Malignant Mucosal Melanoma in the Nasal Cavity: An Uncommon Cause of Epistaxis

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Malignant mucosal melanoma of the nasal cavity is extremely rare. It is usually diagnosed in the elderly. We report a 78-year-old man who had symptoms of occasional episodes of epistaxis and blood-tinged sputum for about 1 month. Physical examination showed a dark-colored mass lesion over the left nasal cavity, and biopsy of it revealed malignant melanoma. Wide excision of the tumor was performed via endoscopic surgery, and adjuvant radiotherapy was also arranged. Diagnosis of malignant melanoma mainly depends on histochemistry and immunostain. Up to now, surgery offers the best chance for local tumor control. However, postoperative radiotherapy or chemotherapy is often needed because mucosal melanomas tend to have distant metastasis and local failure. Immunotherapy may play a role in improving outcome, but evidence is lacking. [J Chin Med Assoc 2010;73(9):496–498]

Key Words: epistaxis, malignant mucosal melanoma, nasal cavity neoplasms

Introduction

Melanomas are tumors arising from melanocytes, which are neuroectodermal-derived cells located in the basal layers of the skin or some mucosal membranes. They usually occur in sunlight-exposed cutaneous areas like the face, neck, and extremities. Melanomas arising from the nasal cavity are extremely rare. Sinonasal melanomas account for less than 1% of all melanomas. The symptoms of malignant mucosal melanoma are nonspecific and they appear according to tumor location; thus, diagnosis is often delayed. The most common clinical presentations of nasal cavity melanomas are epistaxis and nasal obstruction. We report the case of a 78-year-old man who had symptoms of epistaxis. Tumor mass was noted over the left nasal cavity, and biopsy revealed malignant mucosal melanoma.

Case Report

A 78-year-old man suffered from occasional episodes of left side epistaxis and blood-tinged sputum for about 1 month. He visited a local hospital, and a tumor mass was found in his left nasal cavity. Biopsy revealed malignant melanoma and he was then referred to our institute. A dark-colored tumor mass over the left-side nasal floor with easy touch bleeding was found on nasal endoscopy (Figure 1). Head and neck magnetic resonance imaging disclosed a localized tumor mass over the nasal cavity without definite bony destruction or neck metastasis. There was no paranasal sinus, or nasopharyngeal, orbital and intracranial extension (Figure 2). Tumor survey series showed no regional or distant metastasis or any suspicious cutaneous lesions. Therefore, we arranged wide excision of the tumor via endoscopic surgery. During the operation, several small pigmations were noted over the nasopharynx, bilateral torus tubarius, and bilateral nasal floors. Resection of these pigmations was also performed. Tumor pathology revealed malignant mucosal melanoma with epithelioid growth pattern. No vascular or lymphatic invasion was found microscopically. Surgical margins could not be clearly defined because they were only fragmented specimens through endoscopic approach. The pigmented areas mentioned above showed only melanin pigment depositions without malignant tumor cells (Figure 3). The final diagnosis
was nasal cavity malignant mucosal melanoma, and the clinical stage was T3N0M0, stage III, according to the American Joint Committee on Cancer 2010 cancer staging system. The patient received adjuvant radiation about 2 weeks later.

Discussion

Malignant melanoma is a common malignant skin tumor. However, primary malignant mucosal melanoma is a rare entity. Mucosal melanomas are neoplasms with a more aggressive behavior than cutaneous lesions. The reported incidence of mucosal melanomas is only about 0.5–3.0% of all malignant melanomas of all sites in the upper aerodigestive tract. Sinonasal mucosal

Figure 1. Nasal endoscopy shows a dark-colored tumor mass (white arrow) over the left nasal floor. Black arrow = nasal septum.

Figure 2. Magnetic resonance imaging: (A) coronal view; (B) axial view. A hypointense tumor mass (arrows) on the left nasal floor seen on T2-weighted imaging; no paranasal sinus or nasopharyngeal, orbital and intracranial extension are noted.

Figure 3. (A) Tumor cells of malignant mucosal melanoma with epithelioid growth pattern (Fontana-Masson stain, 200×). (B) Only melanin pigment depictions (arrows) without malignant tumor cells were found in the other pigmented areas (Fontana-Masson stain, 100×).
melanomas, although they are the most frequent of head and neck melanomas, account for less than 1% of all melanomas and less than 5% of all sinonasal tract neoplasms. The etiology and pathogenesis of mucosal melanoma is still poorly understood. It is believed that mucosal melanoma arises from melanocytes that migrate from neural crest to sinonasal mucosa.

Sinonasal mucosal melanomas are usually diagnosed in the elderly, with the peak incidence being in the 7th decade of life. They are found equally in both sexes, although some reports showed a slight male predominance. In the nasal cavity, the most common sites are the nasal septum (41%), middle turbinate (29%), inferior turbinate (23%), lateral nasal wall (7%), and nasal floor (1%). Most nasal melanomas appear as pigmented, polypoid, fleshy and friable masses, and thus might be initially diagnosed as benign lesions. The symptoms vary according to the tumor extent and location. Epistaxis is the most common presentation, followed by nasal obstruction, facial pressure, mass lesion, pain, and rhinorrhea. Diagnosis is usually delayed due to the nonspecific symptoms.

Diagnosis of mucosal melanomas is mainly based on histologic findings and immunostain because their microscopic features could be easily misdiagnosed as lymphoma, rhabdomyosarcoma, plasmacytoma, olfactory neuroblastoma, and poorly differentiated carcinoma. They are positive for S100 protein, vimentin, and specific melanocytic markers such as Melan-A and HMB45 antigens. In the tumor, abundance of melanin and specific melanocytic markers such as Melan-A and HMB45 antigens. In the tumor, abundance of melanin can be detected by magnetic resonance imaging, with the characteristics of hyperintensity on T1-weighted imaging and hypointensity on T2-weighted imaging. However, sometimes, the melanin in malignant mucosal melanoma is scanty or absent.

Mucosal melanomas usually have early vasolymphatic invasion, multiple and satellite formation, which result in easy local recurrence. If there is local failure, an increased risk of distant metastasis and poor prognosis can be expected. Other poor prognostic factors include advanced age, obstructive symptoms, tumor size > 3 cm, vascular invasion into skeletal muscle and bone, high mitotic count, marked cellular pleomorphism and distant metastasis.

Up to now, wide excision is the treatment of choice when the tumor is localized. However, adjuvant therapy like radiotherapy or chemotherapy is often needed. Some authors have suggested that radical surgery should be performed at the presentation of localized melanomas. Achievement of local tumor control provides the possibility of an increase in survival rate, although the average 5-year survival rate is only about 15–20%, and is considered to be unsatisfactory. Thus, many clinicians have tried to find effective adjuvant treatments to improve the long-term outcome. Radiotherapy is the most common adjuvant therapy postoperatively, and early intervention was reported to lead to better local control and longer disease-free survival. Chemotherapy drugs such as dacarbazine, cisplatin, ranimustine and tamoxifen are usually reserved for systemic disease or palliation. However, the actual role of postoperative radiotherapy or chemotherapy in head and neck mucosal melanomas has not yet been clearly defined, and the results have varied among different studies. Further studies are therefore needed. Immunotherapy, like interferon and Bacillus Calmette-Guérin vaccine, has been used in some cases and may play a role in the future.

In our patient, we performed wide tumor excision via nasal endoscopy due to the localized tumor extent. Many melanin depositions were found incidentally during the operation, although no tumor cells in these lesions were noted in the final pathology report. As mucosal melanomas have a multicentric characteristic, possible malignant changes in the pigmented areas could not be completely excluded. Thus, postoperative adjuvant radiotherapy is needed and long-term follow-up must be done.

References