International Journal of Surgery Case Reports 87 (2021) 106450

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Malignant mucosal melanoma of paranasal sinuses: A case report

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ARTICLE INFO

Keywords:
Mucosal melanoma
Paranasal sinus
Surgery
Radiotherapy
Prognosis

ABSTRACT

Introduction: Mucosal melanoma of paranasal sinuses is a rare disease with a challenging treatment and a poor prognosis. In this paper, we reported the successful multimodality treatment of malignant mucosal melanoma of frontal sinus.

Case presentation: A 65-year-old female presented with a frequent nosebleed for one month before admission. Computed tomography and magnetic resonance imaging showed a mass in the right frontal and ethmoidal sinuses with adjacent bone erosion and right orbit invasion. Biopsy revealed malignant melanoma. No metastasis was found. The definitive diagnosis was malignant mucosal melanoma of paranasal sinuses AJCC stage IVb (T4bN0M0). We used right frontobasal craniotomy to resect tumor for local control of the disease. Immunohistochemical staining was Melan A(+), S100(+), and HMB45(+). A week postoperative, she received adjuvant radiotherapy and immunotherapy (pembrolizumab). For three months postoperative, the patient had no recurrence and metastasis, no headache and no new neurological deficits. She returned to her daily activities.

Clinical discussion: Mucosal melanoma of paranasal sinuses is usually aggressive and diagnosed at an advanced stage. Management options are surgery, radiation therapy, chemotherapy, and immunotherapy. These options were performed on a case-by-case basis and depend on the extent and location of the tumor. Despite that, the prognosis remains very poor, with a high rate of local recurrences and distant metastases. Therefore, post-treatment lifetime and frequent follow-ups are highly recommended.

Conclusion: The critical issues in management of mucosal melanoma are early diagnosis, multimodality treatment, and frequent follow-ups.

1. Introduction

Paranasal sinuses cancers are defined as tumors arising from maxillary, ethmoid, sphenoid, and frontal sinuses [1]. These cancers are rare, with an incidence of 1 in 500,000 to 1 in 1,000,000 people in the general population [2]. It accounted for only roughly 3% of head and neck malignant tumors [3]. They are usually aggressive and diagnosed at an advanced stage. Adenocarcinoma and squamous cell carcinoma of the maxillary sinus and ethmoid sinus make up the majority of cases (70–85%) [4,5]. In contrast, malignant mucosal melanoma of paranasal sinuses is rare and very devastating [6–8].

Diagnosis of malignant mucosal melanoma of paranasal sinus was challenging due to their complex anatomic sites and their symptoms confused with more common benign tumors. Regarding treatment, surgical treatment still plays the most critical role besides chemotherapy and radiotherapy. Despite that, the prognosis of malignant mucosal melanoma is poor, with a 5-year survival rate of a mere 0–5% [2,9,10]. In this study, we reported the successful multimodality treatment of malignant mucosal melanoma of the paranasal sinuses.

The work has been reported in line with the SCARE criteria [11].

2. Presentation of case

A 65-year-old female with no medical history was presented Hanoi Medical University Hospital (Hanoi, Vietnam) with a frequent nosebleed for one month before admission. Her nosebleed was mild but rapid

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https://doi.org/10.1016/j.ijscr.2021.106450
Received 12 August 2021; Received in revised form 21 September 2021; Accepted 22 September 2021
Available online 25 September 2021

recurrent. She also reported a dull persistent pain and watering in her right eye. On examination, she was alert and oriented. Ophthalmologic examination revealed right moderate ptosis, superior rectus palsy, and visual acuity was normal. She had no motor and sensory deficits. She denied cranial nerve palsies. Computed tomography (CT) showed a hyperdense mass in the right frontal and ethmoidal sinuses with adjacent bone erosion and right orbit invasion (Fig. 1). On magnetic resonance imaging (MRI), the mass measuring 26 × 52 mm was a heterogeneous intensity and enhanced contrast vividly. The tumor invaded the right orbit, superior rectus muscle and compressed the right frontal lobe (Fig. 2). She was screened for metastasis. Thyroid, breast and abdomen ultrasound, chest X-rays were normal. Lymphadenopathy was not found. Tumor markers (AFP, CEA, CA 19-9, CA 12-5, CA 15-3, CA 72-4) were also negative. A transnasal endoscopic biopsy was performed. Histological examination demonstrated malignant mucosal melanoma. The definitive preoperative diagnosis was malignant mucosal melanoma of paranasal sinuses AJCC stage IVb (T4bN0M0).

The patient was indicated tumor resection and cranioplasty for local control of the disease. A dose of preoperative prophylaxis antibiotic (cefotaxime 1 g, intravenous injection) was given. We used bifrontal incision and right frontobasal craniotomy. Intraoperatively, the tumor eroded frontal bone, anterior cranial fossa, right superior orbital wall, frontal and ethmoidal sinuses, and invaded dura but not brain parenchyma. This tumor was hypervascular and fragile (Fig. 3). We extirpated the tumor and involved bone with prevention from tumor cell seeding by surgical sponges. The skull bone was reconstructed by titanium mesh. Immunohistochemical staining of the tumor was Melan A (+), S100 (+), HMB45 (+), LCA (−), and CK (−) (Fig. 4).

After the operation, the patient received antibiotic (cefotaxime 1 g, intravenous injection three times per day), analgesics (acetaminophen 1 g, intravenous administration three times per day), saline solution (sodium chloride 1000 ml per day), and anti-inflammations agents (alpha chymotrypsin 5000UI, intramuscular injection twice a day). Postoperatively, her nosebleed was disappeared, but ptosis and superior rectus palsy had no change. Then, she was transferred to the oncology department for adjuvant radiotherapy and immunotherapy (pembrolizumab) after a week. For three months postoperative, the patient had no recurrence and metastasis, no headache and no new neurological deficits. She returned to her daily activities and still was followed up closely.

3. Discussion

3.1. Diagnosis and staging

Primary mucosal melanoma of the head and neck regions only accounted for 1% of malignant melanoma [8]. In which mucosal melanoma of paranasal sinuses comprised a modest 15% of them. These tumors were more prevalent in men than women, and the overwhelming majority (80%) of them occurred at 45–85 years old [6]. The typical clinical presentations are nasal obstruction, epistaxis, or loss of smell [6].

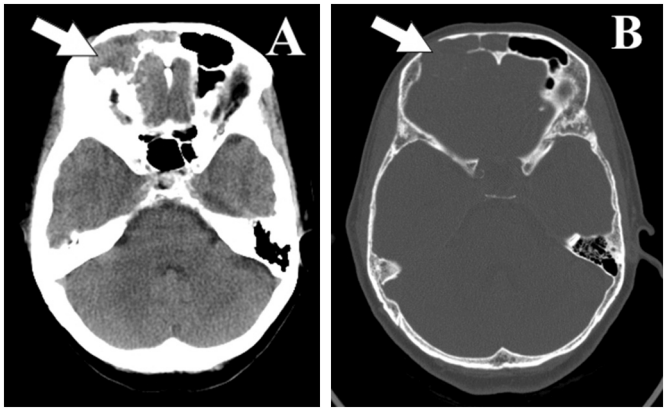

Fig. 1. CT showed a hyperdense mass (white arrow) in the right frontal and ethmoidal sinuses with adjacent bone erosion and right orbit invasion. (A) Brain window. (B) Bone window.

Diagnostic evaluation of mucosal melanoma of paranasal sinuses should include a thorough physical examination of the head and neck and flexible or rigid nasal endoscopy [1]. In addition, further work-ups include computed tomography (CT) and magnetic resonance imaging (MRI) of the head for the detailed local extent of the tumor and CT and PET imaging to evaluate lymphadenopathy and distant metastases [1]. Despite that, the biopsy is a mandatory work-up for a definitive diagnosis of mucosal melanoma. Besides standard histopathological examination, immunohistochemical stains are essential for untypical cases [1]. In melanoma, both S100 and homatropine methylbromide (HMB45) status were positive [6]. Additionally, Melan A is another highly specific marker to differentiate melanoma from other cancers, often used when stains, as mentioned above, were unclear [2].

The American Joint Committee on Cancer (AJCC) staging system for mucosal melanoma of the head and neck is now used more and more popularly [1]. It demonstrates the inferior prognosis of this disease with overall 5-year survival of a mere 0–5% [2,9,10]. Indeed, this staging system starts at stage III as the most limited form of the disease and then comprises three subcategories of stage IV disease (IVA, IVB, and IVC), depending upon the local extent and presence of regional and distant diseases [10]. In our case, the patient was diagnosed at a very advanced stage (IVb, T4bN0M0) because the tumor was involved a dura and skull base but no lymph node and distant metastases. Therefore, surgery was indicated for local control of disease, and then adjuvant radiotherapy and immunotherapy were used to control regional and distant recurrences.

3.2. Treatment and prognosis

Mucosal melanoma of paranasal sinuses has been challenging in treatment and has a poor prognosis. This is because these tumors usually developed silently with confusing symptoms and only were diagnosed at an advanced stage when cancer eroded and invaded adjacent structures. Moreover, tumors often invade vital structures, such as the skull base, orbit, brain, and carotid artery [12]. Finally, another reason for the poor survival of mucosal melanoma is the high rates of recurrences and distant metastases [13].

Management options of mucosal melanoma of paranasal sinuses are surgery, radiation therapy, chemotherapy, and immunotherapy. These options were performed on a case-by-case basis and depend on the extent and location of the tumor. Total tumor resection is the mainstay of treatment for AJCC stage III and IVA mucosal melanomas if feasible. However, surgical removal is not recommended as first-line therapy for AJCC stage IVB (lymph node involvement) and IVC (distant metastasis) mucosal melanomas except for local control of the disease [10]. In case of skull base invasion of the tumor, craniofacial resection should be performed with a multidisciplinary team, including ear, nose, and throat surgeons, neurosurgeons, and maxillofacial surgeons [6]. Radiotherapy also plays a crucial role, either as adjuvant therapy following surgical resection or when surgery is not appropriate or feasible. Nowadays, complete surgical removal of the tumor and postoperative radiation therapy is the standard of care for resectable lesions [12,13]. However, radiation therapy is contraindicated if the cancer was nearby the eye or central nervous system. Chemotherapy combined with radiotherapy was shown to enhance local control of the disease. Palliative care should be the treatment of choice in unresectable lesions with brain involvement, carotid artery encasement, and bilateral optic nerve involvement.

Regarding follow-ups, the most concerning issues of mucosal melanoma of paranasal sinuses are the high proportion of local recurrences and distant metastases despite the aggressive surgery. Meleti et al.

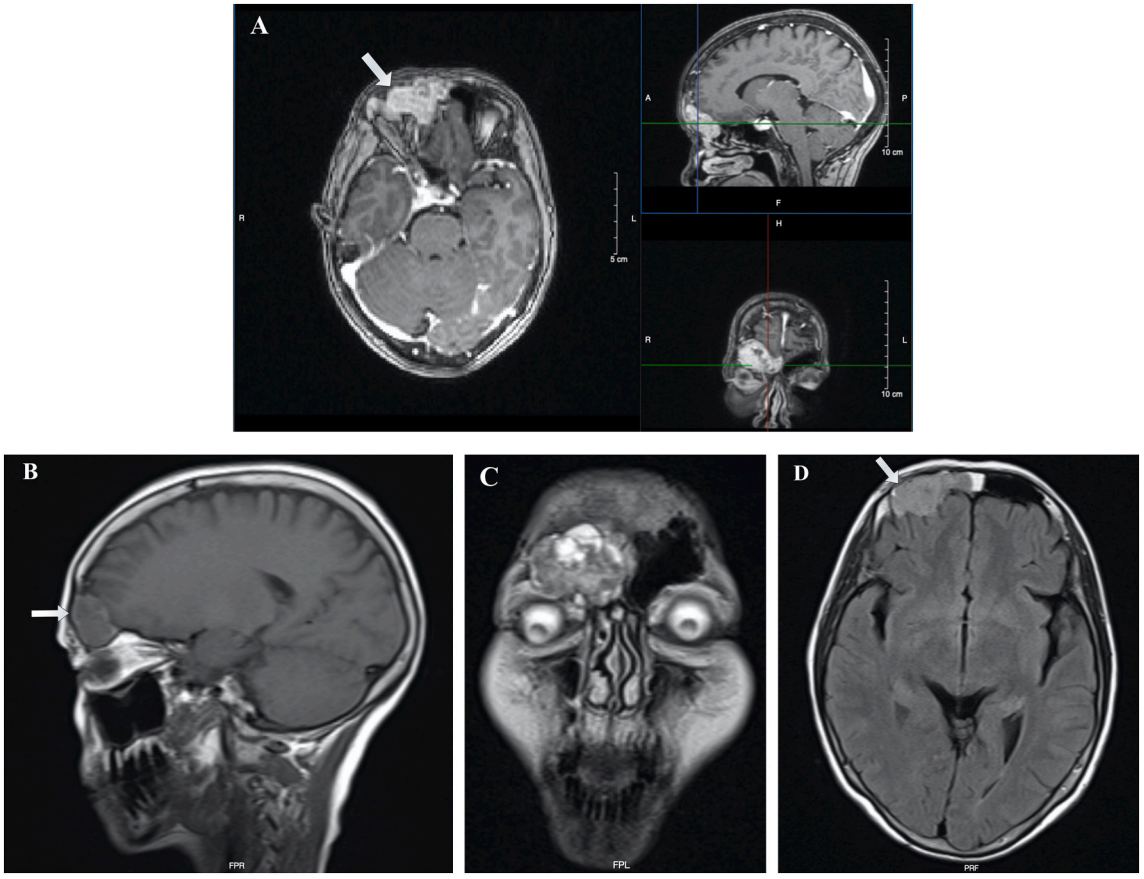

Fig. 2. On MRI, the mass (white arrow) was the heterogeneous intensity and enhanced contrast vividly. The tumor invaded the right orbit, superior rectus muscle, and compressed right frontal lobe. (A) Gadolinium-contrast T1W, (B) T1W, (C) T2W, and (D) FLAIR.

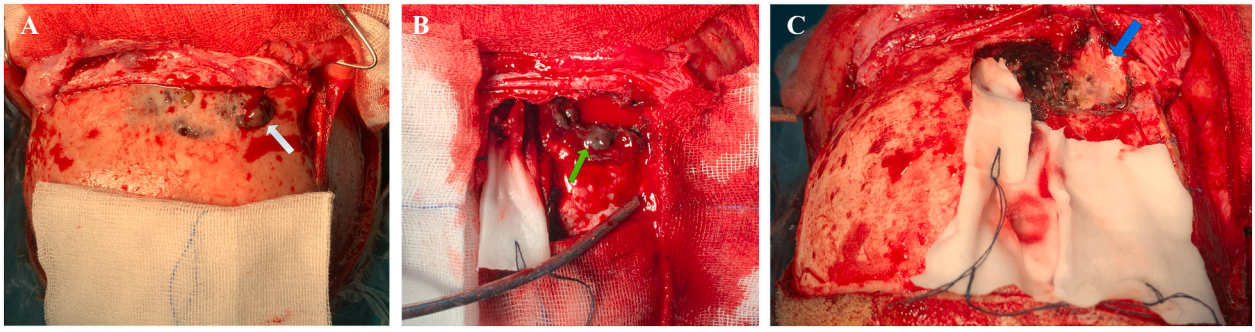

Fig. 3. The tumor eroded frontal bone (white arrow), anterior cranial fossa, right superior orbital wall, frontal and ethmoidal sinuses, and invaded dura, orbit (blue arrow) but not brain parenchyma (A, C). This tumor was hypervascular and fragile (green arrow) (B). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

reported local recurrence occurred in 57.9% and 26.3% of patients with "surgery alone" and "surgery and adjuvant radiotherapy," respectively. The rates of distant metastases were just over a half (52.6%) of patients with "surgery alone" and close to one-half (47.3%) in patients with "surgery and adjuvant radiotherapy" [14]. In a systematic review, the local, regional and distant recurrences rates were 19–61%, 7–36%, and 18–76%, respectively [8]. The overall survival rate was very poor (16–45%) [8]. In another study, Bachar et al. showed that 2-year and 5-year disease-free survivals were 25.8% and 8%, respectively [15]. Factors associated with local recurrence include tumor size, incomplete resection, and vascular invasion [2]. The median time to the first recurrence in the head and neck is 6 to 12 months. Thus, these patients with mucosal melanoma require lifetime and frequent follow-ups because of a high rate of local recurrences and distant metastases.

4. Conclusion

Malignant mucosal melanoma of paranasal sinuses is a rare and complicated disease. The prognosis of this disease remains very poor despite numerous technological developments in surgery and radiation therapy and advances in systemic modalities. Therefore, the critical issues in management are early diagnosis and multimodality treatment, which are surgical excision and radiation therapy with adequate follow-ups.

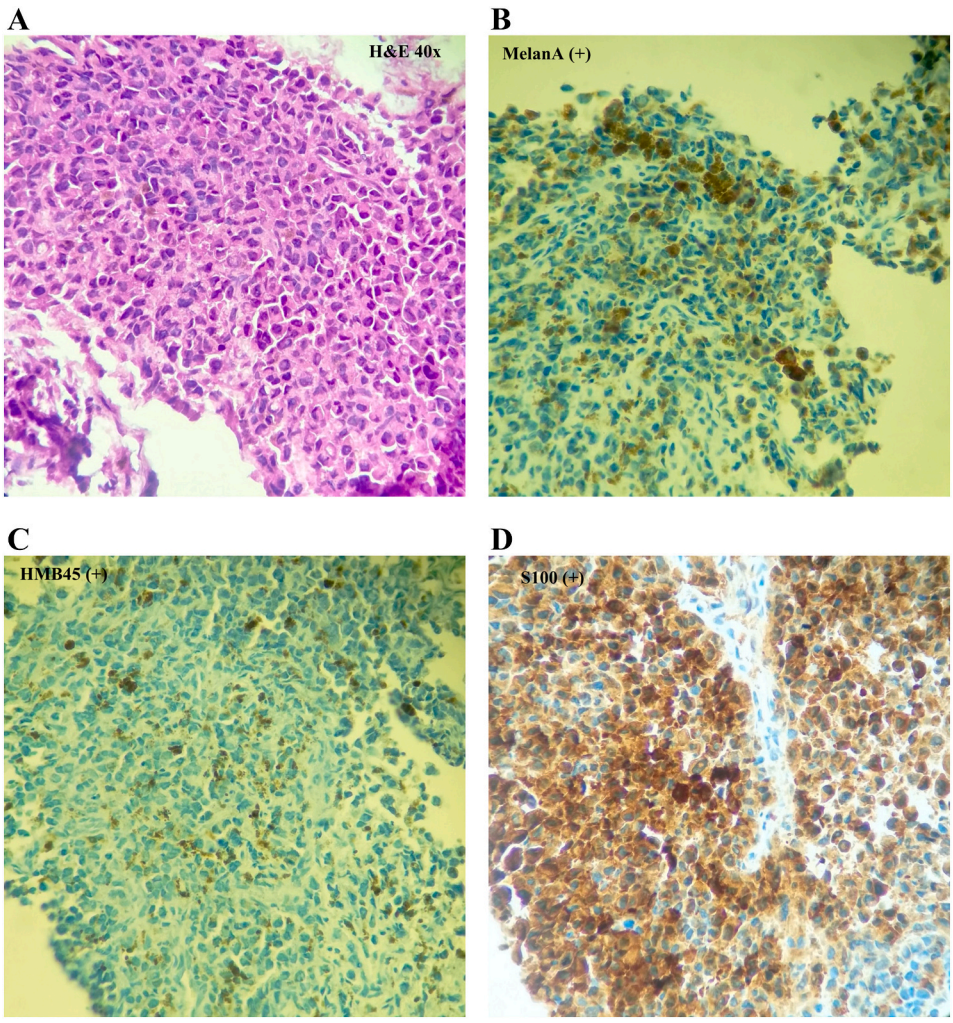

Fig. 4. Immunohistochemical staining of the tumor was Melan A (+), S100 (+), HMB45 (+), LCA (−), and CK (−).

Abbreviations

AJCC	American Joint Committee on Cancer
CT	Computed tomography
MM	Mucosal melanoma
MRI	Magnetic resonance imaging

CRediT authorship contribution statement

Hung Dinh Kieu: Conceptualization, Methodology, Investigation, Writing - Review & Editing, Supervision

Tam Duc Le: Conceptualization, Methodology, Investigation, Writing - Original Draft, Writing - Review & Editing, Visualization, Data collection

Vu Nguyen: Methodology, Resources, Writing - Review & Editing, Data collection

Trung Quang Tran: Visualization, Resources, Writing - Review & Editing, Data collection

Source of funding

This research did not receive any specific grant(s) from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

The study was approved by the Research Ethics Committee of Hanoi Medical University. The procedures used in this study adhere to the tenets of the Declarations of Helsinki.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registry

Not applicable – this is a single case report, not a systematic review or meta-analysis. Moreover, we attest that it is not a 'first in man' study, either.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Guarantor

Tam Duc Le

Declaration of competing interest

None.

Acknowledgments

Nothing to declare.

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