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Primary Nasal Cavity Malignant Melanoma Presenting with Epistaxis

Epistaksis ile Başvuran Primer Nazal Kavite Malign Melanomu

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SUMMARY

Primary mucosal malignant melanoma in the nasal cavity and paranasal sinus is rare, and it has predominance in elderly patients with poor prognosis. It may lead nasal obstruction and/or epistaxis. A 46-year-old man was admitted to the hospital with three months of epistaxis. A black mass filling left nasal cavity was found on endoscopic examination. Histopathology with immunohistochemical staining revealed malignant melanoma. He had surgical resection since he had no distant metastasis. In here, mucosal malignant melanoma is discussed with review of the literature.

Key Words: *Mucosal malignant melanom, malignancy in paranasal sinuses, nasal cavity malignant melanoma, epistaxis.*

ÖZET

Nazal kavitede ve paranazal sinüste mukozal malign melanom nadirdir, yaşlılarda daha sıktır ve kötü seyirlidir. Nazal obstrüksiyona ve/veya epistaksise neden olabilir. Kırk altı yaşında erkek hasta üç aylık epistaksis ile hastaneye başvurdu. Endoskopik incelemede sol nazal kaviteyi dolduran siyah kitle saptandı. İmmünhistokimya boyama ile histopatolojide malign melanom saptandı. Uzak metastazı olmadığı için cerrahi rezeksiyon uygulandı. Burada, literatür gözden geçirilerek mukozal malign melanom tartışılmıştır.

Anahtar Kelimeler: *Mukozal malign melanoma, paranazal sinüs maligniteleri, nazal kavite malign melanoma, epistaksis.*

INTRODUCTION

Malignant melanoma (MM) is a neoplasia caused by malignant transformation of the normal melanocytes. Skin is the most common site for migration of arising precursor melanocytes in the neural crest during first trimester of fetal life. Mucosal MM is a rare form which constitutes of 0.2-8% of all MMs (1). Head and neck are common sites for mucosal MM. Sinonasal mucosal MM is extremely rare and aggressive. It represents less than 1% of all MMs, and 2-8% all malignancies in the sinonasal tract (1). Nasal cavity is the most common site and generally occurs in patients 50 to 70 years of age without gender predominance (2-4). En-bloc resection with or without postoperative radiotherapy is generally preferred in most patients (3-6).

CASE REPORT

A 46-year-old male patient with epistaxis for three months was admitted. On physical examination, his performance status was 1 according to the Eastern Cooperative Oncology Group. He had a black mass filling left nasal cavity on endoscopic evaluation of nasal cavity and a cervical lymphadenopathy (LAP) with a diameter of 0.5 cm in size. A homogenous soft tissue mass, predominantly filling the left nasal space and maxillary sinuses which destroyed the nasal septum medially on magnetic resonance imaging (MRI) of the paranasal sinuses (PNS) and neck. There was also a cervical LAP with a diameter of 0.7 cm (Figure 1). Histopathology of the diagnostic tru-cut biopsy of the mass revealed MM. Immunohistochemical examination confirmed diagnosis of MM (Figure 2). Histologic appearance of the neoplastic cells comprised subepithelial solid groups, including atypical neoplastic cells with plasmocytoid eccentric nucleus and large eosinophilic cytoplasma with clear nucleoli and melanin pigmentation on staining of hematoxylin and eosin (H & E). The atypical cells which had diffuse cytoplasmic staining with Melan A and HMB45 had also weakly cytoplasmic staining with S100 by immunohistochemical staining. He had no distant metastases. The patient underwent endoscopic total tumor resection and adjuvant high dose interferon (20 MU/m^2, five days of a week, four weeks, then 10 MU/m^2, three days of a week, 48 weeks) was planned. He had progression of the cervical LAP (2 cm) in the first month of therapy. Fine needle biopsy of the LAP revealed metastasis of MM. He refused radical neck dissection. Temazolamide failed to control the disease. Palliative cervical radiotherapy was applied. He was given palliative chemotherapy, including dacarbazine and cisplatin since he had intraabdominal LAP, liver and lung metastasis in the following days. However, he died because of progressive disease after 10 months of diagnosis.

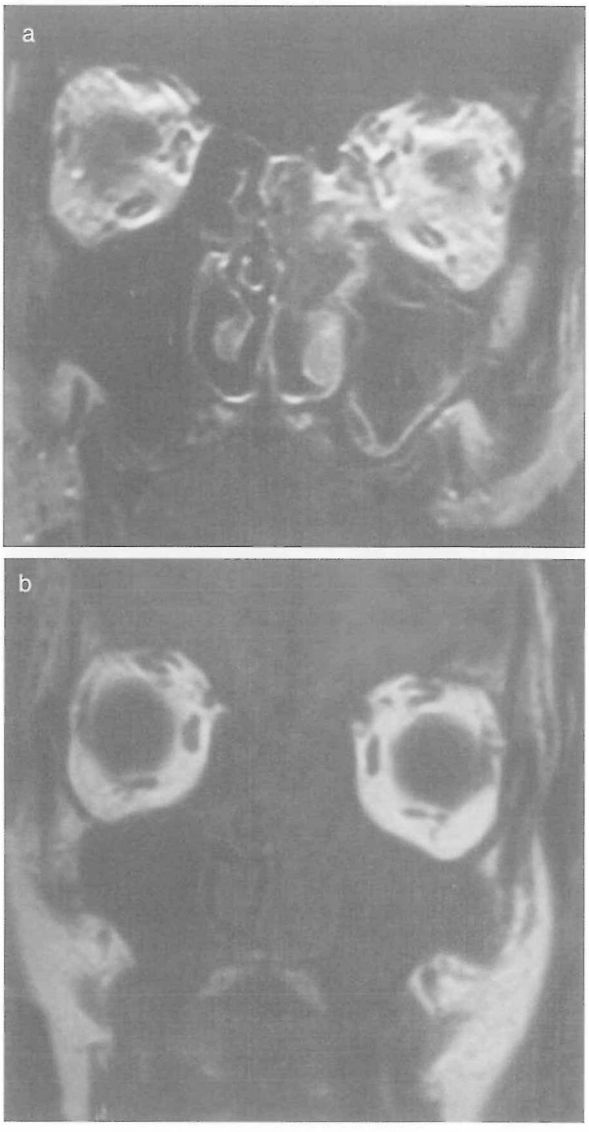

Figure 1. MRI of paranasal sinuses. Arrows show soft tissue mass, predominantly filling in the left nasal space on the coronal nects.

DISCUSSION

Sinonasal mucosal melanoma (SNMM) is a rare disease. Diagnosis of mucosal melanoma is easier when melanin-rich tumor cells are identified on histologic examination. However, one third of the cases are weakly pigmented or lack pigment, and it makes diagnosis difficult (1). S-100, HMB-45 and Melan-A are the most common immunohistochemical stains for MM

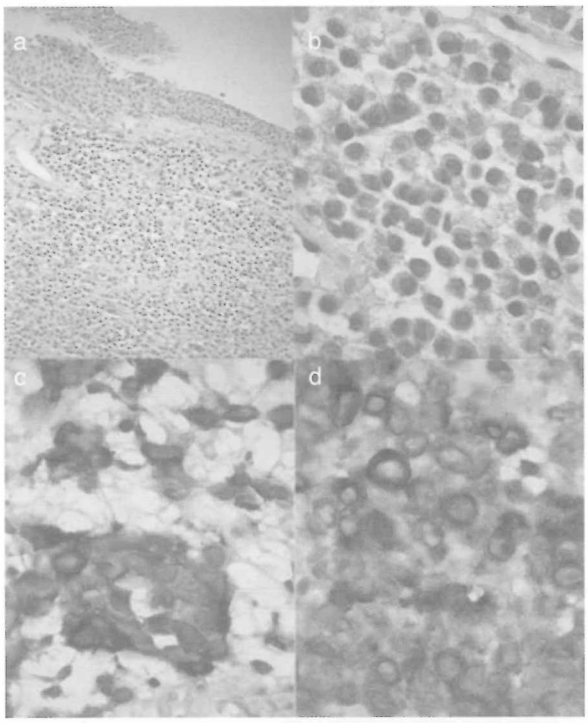

Figure 2. Microscopic view of the mass in the nasal cavity shows subepithelial solid groups including atypical neoplastic cells with plasmocytoid eccentric nucleus and large eosinophilic cytoplasma with clear nucleoli (A,B) (H & E x200, x1000). Atypical neoplastic cells which were diffuse cytoplasmic staining with Melan A and HMB45 by immunohistochemical staining (C,D), (x1000, x1000).

(1,3,4). All of them were positive in our patient. Treatment of SNMM is controversial, since data is based on case reports and small patient populations in the literature. However, surgery is preferred in resectable lesions. Endoscopic resection of the lesion failed to control the disease in our patient.

Our patient had cervical LAP which was resistant to systemic therapy. In contrast to squamous cell carcinoma, primary MM of the nasal cavity and paranasal sinuses metastatize less frequently to cervical lymph nodes whereas distant metastasis is more frequent. Regional lymph node metastasis rate was reported as 0-6% in SNMM cases (1,5,7).

It was claimed that SLNB might have had prognostic importance as a staging tool in mucosal head and neck MM (8). However, it needs further investigation before application of sentinel lymph node biopsy (SLNB) as a routine part of staging. We did not apply SLNB to our patient because of smaller size of cervical LAP on physical examination at diagnosis besides its unclear role. However, elective neck dissection is also unclear since it has low rate of regional lymph nodes metastasis at presentation (2).

Convantional radiotherapy fractionation schedules may be ineffective because of the ability of melanoma cells to repair sublethal damage. So, higher doses per fractionation may lead beter aoutcomes in SNMM (1). Adjuvant radiotherapy may improve locoregional control of the disease, especially in in higher risk groups. Radiotherapy may also have role in palliation of the symptoms. However, our patient did not benefit from palliative radiotherapy.

Adjuvant therapy is controversial. However, adjuvant systemic therapies such as interferon might contribute to better outcomes because of higher risk of hematogenous metastasis (6). We started adjuvant interferon, but our patient had progression of cervical LAP during treatment.

Five year overall survival rate was reported as 15.6% (1). Our patient died because of agressive disease progression in the first year despite systemic therapies.

In conclusion, primary mucosal MM in the nasal cavity is a rare entity with poor prognosis. The rarity of SNMM makes it difficult to determine the most appropriate treatment modality. Treatment of the patients should be individualized.

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