CASE REPORT

Dramatic response to radiation of a sinus melanoma: A case report

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Abstract

In this report, we shared our experience in the treatment of a patient with unresectable sinonasal melanoma, in whom a complete radiographic response was seen after a course of conventional external beam radiotherapy. Given that local control has an important role in maintaining the quality of life of patients.

KEYWORDS radiotherapy, sinonasal melanoma

1 **INTRODUCTION**

The origin of mucosal melanoma is from melanocyte cells in the mucous membrane in the paranasal sinuses, nasal cavity, oral cavity, lips, pharynx, vulva, vagina, and anorectal areas.¹ Primary sinus melanoma is rare and accounts for only 0.5%–2% of melanomas cases.² However, its prevalence seems to have increased according to SEER data, especially in women.³ Further use of diagnostic endoscopy or the presence of an unknown factor may be the reason.⁴ Unlike cutaneous melanoma, there is no known major risk factor for sinus melanoma, but hereditary and environmental factors may be the cause of the different prevalence between Asians and Caucasians.⁵



FIGURE 1 Pretreatment MRI (soft tissue mass in the ethmoidal sinus, nasal cavity, and maxillary sinus).

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The most common sites of sinus melanoma are the nasal cavity (70%) and the maxillary sinus (14%). The nasal cavity has been shown to have a better prognosis than maxillary sinus involvement,⁶ because sinus melanoma is asymptomatic until the local invasion and advanced disease.⁷ These patients are generally difficult to treat and the prognosis is poor. Five-year survival is less than 40% in literatures. The most commonly used treatment modality in these patients is surgery and radiotherapy.⁸ In this report, we introduce a rare case of advanced sinonasal melanoma with a dramatic response to conventional external beam radiotherapy.

2 CASE PRESENTATION

A 48-year-old healthy woman was consulted in July 2021 for nasal obstruction and epistaxis. Paranasal sinus MRI revealed intense but heterogeneously enhancing T1 and T2 mixed signal intensity soft tissue mass in the ethmoidal sinus, nasal cavity, and maxillary sinus as well as mild extension into the left frontal sinus with bone erosion and protruding into the left orbital cavity (Figure 1). Core biopsy revealed undifferentiated malignant neoplasm (Figure 2). IHC was positive for S100, HMB45, and KI67 (Figure 3) both histomorphology and IHC were compatible with malignant mucosal melanoma. Thoracic, abdominal, and pelvic CT scans showed no metastasis. The mass was unresectable due to wide extension. The multidisciplinary team decided to perform definitive radiation of the mass because of the poor response of chemotherapy in melanoma and high cost of immunotherapy and the patient was symptomatic and needed early response to treatment. Radiotherapy was performed with 3D conformal planning and a total dose of 64 Gy in 32



FIGURE 2 Microscopic view of the mass (undifferentiated malignant neoplasm).



FIGURE 3 IHC (positive for S100 (A), HMB45 (B), and KI67(C)).

fractions. At the end of the radiation therapy, signs and symptoms were improved. MRI 1 month later showed significantly decreasing tumor volume and appropriate response to therapy (Figure 4). After 3 months, the patient is symptom-free and there is no progression in 3 months follow-up MRI.



3 DISCUSSION

Primary mucosal sinonasal melanoma accounts for about 4% of head and neck cancers.⁶ Patients often present in the advanced stages because the tumor has no specific symptoms in the early stages.³ The most common symptoms at presentation are epistaxis and nasal obstruction.¹ The preferred option for assessing local tumor extension especially invasion to the base of the skull is MRI. Due to the high probability of metastases all patients should undergo PET-CT and thoracic CT-scan.³ Although the main treatment for these patients is surgery, its complete removal is challenging due to the complications of invasive surgery and complex anatomy. Recent data are in favor of using post-op radiotherapy in these patients.^{3,9} Compare to cutaneous melanoma, immunotherapy is less effective in mucosal melanoma and BRAF mutation rarely occurs.⁹ But according to KIT mutation use of imatinib was promising in some studies.¹⁰ These patients with a 5-year survival of 25% have a poor prognosis.³ Because of the low prevalence of mucosal melanoma, prospective data are not available. In a meta-analysis of the studies on mucosal melanoma patients, head and neck involvement was the most frequent site of involvement and also had the highest overall mortality and disease-specific mortality due to the difficulty of surgery. Radiotherapy and immunotherapy such as PDL1-Inhibitors and CTLA4-Inhibitors seem to be useful in un-resectable tumors.¹¹ In a retrospective study by Modreno et al., 44 patients with sinonasal melanoma were investigated. About 95.5% of the patients underwent surgery and 50% of them received post-operation radiotherapy. Distant metastases occurred in 50% of the patients, mostly in the liver, bones, and lungs and 3-year survival was less than 50%. With postoperative radiation therapy, a trend toward improved local control was observed.³

Extensive resection plays an important role in achieving proper local control because most sinus melanomas are multifocal.¹⁰ In the review 22 patients'

negative surgical margin was a major prognostic factor in 5-year survival and six of seven patients died within 24 months.¹² In a study rate of local control in melanoma of the head and neck was 0%-61% with definitive radiotherapy.¹⁰ Due to the high recurrence rate with definitive radiotherapy, Hanaoka et al. designed a study and used anti-PDL1-Inhibitors with radiotherapy. After a median follow-up of 46 weeks local control was 100% but complications like hypopigmentation, pneumonitis, anorexia, and mucositis were observed.⁹ In a recent study particle therapy by carbon-ions was used for definitive treatment of locally advanced sinonasal tumors. 2 years of local control was 82.5% in the patients with melanoma.¹³

In this report, we shared our experience in the treatment of a patient with unresectable sinonasal melanoma, in whom a complete radiographic response was seen after a course of conventional external beam radiotherapy. Because local control plays an important role in maintaining patients' quality of life, in cases where modern radiotherapy methods such as ion/proton therapy are not available, a favorable and durable response may still be available with conventional radiation therapy.

AUTHOR CONTRIBUTIONS

Drafting of the manuscript: R. H, M. G; critical revision of the manuscript for important intellectual content: R.B, and P. F; Pathologist: T.Y.

ACKNOWLEDGEMENTS

We would like to thank Dr. Nima Mousavi Darzikolaee for his generous help in this study.

CONFLICT OF INTEREST STATEMENT

The authors declared no conflict of interests.

DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

ETHICAL APPROVAL

The patient signed the informed consent. Consent form attached.

CONSENT

Published with the written consent of the patient.

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How to cite this article: Garousi M, Hariri R, Fadavi P, Yousefi T, Bayani R. Dramatic response to radiation of a sinus melanoma: A case report. *Clin Case Rep.* 2023;11:e7037. doi:<u>10.1002/ccr3.7037</u>