PRIMARY MUCOSAL MALIGNANT MELANOMA OF THE NASOLACRIMAL SYSTEM

NAZOLAKRİMAL SİSTEMİN PRİMER MUKOZAL MALİGN MELANOMU

Sümeyra DOLUOĞLU, MD;[1] Aydın ACAR, MD; [1] Serdar ÖZER, MD;[1] Gülüşan ERGÜL, MD;[2] Adil ERYILMAZ, MD[1]
[1]Department of ENT III Clinic of Ankara Numune Education and Research Hospital, Ankara, Turkey
[2]Department of Pathology Clinic of Ankara Numune Education and Research Hospital, Ankara, Turkey
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ABSTRACT

Mucosal malignant melanoma of the nasolacrimal system is rare and usually has an aggressive clinical course. These tumors present themselves with epiphora, bloody tears and epistaxis, and as the tumor grows, with a mass. Regarding these tumors, the number of studies in the literature is relatively small in number. Our case was a 92-year-old male who had presented with bloody lacrimation in the left eye for the previous three months and epistaxis for the previous two months. The histopathological diagnosis of the biopsy obtained from the mass observed at the inferior meatus during nasal endoscopy was "Malignant melanoma of the nasolacrimal system". The patient is presented under the light of the relevant literature.

Keywords: *Malignant melanoma, nasolacrimal system, epistaxis, epiphora*

ÖZ

Nazolakrimal sistemin mukozal malign melanomu nadir görülür ve son derece agresiv seyreder. Bu tümörler kendini epifora, kanlı gözyaşı, epistaksis ve tümör büyüdükçe de kitle oluşturarak belli eder. Literatür incelendiğinde bu tümörlere ait çalışmalar oldukça az sayıdadır. Bizim vakamız 3 aydır devam eden sol gözde kanlı sulanma ve 2 aydır burun kanaması şikayeti olan, 92 yaşında erkek hasta idi. Nazal endoskopide alt meatusta görülen kitleden alınan biyopsi ile nazolakrimal sistem malign melanomu tanısı konulup, literatür eşliğinde tartışılmıştır.

Anahtar Kelimeler: *Malign melanom, nazolakrimal sistem, epistaksis, epifora*

Yazışma adresi/Correspondence Address: Dr. Sümeyra DOLUOĞLU, Department of ENT III Clinic of Ankara Numune Education and Research Hospital, Ankara, Turkey
Tel: 0 3125962137 **e-mail:** sumeyradoluoglu@gmail.com

INTRODUCTION

Malignant melanomas are cutaneous tumors originating from the melanocytes of the basal layer of the epidermis. They typically arise in scarlet or fair people with blue or green eyes who bronze with difficulty after sun exposure and tend to develop freckles (1). The tumor's incidence is approximately 5 in 100,000 in USA, and develops in the head and neck in 20% of cases (2). Malignant melanoma reveals itself by its metastases rather than the primary tumor, and there is a three- fold increase in its incidence for its all clinical forms (3).

Mucosal melanoma represents 1-2 % of all melanomas. In all type melanomas, 20-25 % of mucosal melanomas occur in the head and neck (4). Sinonasal malignant melanoma is the most frequent form of mucosal melanoma occurring in the head and neck. These tumors are mostly seen at 5th – 8th decades with a slight preponderance in males. They are more frequent in black people than white population (5).
Malignant tumors of the lacrimal system are scarce. Malignant melanoma of the lacrimal sac is quite rare and may remain undiagnosed until advanced stages. Less than 30 lacrimal system malign melanoma cases have been reported in the literature. Malignant melanoma of the nasolacrimal duct has been reported only in 3 cases up to date (6, 7). In this case report, we presented a patient with malignant melanoma of the nasolacrimal system and discussed in the light of the literature.

CASE REPORT

A 92-year-old fair male was admitted to the emergency department with the complaints of bloody tearing in his left eye lasting for 3 months and recurrent epistaxis for 2 months. He was initially treated with nasal packing, prophylactic antibiotics and antihistamines. He was later hospitalized because he didn't accept treatment.

Physical examination of the patient after removal of the nasal packing revealed a concave nasal septum deviation towards the right side, and bilateral macerated and edematous nasal mucosa. There were no polyps, mass or discoloration of the mucosa. On nasal endoscopy, a hemorrhagic, dark colored and 10 x 5 mm sized mass was seen originating from the inferior meatus and extending to the nasal floor. Multiple punch biopsies were taken from the nasal floor and inferior meatus and sent for histopathologic examination.

Orbital computerized tomography (CT) of the patient revealed a nasolacrimal ductal mass causing enlargement of the nasolacrimal duct and the sac, and extending to the inferior meatus through the nasolacrimal duct (Figure 1). The mass caused an indentation in the nasal bone. Maxillary, frontal, ethmoid and sphenoid sinuses were normal on paranasal CT. On orbital and paranasal magnetic resonance imaging (MRI), a densely contrast enhanced soft tissue mass was seen extending through the left nasolacrimal duct, causing expansion of the duct and the sac (Figure 2). The patient had no lymphadenopathies on the palpation of the neck.

Figure 1: Nasolacrimal ducal mass enlarging left nasolacrimal canal, causing enlargement of canal and sac and extending into the middle meatus.

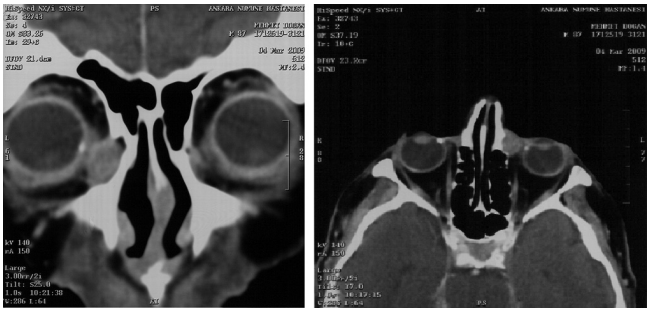
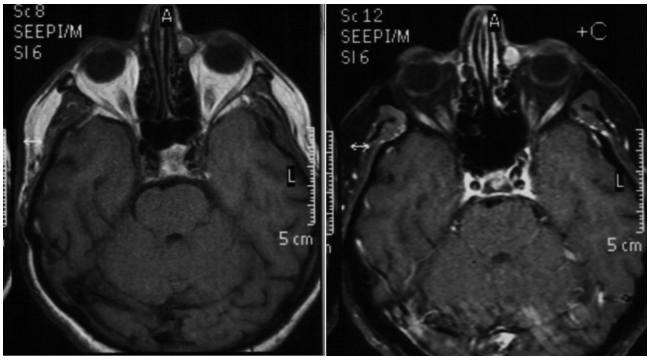


Figure 2: Densely contrast enhanced nasolacrimal soft tissue mass extending through left nasolacrimal canal and causing enlargement of the canal and the sac.



All brown colored tissue obtained by biopsy (bigger one with a diameter of 1 cm, and the smaller one with a diameter of 0.5 cm) was histopathologically sampled. On microscopy, it was noticed that all of the material consisted of tumor (Figure 3). The tumor cells were atypical, formed solid islands, had vesicular nuclei, significant eosinophilic

nucleoli and eosinophilic cytoplasms (Figure 4). Extensive intracellular and extracellular melanin pigment was noted. Immunohistochemistry showed S-100 and Melan–A staining in the tumor cells (Figure 5). With the aforementioned histological and immunohistochemical findings, the histopathological diagnosis was malignant melanoma.

Figure 3: Hemorrhagic tumoral mass forming solid islands (HE, x100).

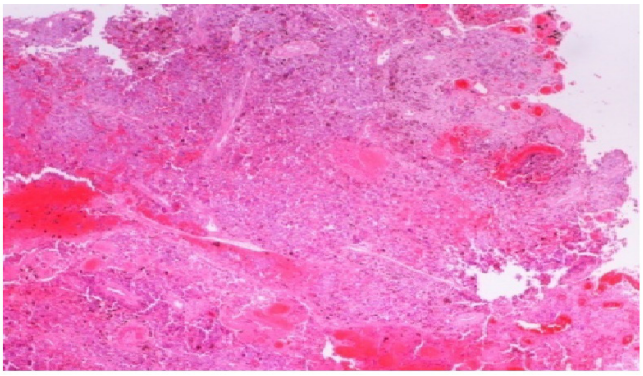


Figure 4: Tumor cells with vesicular nucleus, significant eosinophilic nucleoli and with melanin pigment in their cytoplasms (HE, x400).

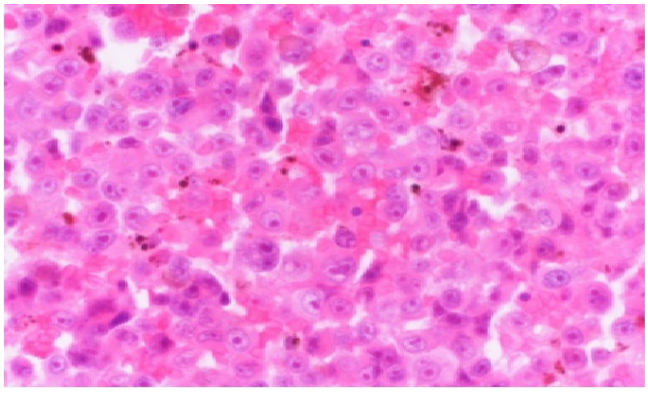
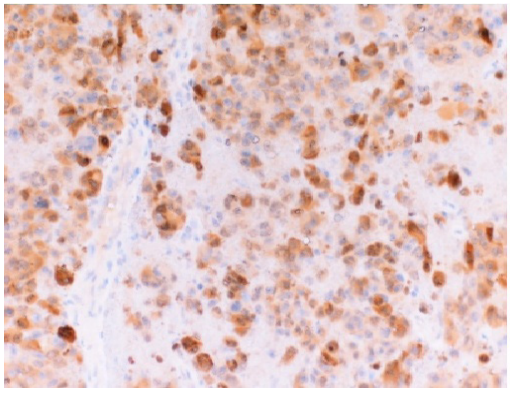
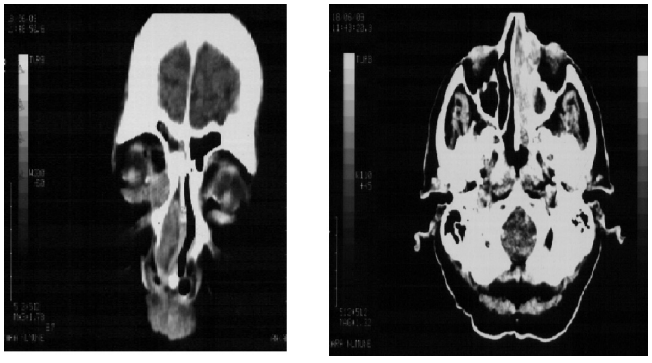


Figure 5: S- 100 positivity in tumor cells (S-100, x200).



The patient was diagnosed with left nasolacrimal malignant melanoma and was offered surgery. However, he did not agree with surgery and therefore directed to radiotherapy. But the patient did not want to receive radiotherapy as well due to the risk of blindness. Following this, the patient took palliative medical treatment (antihistamines and nasal steroids). On his follow up 2 months later, he still had epistaxis and bloody tearing. His orbital CT showed a mass with slight peripheral contrast enhancement medial to left ocular bulb and left - lateral to nasal bone, extending through the nasolacrimal duct. On the 6th month follow up of the patient, his complaints were found to have increased and a maxillofacial CT was obtained in axial and coronal planes. On maxillofacial CT, a 12 x 10 mm sized, heterogenous, hyperdense and centrally more hypodense mass was seen at the localization of left nasolacrimal canal and sac, caused thinning of the nasal bone and destruction of lamina papyracea, extending anterosuperior neighborhood of left medial rectus muscle and medial border of ocular bulb. The borders of the mass and the ocular bulb could not be separated on one section of the CT scan. There were highly hyperdense soft tissue densities filling the left ethmoid cells and the nasal cavity, and extending into the choana (Figure 6). The patient did not agree to palliative radiotherapy and died 14 months after the diagnosis.

Figure 6: A 12 x 10 mm sized, heterogenous, hyperdense and centrally more hypodense mass at the localization of left nasolacrimal canal and sac, causing thinning of the nasal bone and destruction of lamina papyracea, extending anterosuperior neighborhood of left medial rectus muscle and medial border of ocular bulb. The borders of the mass and the ocular bulb could not be separated on one section of the CT scan.



DISCUSSION

Malignant melanoma is a rare tumor with a poor prognosis. Melanocytes originate from neural crest cells and are found in all epithelia of ecdodermal origin (8). Nasal and oral mucosa arise from ecdodermal origin. Zak and Lawson are among the first investigators who claimed that melanocytes could be found dispersed throughout the nasal mucosa (9).

The etiology of melanomas originating in the respiratory tract is debated. Although some authors claimed that irritant exposure increased the risk of mucosal melanoma, Conley and Pack could not find any correlation of sinonasal melanoma with local irritation, chronic infection or allergy (10, 11).

Melanoma of the nasal cavity usually presents itself with nasal mass and epistaxis. However, patients with lacrimal system melanoma usually are admitted with bloody tears, epiphora and epistaxis. Due to obscure symptoms, patients usually present at an advanced stage. The mass can be polypoid or may involve the entire mucosa. Discoloration is common and the mass may be white, pink, gray or black (12).

Lacrimal sac tumors are rare and melanomas comprise 5% of them (13, 14). To date, fewer than 30 cases have been reported. Primary nasolacrimal duct malignant melanoma is quite rare and only 3 cases have been reported in the literature. Esteban et al. reported 2 cases (15). The first patient had a small melanoma at the inferior turbinate, after definitive surgery, the patient died 6 months later due to multiple liver metastasis. The other patient developed bilateral neck metastasis 6 months after definitive surgery and bilateral neck dissection was performed. The patient was tumor-free after one year (15). The third case in literature was reported by Lewis. He had a melanoma originating from the distal part of nasolacrimal duct, and he had definitive surgery and adjuvant radiotherapy. The patient died due to multiple bone metastasis three years later (16).

Multiple treatment modalities have been described for malignant melanoma including primary excision, external radiotherapy and chemotherapy. Radical surgical excision including the removal of all mucosa, soft tissues and involved bone is the standard therapy. However, a large tumor mass, localization of the tumor, and the general condition of the patient may make radical resection of the tumor impossible. Although mucosal melanomas have been reported to be relatively radioresistant, some authors claim that it may increase survival when applied with surgery (17). The results for chemotherapeutic regimens are variable (18- 20).

Mucosal melanomas are usually more aggressive and lethal compared to melanomas of the skin (21). The size of the tumor is more important than its thickness for the prognosis. However, there are no well defined staging criteria.

Our patient was 92 years old, his general condition was good and and he was admitted with the complaints of bloody tears and epistaxis. He was diagnosed with nasolacrimal duct malignant melanoma after physical examination, imaging and biopsy. The patient was offered comprehensive surgery including neck dissection, but he did not accept. Thereafter, he was administered palliative medical treatment (antihistamines and nasal steroids) and he followed up regularly. His complaints increased 6 months later. He had more frequent epistaxis attacks. He did not agree to palliative radiotherapy and died 14 months after the diagnosis. Although we could not treat this patient because he did not agree to treatment, this case report is important for supplying information about the natural course of malignant melanoma in this extremely rare localization.

In conclusion; patients with mucosal malignant melanomas are usually admitted at advanced stages. The disease has a poor prognosis. A patient complaining of epiphora, bloody tears, and epistaxis must have a complete physical examination including nasal endoscopy and radiological imaging. Delayed diagnosis and treatment may be life-threatening.

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