Case Report

Radiation therapy for a rare association of maxillary neoplasm in xeroderma pigmentosum: Is it really contraindicated?

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ABSTRACT

Dermatologic malignancies are common in xeroderma pigmentosum (XP) patients; they can develop maxillary sinus cancers on rare occasions. Despite their extreme sensitivity to ultraviolet light, the patients of XP can be treated with standard doses of ionizing radiation for the treatment of cancers. The examples of use of radiotherapy as a treatment modality for maxillary neoplasms in patients of XP are rare. This report highlights a rare association of maxillary carcinoma in a patient of XP who received the tumoricidal doses of therapeutic X-rays with acceptable toxicities.

Key words: Ionizing radiation, maxillary sinus carcinoma, toxicities, xeroderma pigmentosum

INTRODUCTION

Xeroderma pigmentosum (XP) is a hereditary autosomal recessive disorder characterized by mucocutaneous and ocular hypersensitivity to ultraviolet (UV) radiation with irreversible DNA damage and subsequent malignancies. The basic defect in XP is in nucleotide excision repair (NER), leading to deficient repair of DNA damaged by UV radiation. There are eight complementation groups identified from XPA to XPG and a variant form XP-V. These patients frequently develop a variety of skin cancers such as basal cell carcinoma, squamous cell carcinoma, and melanoma. This case report illustrates the rare occurrence of maxillary carcinoma in a young patient of XP. It also highlights the fact that even though the patients of XP are sensitive to the ionizing effects of UV rays, use of therapeutic X-rays in the form of external beam radiotherapy (EBRT) can be a viable option in treating locally advanced maxillary cancers.

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Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.186110

CASE REPORT

A 26-year-old male, farmer by occupation and resident of Mysore district, Karnataka, India, was referred to the Department of Radiation Oncology, Victoria Hospital, Bengaluru, Karnataka, India. He had chief complaints of right-sided headache, swelling on the right side of the face, constant watering, and diminished vision of right eye since 3 months. Patient had been operated for recurrent skin malignancies of right nasal ala in past. He underwent wide local excision for basal cell carcinoma 4 years back and wide local excision with nasolabial flap reconstruction for dermatofibrosarcoma 1 year later.

On physical examination, there was diffuse freckling with multiple hyper- and hypo-pigmented macules on the face, neck, and extremities along with proptosis and exposure

Cite this article as: Samdariya S, Jain SC, Onkar A, Viswanthan C. Radiation therapy for a rare association of maxillary neoplasm in xeroderma pigmentosum: Is it really contraindicated?. Clin Cancer Investig J 2016;5:339-41.

keratitis of the right eye. There was a diffuse, nontender, soft swelling over right maxillary sinus region. Anterior rhinoscopy showed a mass obstructing the right nasal cavity. A contrast-enhanced computed tomography scan showed moderately-enhancing mass lesion measuring approximately 5 cm × 4.5 cm × 3 cm in right maxillary sinus extending into nasal cavity, right ethmoid air cells, infratemporal fossa, pterygoid plates, right orbit, middle cranial fossa (medial to temporal lobe) [Figure 1]. There were no significantly enlarged lymph nodes on examination and imaging. Fine needle aspiration cytology from maxillary sinus mass revealed squamous cell carcinoma of right maxillary sinus. Hence, a final diagnosis of squamous cell carcinoma of right maxillary sinus cT4bN0M0 UICC Stage IVB was made.

The patient was planned for concomitant chemo-irradiation. He was counseled regarding the possible radiation-induced skin and mucosal reactions during the course of radiotherapy. After taking his informed written consent, he was treated by concomitant chemo-irradiation with 5 cycles of weekly cisplatin (40 mg/sqm) and radical EBRT using telecobalt gamma rays to a total dose of 66 Gy in 33 fractions using ipsilateral wedge pair and isocentric method for 6 weeks. He tolerated the treatment well with Radiation Therapy Oncology Group Grade II skin and Grade III mucosal toxicities and is being followed up. At 3 years of follow–up, he is disease free.

DISCUSSION

XP is a rare autosomal recessive disorder of DNA repair,[1] with a prevalence of 1 in a million.[2] It was first described by Hebra and Kaposi in 1874.[3] It is characterized by multiple pigmented lesions, marked photosensitivity, and xerosis.[4] The underlying cause of XP is defective DNA NER mechanism. Affected patients are at 1000 times higher risk to develop a variety of skin cancers such as basal cell carcinoma, squamous cell carcinoma, and malignant melanoma,[5] but association of maxillary carcinoma is very rare.

Sinonasal malignancies are rare entities with an incidence of approximately 1 in 500,000–1 in 1,000,000 people.[6] Maxillary neoplasms are the most common among this category of tumors. They are more commonly seen in males of middle age group. Surgical resection with negative microscopic margins is the preferred treatment option. Early infrastructure lesions may be excised and cured by surgery alone, but for most other cases, irradiation is given postoperatively even if margins are negative. Extension of cancer to the base of the skull, nasopharynx, or sphenoid sinus contraindicates surgical excision.[7] As in our patient, the tumor had spread to temporal lobe of brain, henceforth treated with concurrent chemoradiation.

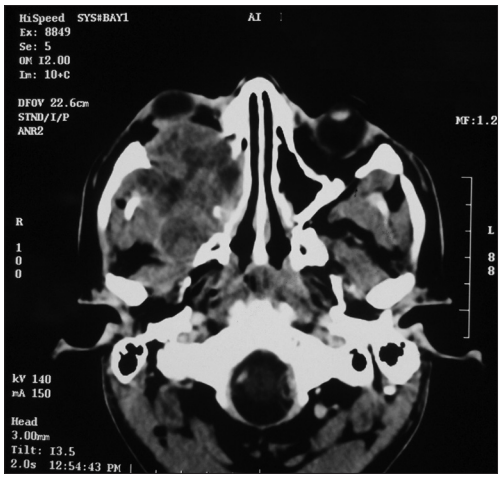

Figure 1: Axial computed tomography image showing enhancing mass lesion in right maxillary sinus extending into infratemporal fossa, pterygoid plates, right orbit

The basic principles of management of carcinoma maxilla in XP patients remain unchanged. Literature review showed that patients of XP show difference in sensitivity to ionizing radiation.[8-10] Some patients develop severe radiation-induced reactions during radiotherapy and some tolerate complete tumoricidal doses well with minimal reactions. Despite their extreme sensitivity to UV light, the patients of XP can be treated with standard doses of ionizing radiation for the treatment of cancers. The reports of use of radiotherapy as a treatment modality for maxillary neoplasms in patients of XP are rare. Our patient tolerated the treatment well with acceptable skin and mucosal toxicities.

CONCLUSION

XP patients are predisposed to develop skin malignancies, but rarely, squamous cell carcinoma of maxillary sinus may develop in these patients affecting the overall prognosis and quality of life. Management of these patients by concurrent chemo-irradiation is a good option in these types of cases. In contrast to exaggerated sensitivity to UV radiation, XP patients may tolerate therapeutic doses of ionizing radiation. Radiation therapy is an effective therapeutic modality for the treatment of maxillary neoplasm with XP. In view of short survival period of these patients, long-term clinical outcome and late effects of radiation therapy remain to be elucidated. While planning treatment by ionizing radiation, detailed counseling is needed regarding the precautions and intensive treatment of possible severe acute radiation reactions during the course of treatment in these patients.

Acknowledgment

The authors would like to thank the patient, his relatives, and every hospital staffs.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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