CASE REPORT

Malignant Mucosal Melanoma of the Nasal Cavity: No Local Recurrence with a Follow-Up of 3 Years†: Case Report

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Background: Malignant mucosal melanoma of the nasal cavity is a rare and aggressive tumor with a poor prognosis. Epistaxis and unilateral nasal obstruction are the most common presenting symptoms. Accurate diagnosis is immunohistochemical staining analysis for HMB-45 and S-100. The treatment of choice is surgical resection if the tumor is resectable. The 5-year survival rate is between 5% and 30%.

Case Report: A 72-years-old male came with left epistaxis for two months. An endoscopic examination showed a bleeding tumor at the left inferior turbinate with protrusion into the left sided nasopharynx. Computed tomography scan revealed the enhancing mass in posterior left nasal cavity involving left inferior turbinate with protrusion into left sided nasopharynx, which measured about 3.3×1.5×1.4 cm. No gross bony destruction was detected. An incisional biopsy was performed and the immunohistochemical staining analyses were positive for HMB-45 and S-100. The findings indicated stage III, T3N0M0. Endoscopic medial maxillectomy with partial nasopharyngectomy with pressure equalization tube insertion was performed under general anesthesia and the frozen sections were negative intraoperative margins. He then received postoperative radiotherapy. At a 3-year follow-up, the patient showed no evidence of local recurrence.

Conclusion: The author presented a rare case of malignant mucosal melanoma of the nasal cavity. Although the diagnosis of these tumors was challenged as the tumors were confused with other tumors when using immunohistochemistry, which lead to delay of treatment, this patient showed no evidence of local recurrence at 3-year follow-up after endoscopic resection and postoperative radiation.

Keywords: Amelanotic melanoma; Nasal cavity; Transnasal endoscopic surgery

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Melanomas are tumors arising from melanocytes and located in the basal layers of the skin or in the mucosa. The sun-exposed areas such as extremities and head and neck are the most commonly involvement[1]. Malignant melanomas have two origins, cutaneous and mucosal form. The malignant mucosal melanomas have worse prognosis due to the aggressiveness of local invasion, distant metastasis, and recurrences compared with the other cutaneous form. Primary malignant melanoma of the nasal cavity and paranasal sinus is a rare disease and less than 1% among all melanomas[2]. They are more common in males older than 50 years old[3]. Epistaxis and unilateral nasal obstruction are the most common presenting symptoms[1]. The tumors typically present at an advanced stage with a 5-year survival rate between 5% and 30%. The diagnoses of these tumors are challenged due to the locations and symptoms as they are confused with other benign tumors. The treatment of choice is resection with negative margins[4].

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Case Report

A 72-year-old man presented with left recurrent epistaxis for two months. The patient had a known history of diabetes mellitus and hypertension. He had no past traumatic or surgical history. An endoscopic examination showed a smooth pinkish mass with oozing blood at the left inferior turbinate with protrusion into the left sided nasopharynx. The clinical picture of the tumor was not performed in

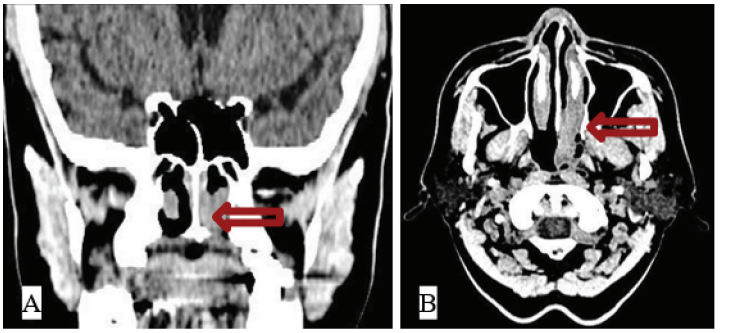

Figure 1. Computed tomography (CT) scan of paranasal sinuses; (A) Coronal view, (B) Axial view; CT scan revealed an enhancing mass in posterior left nasal cavity involving left inferior turbinate with protrusion into left sided nasopharynx, measured about 3.3×1.5×1.4 cm (arrow). No gross bony destruction is detected.

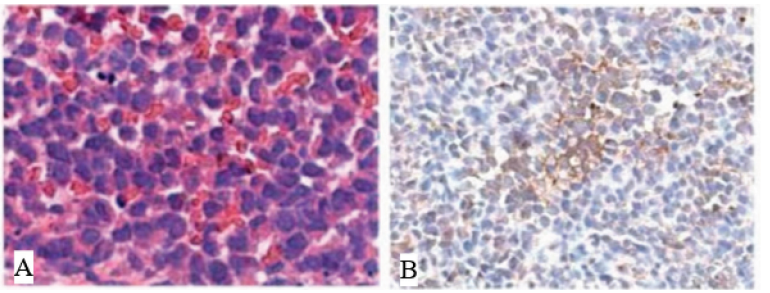

Figure 2. Pathological report of left nasal mass; (A) Plasma cell neoplasm (submucosal dense infiltration of atypical plasma cells with mature and immature nuclei); (B) Immunostains of left nasal mass: positive focal and faint CD138, no restriction Kappa and Lambda.

this patient. Computed tomography (CT) scan of the paranasal sinuses revealed an enhancing mass in posterior left nasal cavity involving left inferior turbinate with protrusion into left sided nasopharynx that measured about 3.3×1.5×1.4 cm. No gross bony destruction was detected (Figure 1).

Following biopsy under general anesthesia, the pathologists identified the lesion as plasma cell neoplasm and suggested using immunohistochemistry such as CD138, Kappa and Lambda to evaluate clonality of plasma cells (Figure 2A). Immunostains of left nasal mass were positive focal and faint CD138. There was no restriction Kappa and Lambda, which indicated that those plasmacytoid cells were not true plasma cells (Figure 2B). A round cell tumor of the sinonasal tract was suspected. The differential diagnoses included sinonasal carcinoma, melanoma, lymphoma, olfactory neuroblastoma, Ewing sarcoma, and rhabdomyosarcoma. Additional immunostains were sent for diagnosis such as AE1/AE3, S-100, CD45, Myogenin, CD99, Chromogranin, Synaptophysin, and Human Melanoma Black-45 (HMB-45). Finally, the tumor cells were positive for S-100 (Figure 3A), CD99 (Figure 3B), and HMB-45 (Figure 3C), but negative for AE1/AE3, CD45, fMyogenin, Chromogranin, and Synaptophysin. The final diagnosis was mucosal melanoma.

The positron emission tomography and computed tomography (PET-CT) scans were done for metastatic workup. They did not detect distant metastasis. The findings indicated T3N0M0, stage III, according to the American Joint Committee on Cancer (AJCC) staging system for head and neck mucosal melanoma. The patient was referred for endoscopic medial maxillectomy with partial nasopharyngectomy with pressure equalization (PE) tube insertion under general anesthesia and the frozen sections were negative intraoperative margins. He had no serious postoperative complications. The patient was discharged from the hospital, five days postoperatively. The patient was receiving postoperative radiotherapy to improve locoregional control. He underwent the intensity modulated radiation therapy (IMRT) that delivered 50 gray (Gy) in 20 fractions administered daily five days per week. There were no serious complications in this patient. The follow-up of magnetic resonance imaging (MRI) was performed every six months. At a 3-year follow-up, the patient showed no evidence of local recurrence.

Figure 3. Immunostains of left nasal mass: The tumor cells are positive for S-100 (A), CD99 (B), and HMB-45 (C), but negative for AE1/AE3, CD45, Myogenin, Chromogranin, and Synaptophysin.

Discussion

The malignant mucosal melanoma of the nasal cavity is a rare tumor and has poor prognosis[1-3].The most common area of nasal cavity is nasal septum followed by middle and inferior turbinates[5,6]. In the present case, he had a left inferior turbinate nasal mass with no aggressive local invasion and distant metastasis. However, the diagnosis of this tumor is difficult because it was confused with other tumors. Histopathologically, the presence of melanin originating from melanocytes is the hallmark feature of melanoma but the amelanotic variant becomes a diagnostic challenge for pathologists[7,8]. The immunohistochemistry was helpful in this case for definite diagnosis, but it took a long time to send each immunostains leading to a delay of treatment.

Tumor cells of melanomas are oval and round nuclei with abundant nucleoli. A similar histological structure may be recognized in other malignancies such as small round cell tumors[9]. Accurate diagnosis is immunohistochemical staining analysis for HMB-45 and S-100[10]. The first pathological report of this patient was plasma cell neoplasm, which was necessary for immunostains. However, the second report did not show the final diagnosis because there were plasmacytoid cells and not true plasma cells. Round cell tumors were suspected. Finally, the tumor cells were positive for CD99, S-100, and HMB-45, which confirmed malignant mucosal melanoma. CD99 is a stain on a variety of tumors as non-specific for Ewing/PNET but may be up to 25% of melanoma[11]. It took two months for immunohistochemical analyses.

Patients with malignant mucosal melanoma present a poor overall survival rate with a five-year survival rate of between 5% and 30%. Some patients have concomitant metastasis at the time of diagnosis[12]. Early diagnosis could help for earlier treatment and potentially improve prognosis and the overall survival rate. It would also reduce distant metastasis[8]. The location and size of the tumor are important for the surgery choice, which is resection either endoscopic or open surgery[13]. Postoperative radiation can improve locoregional control[14]. Although the endoscopic resection of this tumor becomes a treatment of choice when the surgical margin can be secured, it is still controversial[15]. However, recent research suggests that endoscopic resection has better survival outcomes than open surgery because of the simple and less invasive procedure, especially in the elderly who cannot tolerate invasive open surgery[16,17]. Suzuki et al. reported two cases of malignant mucosal melanoma who underwent endoscopic resection using temporary transseptal access. They were successful in removing the tumors from the inferior turbinate[15]. Ruggeri also reported two cases of malignant mucosal melanoma of nasal cavity with endoscopic resection. They had no evidence of local recurrence at the 5-year follow-up[18]. Karli et al. reported three cases of malignant mucosal melanoma who underwent endoscopic resection that were successful[19]. Therefore, the author prefers endoscopic resection as a treatment for resectable tumors. In the present case, the lesion was identified only in the inferior turbinate and there were no lymph nodes and distant metastases. Endoscopic surgery played an important role in the treatment. The patient underwent endoscopic medial maxillectomy with partial nasopharyngectomy with PE tube insertion and the frozen sections were negative intraoperative margins. The patient received postoperative radiotherapy and had no evidence of local recurrence at 3-year follow-up with MRI.

Conclusion

The author illustrated a rare case of malignant mucosal melanoma of the nasal cavity, an aggressive tumor with a poor prognosis that required aggressive

surgery with negative margins and postoperative radiation for patient survival. Although the diagnosis of these tumors was challenged as it was confused with other tumors leading to delayed treatment, using immunohistochemistry, the final diagnosis was made. This patient showed no evidence of local recurrence at 3-year follow-up after endoscopic resection and postoperative radiation.

What is already known on this topic?

Immunohistochemical staining analysis is helpful for the accurate diagnosis of malignant mucosal melanoma of the nasal cavity.

What does this study add?

The early diagnosis and treatment of malignant mucosal melanoma of the nasal cavity is important for patient survival.

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Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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