's sarcoma (KS) in HIV-seronegative people is extremely rare. Mucosal lesions in VIH seronegative patients have been reported, making it a challenging diagnosis to identify and manage. Here, we present a 54-year-old seronegative patient who underwent radiation therapy as the only course of treatment for a locally advanced palatal mucosal kaposi sarcoma. The outcomes of the treatment in terms of tumor control and the immediate relief of our patient's symptoms illustrate its effectiveness and support kaposi sarcoma radiosensitivity.

Introduction:-

Moritz Kaposi originally described Kaposi's sarcoma (KS) as vascular mucocutaneous tumor of endothelial origin, in 1872 (4). There are four clinical subtypes of the disease, including classic, endemic, iatrogenic, and epidemic KS, each having its own natural history, site of predilection, and a direct connection with Human immunodeficiency virus (HIV) and Human herpesvirus-8 (HHV-8). (3)

Oropharyngeal involvement is unusual in classic KS cases with negative VIH serology as it might be the initial symptom of immunodeficiency (3). Oral KS typically affects the tongue, gingiva, and hard palate. Clinical manifestations can involve a variety of solitary or multifocal plaques and nodules of red-purplish color that grow gradually and possibly invade alveolar bone (3, 4).

Classic KS is generally considered to be sensitive to radiation therapy ensuring rapid symptom palliation and good esthetic results with doses exceeding 20 Gy (5, 6).

Case presentation:

A 54 years old woman consulted a maxillofacial surgeon after a year of neglecting a lesion in the hard palate that was gradually growing in size, until it began to interfere with mastication, speech, sleep quality and oral hygiene. The patient was seronegative and had no risky sexual behavior. She had no medical history or drugs intake.

The clinical inspection revealed a left jugal swelling deforming the face with no skin lesion (Figure 1.a). The intraoral examination objectified a large exophytic purplish mass of the hard palate, friable and infected with yellow deposit. Anteriorly, the lesion invaded the left gingival mucosa damaging the teeth (figure 1.b). Posteriorly, it invaded the hard palate, crossing the midline and reaching the soft palate (figure 1.b)

Corresponding Author: - B. Assenhaji Louizi
Address**:-** Department of Radiotherapy and Brachytherapy, Hospital of Oncology, CHU Hassan II, Fez, Morocco.

The patient had an extremely poor oral health, halitosis and was experiencing moderate trismus.

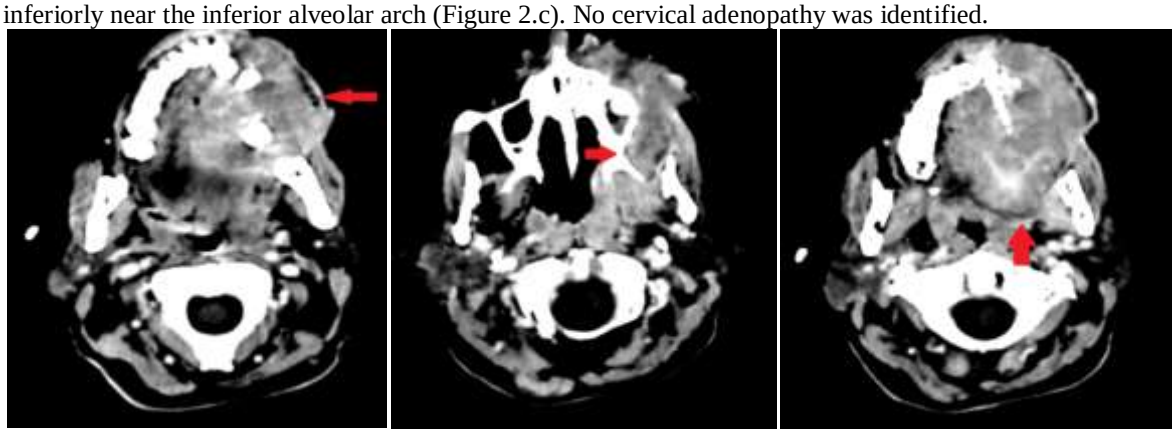
There was no adenopathy detected in the cervical lymph nodes examination

(a) **(b)**

Figure 1:- Clinical examination of the patient showing a left jugal swelling (a) with, at the oral examination, a tumoral process of the hard palate invading the gingival mucosa (b)

Anatomo-pathological analysis of the tumor biopsy revealed sarcomatous spindle cell proliferation CD20+, CD30+, with the presence of few atypical cells. The identification of HHV-8 infection in the immunohistochemical study supported the final dignosis of oral Kaposi's sarcoma. All cells were CD40 positive and CK (AE1/AE3), AML, and Psl00 negative

A maxillary and cervical computed tomography revealed the presence of a heterogeneous tumor mass on the left upper alveolar arch , with dimensions of 75x61x92 mm , spontaneously hypodense with irregular poorly-identified contours. Jugal soft tissues were deeply infiltrated anteriorly (Figure 2.a), reaching the symphyseal area, medially the mass infiltrates the floor of the mouth. Superiorly it invades the hard palate with bone lysis and erosion of the floor of the left maxillary sinus (Figure 2.b), posteriorly it infiltrates the parapharyngeal space and arrives inferiorly near the inferior alveolar arch (Figure 2.c). No cervical adenopathy was identified.



(a) (b) (c)

Figure 2:- computed tomography (CT) scan showing locally advanced oral kaposi's sarcoma invading Jugal soft tissues and upper left alveolar arch (a), with maxillary bone lysis (b), arriving to parapharyngeal space and the floor of the mouth (c).

The results of new blood tests were also negative for both HIV and HCV supporting the diagnosis of HIV-negative classic form of this disease. A total blood count showed normal results as well as a gastric endoscopy.

The case was discussed in a multidisciplinary board, and the lesion has been judged non-resectable, hence the indication for local treatment by radiotherapy with no systemic therapy.

The patient received intensity-modulated external beam radiation therapy with a total dose of 30Gy in 10 fractions, 3Gy per fraction, 5 over 7 days. On an injected dosimetric CT scan, the gross tumor volume was delineated, and a 5 mm expansion was performed as a margin to determine the CTV. In order to acquire the PTV while taking into account a thermoformed mask's contention, a 3 to 5mm margin was added to the CTV.

No acute radiation-induced toxicity was noticed during the treatment period. Bicarbonate mouthwashes were prescribed to avoid mucositis for the whole course of radiotherapy and for 15 days afterward.

One month after the completion of radiotherapy, the clinical control revealed a considerable reduction in the patient's jugal deformation (Figure 3.c). at the oral cavity examination, when compared to the original aspect, the maxillary process has significantly regressed, becoming less infected, smooth, and quite well-delimited (figure 3.a,b), with improvements in trismus, breath, and speech. At Three months post-treatment, the tumor is still shrinking, currently measuring 2 cm in major axis, the patient remains asymptomatic.

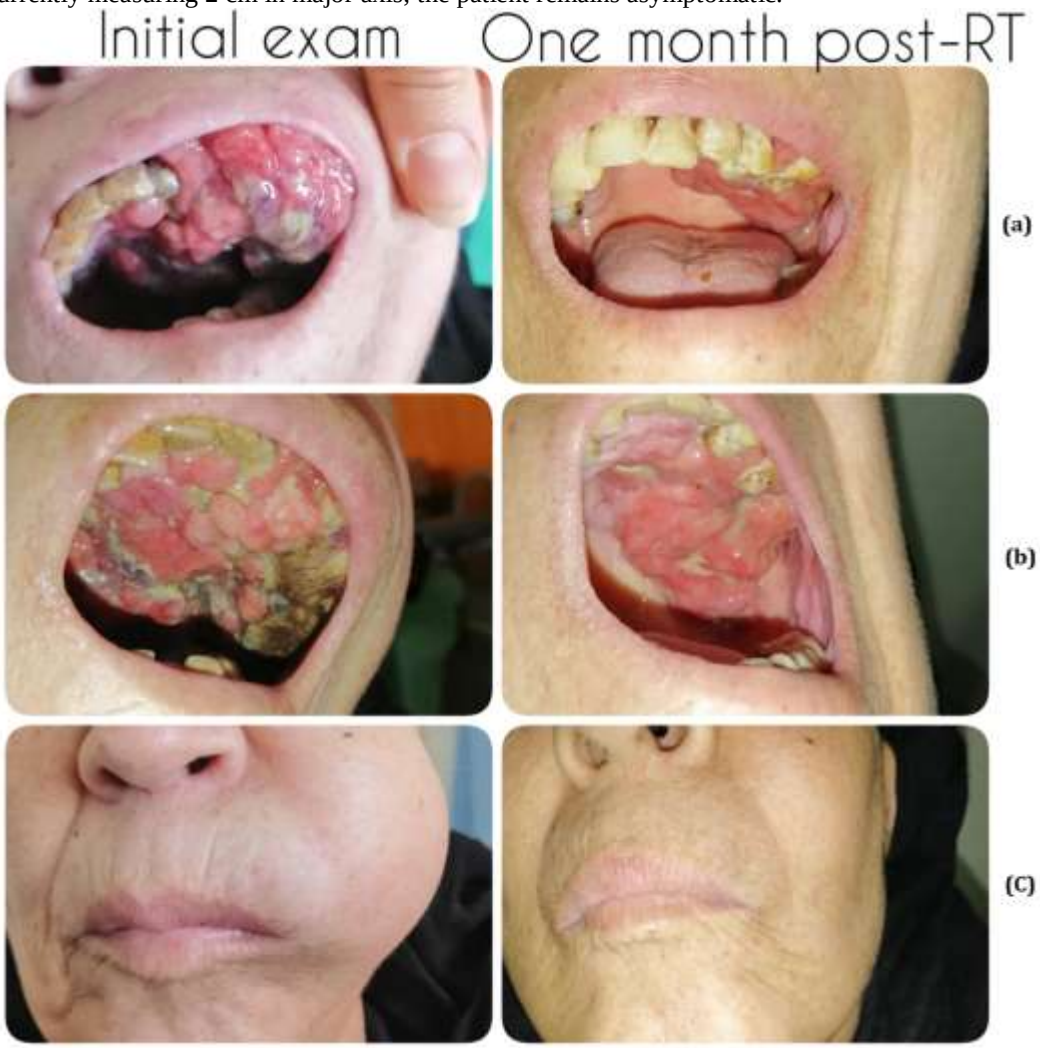


Figure 3:- Non-resectable oral kaposi sarcoma treated with radiotherapy has remarkably reduced in clinical size one month after the completion of treatment.

Discussion:-

Kaposi's sarcoma (KS) is an angio-proliferative, mucocutaneous malignant neoplasm. it is classified as an opportunistic HIV-related tumor (human immunodeficiency virus). Risk factors may also include the human herpesvirus (HHV) 8, immunosuppression, the use of angiotensin-converting enzyme inhibitors or antimalarial therapies. (1,2)

There are four clinical variants of Kaposi's sarcoma, each of which has different clinical, sociodemographic, and histological characteristics. (3)

Classic KS is mainly cutaneous affecting the lower extremities of eldery males (>60) of Mediterranean, Eastern Europe, Jewish, and South American origin. Major risk factors are advanced age and HHV-8. It mainly appears as slowly-growing patchy pigmented skin lesions in the lower limbs, and rarely involves the mucosa, viscera, or lymph nodes. (5)

Endemic KS was first discovered in HIV negative children and young men from Central and South Africa. This variant is characterized by HHV-8 incrimination, agressive progression, and common lymphatic and visceral involvement, causing severe systemic disorders. (18)

Epidemic KS is an aggressive HIV-related KS subtype that is mainly observed in homosexual males (7). Involvement of the viscera, lymph nodes, and mucosa in addition to cutaneous lesions are frequently seen. (8) Antiretroviral treatments have been shown to be effective in decreasing the prevalence and severity of KS. (9,10)

Iatrogenic KS is mostly induced by long-term immunosuppressive therapies for autoimmune disorders or after solid organ transplantation. (7) Corticosteroids may also increase the susceptibility to IKS. Clinical forms are rarely agressive, with an equal prevalence of cutaneous, mucosal, lymphatic and visceral involevement. (11)

Oral cavity involvement may be seen in all four variants, although AIDS-KS association is the most common situation. (2) it has been strongly related to human herpes virus type 8 (HHV-8) which replicates and remains latent in oral cavity's endothelial and B cells, up until the emergence of an immunodepressive condition.(12) The most common sites for oral Kaposi Sarcoma are hard and soft palates, gingiva, and dorsum of the tongue. (3, 13) lesions may be solitary, multipls oral lesions or associated with cutaneous and/or visceral forms. (2,3)

The clinical presentation includes pink purplish macules progressing secondarily to plaques or nodular then exophytic lesions.(2) Patches of Oral KS are asymptomatic, whereas exophytic KS can cause irritation, bleeding, ulceration, or infection. Exophytic gingival KS may also interfere with mastication and oral hygiene, or induce tooth instability and facial deformity. (1, 3)

All KS variants have same microscopic features, including abundant proliferating mononuclear inflammatory, spindle cells and poorly bounded anarchic vessels. The absence of smooth muscle cells called pericytes in newly formed arteries can lead to erythrocytes extravasation and hemorrhage. (15, 16) The clinical progression is associated to a histopathological transition from focally proliferative miniature vessels in the early patch stage, to tumor-like fascicles of an anarchic vascular network with spindle cells and abnormal extravasated erythrocytes. (1, 2, 15)

Herpes virus DNA can be detected by PCR in order to distinguish KS from other clinically similar lesions; however it is also possible to isolate the herpesvirus nuclear antigen related to the latency (LANA) in the immunohistochemical analysis. (17)

Multiple modalities are considered for oral KS treatment, depending on the clinical form and the staging, including intralesional interferon alpha, cryotherapy, laser removal, infrared coagulation, radiation, and systemic therapy.(2,5)

Oral KS is highly radiosensitive; hence radiation therapy is widely suggested as a local treatment with a great effectiveness in symptom relief and tumor control. However, there is no approved standard for RT dose or technique. Regimens range from a single 8-Gy dose to a total dose of 30 Gy delivered in 10 to 15 fractions. Fractional regimens are suggested in mucosal lesions with large irradiated volume. (19-20-21)

Yet, radiation-induced oral toxicities like mucositis, taste alterations, and oral infections usually appear, needing prophylactic and palliative measures. (22)

Since oral KS in seronegative patients is a rare neoplasm, it can be challenging to identify its occurrence. It is noteworthy that all KS subtypes can be effectively and safely treated with RT, and all RT schemes have shown great control rates and symptom relief. (10-21-22)

References:-

1. Nicolas Dupin, Aude Jary ,Samia Boussouar ,Charlotte Syrykh ,Amir Gandjbakhche,Sébastien Bergeret ;Romain Palich Current and Future Tools for Diagnosis of Kaposi's Sarcoma. Cancers 2021, 13(23), 5927; https://doi.org/10.3390/cancers13235927
2. Mahnaz Fatahzadeh, DMD, MSD, and Robert A. Schwartz, MD, MPH, FRCP (Edin). Oral Kaposi's sarcoma: a review and update . International Journal of Dermatology 2013, 52, 666–672
3. Fatahzadeh, M., 2012. Kaposi sarcoma: review and medical management update. Oral Surg. Oral Med. Oral Pathol. Oral Radiol. 113, 2–16.
4. Kaposi M. Idiopathisches multiples Pigmentsarkom der Haut. Arch Dermatol Syph (Prague) 1872;4:265-73.
5. Régnier-Rosencher E, Guillot B, Dupin N. Treatments for classic Kaposi sarcoma: A systematic review of the literature. J Am Acad Dermatol 2013;68(2):313–311.
6. Akmansu M, Goksel F, Erpolat OP, Unsal D, Karahacioglu E, Bora H. The palliative radiotherapy of classic Kaposi's sarcoma of foot region: retrospective evaluation. Int J Hematol Oncol. 2014;24:147–52.
7. Curtiss P, Strazzulla LC, Friedman-Kien AE. An update on Kaposi's sarcoma: Epidemiology, pathogenesis and treatment. Dermatol Ther (Heidelb) 2016;6(4):465– 70.
8. Hoffmann C, Sabranski M, Esser S. HIV-associated Kaposi's sarcoma. Oncol Res Treat 2017;40(3):94–8.
9. Dalla Pria A, Pinato DJ, Bracchi M, Bower M. Recent advances in HIV-associated Kaposi sarcoma. F1000Res 2019;8: F1000 Faculty Rev–970.
10. Kalpidis, C.D.R., Lysitsa, S.N., Lombardi, T., Kolokotronis, A.E., Antoniades, D.Z., Samson, J., 2006. Gingival involvement in a case series of patients with acquired immunodeficiency syndromerelated Kaposi sarcoma. J. Periodontol. 77, 523–533. http://dx. doi.org/10.1902/jop.2006.050226.
11. Rabate C. Prise en charge du sarcome de Kaposi posttransplantation rénale en ile de France: Analyse rétrospective de 89 patients. Ann Dermatol Venereol 2013;140:404.
12. Dittmer, D.P., 2011. Restricted Kaposi's Sarcoma (KS) Herpesvirus Transcription in KS Lesions from Patients on Successful Antiretroviral Therapy. Am. Soc. Microbiol. 2 (e00138-11- e00138-11).
13. Patrikidou, A., Vahtsevanos, K., Charalambidou, M., Valeri, R., Xirou, P.K.A., 2009. Non-AIDS Kaposi's sarcoma in the head and neck area. Head Neck 31, 260–268
14. İpek Pınar ARAL, Gonca ALTINIŞIK İNAN, Süheyla AYTAÇ ARSLAN, Selcan TEKİN, Neşe GÖÇER GÜROK, Alaettin ARSLAN, Yılmaz TEZCAN. Role of Radiotherapy in Kaposi's Sarcoma: Review of the Literature. Turk J Oncol 2021;36(3):389–400 doi: 10.5505/tjo.2021.2563
15. Tappero JW, Conant MA, Wolfe SF, Berger TG. Kaposi's sarcoma. Epidemiology, pathogenesis, histology, clinical spectrum, staging criteria and therapy. J Am Acad Dermatol 1993;28:371-95.
16. Kang T, Ye FC, Gao SJ, Wang LD. Angiogenesis, Kaposi's sarcoma and Kaposi's sarcoma-associated herpesvirus. Virol Sin 2008;123:499 –58
17. Feller L, Lemmer J, Wood NH, Jadwat Y, Raubenheimer EJ. HIV-associated oral Kaposi sarcoma and HHV-8: a review. J Int Acad Periodontol 2007;9:129-36.
18. Slavin G, Cameron HM, Forbes C, Mitchell RM. Kaposi's sarcoma in East African children: A report of 51 cases. J Pathol 1970;100(3):187–99.
19. Ramirez K, Zavala J, Moran D, Hernandez D, Jimenez A. Classic Kaposi's sarcoma: complete response to radiation therapy: a case report. J Med Case Rep 2016;10:322.
20. Teke F, Akkurt Z. Evaluation of 14 patients performed radiotherapy due to Kaposi sarcoma. Dicle Med J 2015;42:326-30.
21. Gonca Altinisik Inan, Ipek Pinar Aral, Suheyla Aytac Arslan, Yilmaz Tezcan. Palliative treatment of Kaposi sarcoma with radiotherapy: a single center experience. Radiat Oncol J 2021;39(1):41-47.
22. Housri N, Yarchoan R, Kaushal A. Radiotherapy for patients with the human immunodeficiency virus: are special precautions necessary? Cancer 2010; 116: 273– 283.