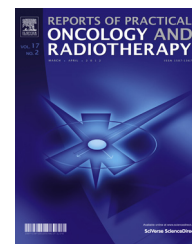


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Case report

A Kindler syndrome-associated squamous cell carcinoma treated with radiotherapy



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ABSTRACT

Kindler syndrome^{1,2} is a genetic disorder mainly characterized by increased skin fragility and photosensitivity,^{3,4} making the use of treatments based on radiation difficult or even prohibited. Thus, cases reporting Kindler syndrome patients treated with radiotherapy are rare. In this study, we report clinical outcomes and care provided for a rare case of a Kindler syndrome patient submitted to radiotherapy. Diagnosed with squamous cell carcinoma involving the buccal mucosa, the patient was exclusively treated with radiotherapy, with 70 Gy delivered on the PTV with the Volumetric Modulated Arc technique. The patient's reaction regarding control of the lesion is relevant compared to patients not affected by the syndrome. We noticed acute reactions of the skin and buccal mucosa after few radiotherapy sessions, followed by a fast reduction in the tumor volume. The efficacy of radiotherapy along with multidisciplinary actions allowed treatment continuity, leading to a complete control of the lesion and life quality improvement and showed that the use of radiotherapy on Kindler syndrome patients is possible.

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1. Case representation

A female patient, previously diagnosed with Kindler syndrome, presented with an initial lesion on the right cheek,

believed to be due to the syndrome. The lesion was, however, diagnosed as squamous cell carcinoma involving buccal mucosa. Six months after the initial complaint, the disease had already extended to the skin of upper and lower right lips and jugal mucosa. Due to the condition

Abbreviations: KS, Kindler syndrome; GTV, gross tumor volume; CTV, clinical tumor volume; PTV, planning tumor volume; HDMLC, high definition multileaf collimator; OAR, organs at risk; PRV, planning risk volume; QUANTEC, quantitative analysis of normal tissue effects in the clinic; NTO, normal tissue optimization; RTOG, Therapy Oncology Group.

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of the lesion, radiotherapy was ruled the best treatment option. However, radiotherapy represents a challenge to head and neck cancer patients with Kindler syndrome due to their characteristic photosensitivity and to unknown consequences of high energy exposure. By the end of the treatment, the external visible tumor mass resolved completely with controlled toxicity of epithelitis and mucositis.

2. Introduction

Radiotherapy applied to head and neck pathologies yields good responses regarding local control of the disease and toxicity on surrounding tissues. It was shown efficient, both exclusively and combined with chemotherapy or surgery, in several oral cavity tumor studies in patients with favorable conditions.^{5–7} However, there are few results described from patients affected by syndromes that hinder the application of radiotherapy, such as Kindler syndrome (KS).⁸ These patients present increased skin and organ toxicity in response to radiation due to photosensitivity, preventing the use of radiotherapy. However, although usually not indicated, radiotherapy can sometimes be the only treatment option for KS patients with head and neck tumors. In this work, we describe the use of radiotherapy to treat an oral cavity tumor on a KS patient. We also discuss its evolution and care provided to allow the treatment.

The patient was diagnosed with a primary lesion on the oral cavity, on the jugal mucosa, with oro-cutaneous fistula, and bone invasion detected on tomography. The lesion was previously assessed by the surgical team and deemed unresectable and at high risk of complications with chemotherapy, which would have only a palliative role if exclusive. Therefore, in a multidisciplinary assessment, exclusive radiotherapy was chosen as a potentially curative option. Radiation was applied in 33 fractions, with a total dose of 70Gy on the lesion primary volume and 54Gy on lymph drainage. The Volumetric Modulated Arc Therapy was used to deliver conformational doses to the tumor volume with high gradient on the boundaries between the lesion and healthy tissues to reduce skin toxicity.^{9–11}

The treatment definition demanded a multidisciplinary team effort, including weekly reviews with the radiation oncologist, monitoring by the dental team with 560 nm laser applications, and adequate nutrition by tube feeding due to the esophageal stenosis caused by the syndrome.

During treatment, we observed a reduction in the tumor mass and acute toxicity, with epithelitis and mucositis, reaching Grade I according to the RTOG¹² study on week two, evolving to Grade II throughout treatment, affecting mainly the primary lesion surroundings and lips. Local response is observed on week one, with a tumor regression of 10%, followed by relief of neoplasia-related pain. Fistula improved continuously until full response by the end of week seven, showing that radiotherapy is a viable prescription, even in KS patients.



Fig. 1 – Status of the lesion before treatment. Fissure on right cheek showing the tumor extending to the lips, and blisters on the lips caused by Kindler syndrome.

2.1. Reported case

A 55-year-old female patient, with no history of smoking, previous or current, in good general condition (ECOG performance status 1) and tumor staging cT4cN0M0. Before treatment, the lesion was ulcerated, with oro-cutaneous fistula, affecting the skin of upper and lower right lips, labial commissure, and right jugal mucosa. The patient also presented skin alterations such as fibrosis, cutaneous atrophy and bullous lesions on the oral mucosa and lips, as shown in Fig. 1. The patient also has a history of esophageal stenosis previously treated with endoscopic dilation.

The patient was diagnosed with KS, a type of autosomal recessive epidermolysis bullosa, characterized by blisters on the skin, photosensitivity, progressive poikiloderma, and extensive skin atrophy.^{1–4,13} The oral cavity is frequently affected and common aspects include: microstomia, tongue mobility abnormalities, ulcerations, and occurrence of malignant tumors from the metaplasia of ulcerations. In addition to cutaneous involvement, the illness can also affect the mucosa, worsening progressively with age, and can cause dental alterations such as gingivitis and periodontitis. Extra-cutaneous affections such as esophageal stenosis, colitis and other gastrointestinal affections are also present. However, the main reason for concern is patient's photosensitivity and skin fragility upon high energy radiation exposure whose consequences are still unknown.

The patient was submitted to computerized tomography for the reconstruction of risk and treatment volumes. GTV₁ delimitation corresponds to the primary tumor volume boundaries (43.1 cm³). Clinical CTV₁ (138.8 cm³) and planning volume PTV₁ (192.3 cm³) are estimated from GTV₁ expansion, with margins of 1 cm and 1.4 cm, respectively.¹⁴ Lymph node chains are associated to CTV₂ and PTV₂ relative to the elective treatment chains, at Level IA/B, IIA/B, III, IV. Level V and retrofaryngeal chain were, thus, excluded because they are considered a low risk of commitment by the radiation oncologists.

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