OLGU SUNUMU CASE REPORT

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Two Mucosal Malignant Melanoma Case Reports Presented During COVID-19 Pandemic

COVID-19 Pandemi Döneminde Sunulan İki Mukozal Malign Melanom Olgu Sunumu

Erdem BAYRAKCI[a], Fakih Cihat ERAVCI[a], Hacı Hasan ESEN[b], Okan Can YILMAZ[b]

[a]Department of Otorhinolaryngology, Head and Neck Surgery, Necmettin Erbakan University Meram Faculty of Medicine, Konya, Türkiye
[b]Department of Pathology, Necmettin Erbakan University Meram Faculty of Medicine, Konya, Türkiye

ABSTRACT Mucosal malignant melanoma of the head and neck are fairly rare pathologies. Even if it is not macroscopically pigmented, malignant melanoma should be included in the differential diagnosis in mucosal lesions. Mucosal malignant melanoma cases which were admitted during the coronavirus disease-2019 pandemic in 2020 were retrospectively screened. Two cases were detected. The first case was a 42-year-old male patient with tonsillar malignant melanoma presenting with a nonpigmented ulcerated mass. Second case was a 56-year-old male patient with a pigmented lesion in the left pyriform sinus. In the first case, modified radical neck dissection and lobectomy for the mass in the lung were performed at different times after whole body positron emission tomography-computed tomography scanning. Lobectomy material and two metastatic lymph nodes in the neck dissection were reported as invasive malignant melanoma. In the second case, retropharyngeal lymph node metastasis was detected accompanying the primary lesion after whole body positron emission tomography scan. Both patients were referred to oncology department for adjuvant therapy after surgical excisions.

Keywords: Melanoma, amelanotic; head and neck neoplasm; malignant melanoma

ÖZET Baş ve boyun mukozal malign melanomları oldukça nadir görülen patolojilerdir. Makroskobik pigmentli olmasa bile malign melanom, mukozal lezyonlarda ayırıcı tanıya dâhil edilmelidir. Koronavirüs hastalığı-2019 salgını sırasında 2020 yılında başvuran mukozal malign melanom olguları retrospektif olarak tarandı. İki olgu tespit edildi. İlk olgu, pigmente olmayan ülsere kitle ile başvuran tonsiller malign melanom olan 42 yaşında bir erkek hastaydı. İkinci olgu, sol piriform sinüste pigmente lezyon bulunan 56 yaşında erkek hastaydı. İlk olguda tüm vücut pozitron emisyon tomografi-bilgisayarlı tomografi taraması sonrası farklı zamanlarda modifiye radikal boyun diseksiyonu ve akciğerdeki kitle için lobektomi yapıldı. Lobektomi materyali ve boyun diseksiyonundaki 2 metastatik lenf nodu invaziv malign melanom olarak rapor edildi. İkinci olguda pozitron emisyon tomografi-bilgisayarlı tomografi taraması sonrası primer lezyona eşlik eden retrofaringeal lenf nodu metastazı saptandı. Her iki hasta da cerrahi eksizyonlardan sonra adjuvan tedavi için onkolojiye yönlendirildi.

Anahtar Kelimeler: Melanom, amelanotik; baş ve boyun neoplazileri; malign melanom

Malignant melanoma (MM) is a neoplasm that develops from pigment-producing melanocytes. Although MM is a malignancy originating mostly from the skin, there are types of extracutaneous melanoma originating from the digestive system, nasal cavity and paranasal sinus mucosa, ocular mucosa, and leptomeninges.[1] Although more than half of all primary mucosal malignant melanomas (PMMM) are seen in the head and neck region, these cases constitute only 1.3% of all melanomas.[2] MM is twice as common in men.[1] Since oral pigmented lesions are mostly seen in smokers, smoking is reported as a main risk factor.[3] Oropharyngeal MM is most commonly seen in the hard palate and maxillary gum areas.[4,5] PMMM seen in the head and neck region is distributed among sites such as the nose, paranasal sinuses, oral cavity, pharynx, and larynx with decreasing frequency.[2] Less than 30 cases of tonsillar PMMM were reported or

Correspondence: Erdem BAYRAKCI
Department of Otorhinolaryngology, Head and Neck Surgery, Necmettin Erbakan University Meram Faculty of Medicine, Konya, Türkiye
E-mail: erdem_bayrakci@hotmail.com

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referenced in the literature.[6] Hypopharyngeal PMMM is scarce in the literature. We aimed to present an uncommon diagnosis that can be overlooked and that can change the prognosis with early diagnosis, with the literature, during the coronavirus disease-2019 (COVID-19) pandemic to increase the awareness of the otolaryngologists

CASE REPORT 1

A 42-year-old male patient was admitted to our otolaryngology clinic with painless swelling in the right neck for 1 month. He also had a mild sore throat for a long time. There were no additional features on the patient's history. The patient was a smoker and used 25 packs/year of cigarettes. Oropharyngeal examination showed a macroscopically non-pigmented ulcerated lesion on the right tonsil. Neck examination revealed palpable, painless swelling which was about 3×2 cm in size and was located in zone 3, right cervical region. No other pathological findings were detected in the nasal cavity, nasopharynx, and larynx via endoscopic examination.

In the magnetic resonance imaging of the patient, asymmetric hypertrophy was observed in the right tonsil as seen in Figure 1. A punch biopsy from the right tonsil and fine-needle aspiration biopsy from the swelling in the right cervical region were obtained. Since the tonsil and neck biopsies of the patient were not diagnostic, bilateral tonsillectomy was performed. The pathology was reported as invasive MM on the right tonsil and tumor-free lymphoid tissue on the left tonsil, as seen in Figure 2. In order to detect distant metastasis, whole-body positron emission tomography-computed tomography (PET-CT) was performed and a 23×30mm lymph node in the right cervical zone 3 (SUV_{max} value: 13.19) and a bilobular nodular lesion in the upper lobe of the left lung with a size of 24×16 mm (SUV_{max} value: 11.34) were reported. The lesion in the lung was reported as a suspected metastasis or a second primary.

We performed a modified radical neck dissection and thoracic surgery team performed a lobectomy since a biopsy could not be obtained from the lesion in the lung. The pathology report revealed that

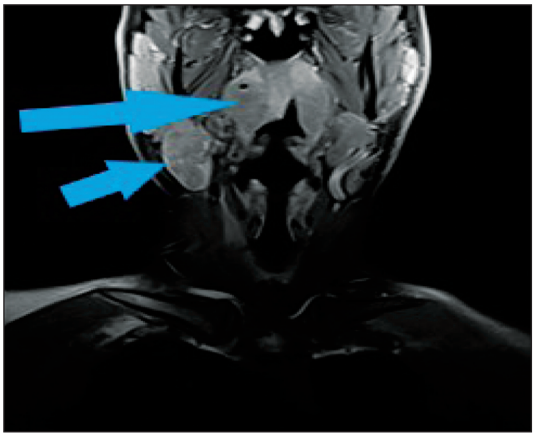

FIGURE 1: Coronal section in neck magnetic resonance imaging with contrast showing asymmetric hypertrophy on right tonsil (long arrow), 3*2 cm sized lymphadenopathy on the right servical chain (short arrow).

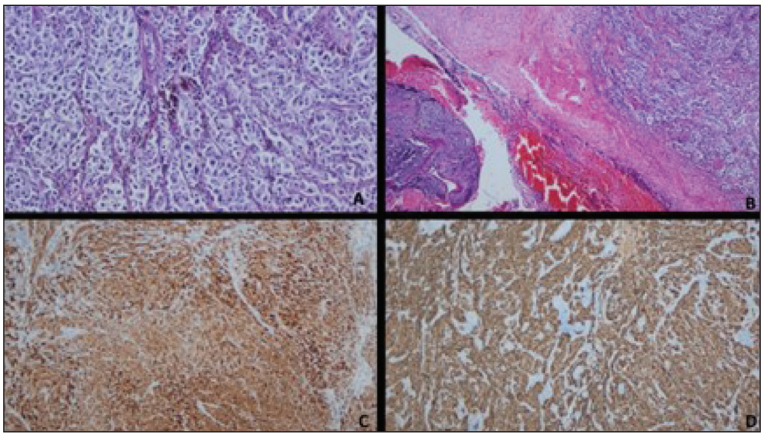

FIGURE 2: Histopathological examination: **A-B:** Tonsillar malignant melanoma cells making alveolar roofed containing mixed type (epithelioid and spindle) atypical melanocytes with prominent nucleoli and hyperchromatic nuclei in (Hematoxylin-Eosin (HE) stain, original magnification x40), **C:** x40, S-100 immunohistochemical staining is positive, **D:** x40, Melan-A immunohistochemical staining is positive.

there were 2 metastatic lymph nodes in the neck dissection material. The largest diameter of the metastatic lymph node was 4 cm, and there was no extranodal spread. Lobectomy material was also reported as an invasive MM. No complication emerged after the surgical excisions. After the recovery process, the patient was referred to the department of medical oncology for adjuvant treatments. A written informed consent was obtained from the patients.

CASE REPORT 2

A 56-year-old male patient was admitted to our clinic with a difficulty in swallowing for 2 months. In the endoscopic examination of the patient, as seen in Figure 3, a mass which was originating from the left posterior pharyngeal wall, obstructed the left pyriform sinus and showed pigmentation. Besides, blistering was observed in the posterior pharyngeal wall at the level of the soft palate, which was estimated as a retropharyngeal lymph node. The patient was a smoker with a smoking history of 40 packs/year and had been followed up for renal cell carcinoma for seven years. The pathology of the biopsy from the pyriform sinus revealed MM. The whole-body PET-CT examination was performed and, as seen in Figure 4, retropharyngeal lymph node (SUV_{max} value: 4.9) was observed in addition to the left hypopharyngeal mass (SUV_{max} value: 10.52). The mass which was adhered to the pharyngeal constrictor muscles was excised using needle cautery with the help of microscopy and endoscopy, in the laryngeal suspension position. The hypopharengeal mass which was pedunculated and originated from a limited area was totally excised with the pharyngeal constrictor muscle. Additionally, the lesion at the level of the soft palate was also excised along with the pharyngeal constrictor muscle. Histopathological examination revealed MM invasion in both the hypopharynx and retropharyngeal lymph node, as shown in Figure 5. The patient was referred to the department of medical oncology for adjuvant treatments. A written informed consent was obtained from the patients.

DISCUSSION

During the COVID-19 pandemic, head and neck cancer patients delayed their hospital admissions, and therefore early-stage patient applications decreased. However, this resulted in an increase in advanced-stage patients in the following period of time.[7] In this present study, we aimed to share two rare cases of PMMM that applied to our clinic during the pandemic to increase the awareness of the otolaryngologist. The first case was an advanced stage tonsillar PMMM with a distant metastasis. The second case was a hypopharyngeal PMMM with only a regional lymph node metastasis, which was diagnosed earlier. In the first case, it was diagnosed at an advanced stage because of the painless neck swelling for 1 month and the sore throat for a long time. In the second case, it was caught at an early stage compared to

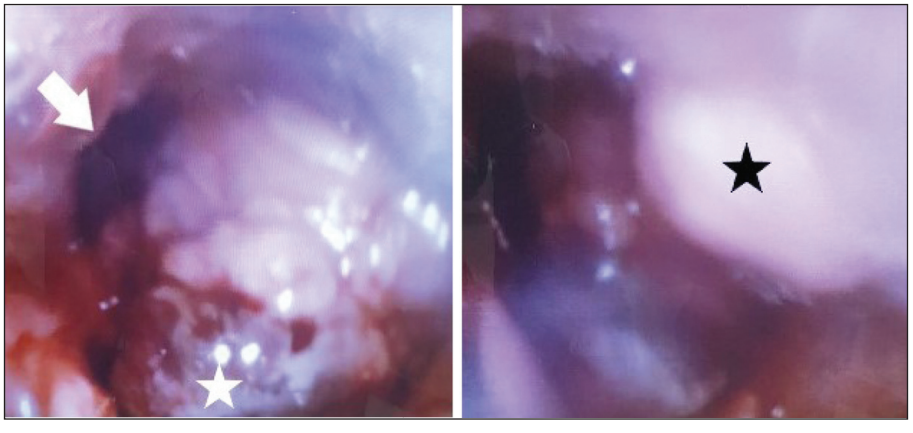

FIGURE 3: Endoscopic scene of the mass that fills the left pyriform sinus level (white arrow) and shows localized pigmentation originating from the left posterior pharyngeal wall. (White asterisk), blistering was observed in the posterior pharyngeal wall at the level of the soft palate, which was precluded as a retropharyngeal lymph node (black asterisk).

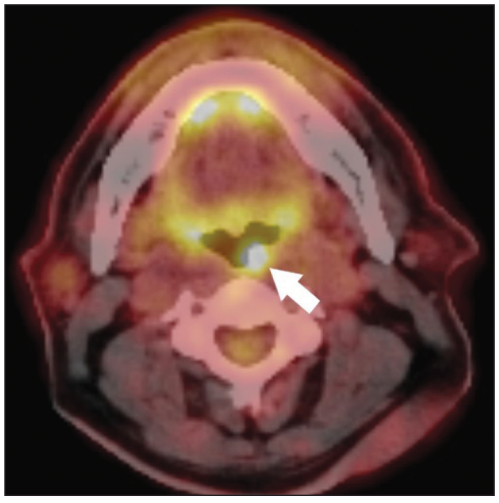

FIGURE 4: Positron emission tomography: retropharyngeal lymph node involvement in addition to the left hypopharyngeal mass (white arrow).

the first case because of the severe difficulty in swallowing. The severity of this situation was shared with the patients with the awareness of the late detection of cancer cases in the COVID-19 pandemic period. Thus their surgeries were performed as soon as possible.

MM cases in the head and neck region are less common than cutaneous melanomas and have a worse prognosis. Oropharyngeal MM is observed much less frequently. Oropharyngeal MM constitute less than 5% of malignancies, since most of the oropharyngeal malignancies are squamous cell carcinomas (SCCs).[2] Hence, there are less than 30 tonsillar PMMM patients reported in the literature.[6] Hypopharyngeal PMMM cases are less common than tonsillar PMMM cases.

Due to the rich lymphatic and vascular drainage of the mucosa, such cases present with nonspecific symptoms and mostly affect the elderly with weak immunity, leading to a late diagnosis with ensuing metastasis in MM's. Additionally, it shows aggressive histological patterns such as high mitotic index and pronounced anaplasia.[5] Approximately one-third of MM seen in the head and neck region is amelanotic and is usually diagnosed at advanced clinical stage.[8] It should be kept in mind that otolaryngologists may encounter MM as a macroscopically non-pigmented lesion, as in our first case. In this case, the differential diagnosis of SCC and lymphoma was considered due to the absence of pigmentation, and therefore, a preoperative photograph of our patient was not taken. On the other hand, the suspicion of MM in our hypopharyngeal case was brought to mind due to macroscopic pigmentation.

The incidence of distant metastasis was reported to be 4-14% for mucosal melanomas and about 5.3% for cutaneous melanomas.[9] In our tonsillar MM case, who was admitted only with swelling in the neck, was

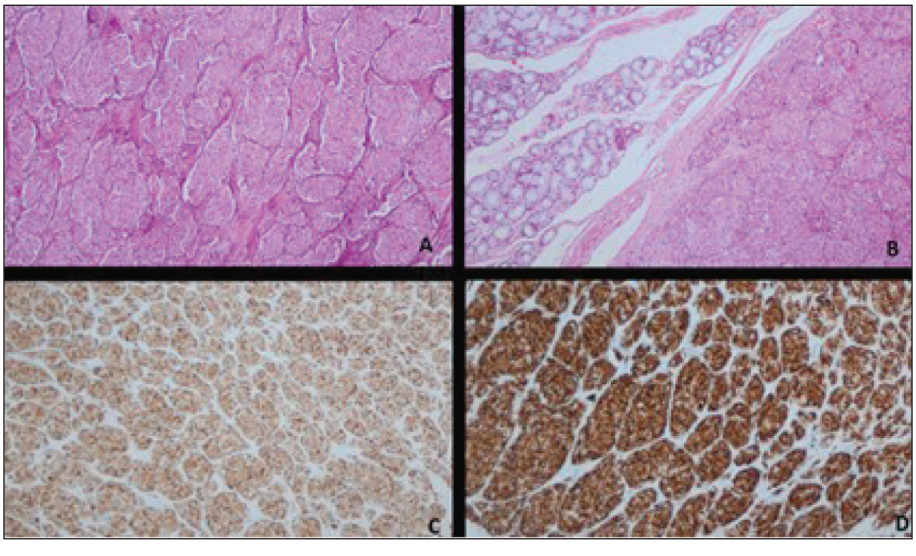

FIGURE 5: Histopathologic examination: **A-B:** Pyriform sinus cells making alveolar roofed containing atypical melanocytes with prominent nucleoli and hyperchromatic nuclei in (Hematoxylin-Eosin (HE) stain, original magnification x40, **C:** x40, S-100 immunohistochemical staining is positiv, **D:** x40, Melan-A immunohistochemical staining is positive.

found to have metastasis in both the cervical region and the lung at the time of diagnosis. In our hypopharyngeal MM case, who presented with a difficulty in swallowing was detected with only a regional metastasis in the retropharyngeal region.

Different staging systems were used for head and neck mucosal MM. Head and neck MM was included as a separate section in Tumor Node Metastasis (TNM) staging (7th version) for the first time in 2010 according to the American Joint Cancer Committee. In the recently published TNM staging (8th edition), due to the aggressive course of this disease, the staging starts directly with T3, while T4a expresses deep soft tissue invasion and T4b refers to the involvement of deep structures (brain dura, carotid artery, cranial nerve, skull base, etc.).[10] According to the guideline, elective neck dissection is not routinely recommended for all patients with T3N1 and T4aN0-1. Since our first case was T4aN1M1, wide surgical resection and elective neck dissection were performed. The second case was T3N1M0, therefore wide surgical resection and retropharyngeal lymph node excision was performed. Prophylactic neck dissection is not recommended since lymph node metastasis is less in mucosal MM cases compared with SCC.[10] However, the rates of lymph node involvement differs relating to the location of mucosal MM. Although the incidence in sinonasal MM is lower; oral cavity, laryngeal, and pharyngeal MM are more likely to metastasize to the lymph nodes. One of the previous studies suggested elective neck dissection since they have showed that lymph node involvement was approximately 77% in oral PMMMs. Additionally, sentinel lymph node biopsy and preoperative PET-CT scan was recommended to guide the elective neck dissection.[2] In our first case therapeutic neck dissection was performed due to the suspicion of metastasis according to PET-CT. Contrary to the first case, elective neck dissection was not performed in the second case, because there was no lymph node involvement in the neck according to PET-CT.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Erdem Bayrakcı, Fakih Cihat Eravcı;* ***Design:*** *Erdem Bayrakcı, Fakih Cihat Eravcı;* ***Control/Supervision:*** *Fakih Cihat Eravcı, Hacı Hasan Esen;* ***Data Collection and/or Processing:*** *Erdem Bayrakcı, Okan Can Yılmaz;* ***Analysis and/or Interpretation:*** *Erdem Bayrak, Okan Can Yılmaz, Hacı Hasan Esen,* ***Literature Review:*** *Erdem Bayrakcı, Okan Can Yılmaz;* ***Writing the Article:*** *Erdem Bayrakcı, Fakih Cihat Eravcı;* ***Critical Review:*** *Erdem Bayrakcı, Hacı Hasan Esen, Okan, Fakih Cihat Eravcı;* ***References and Fundings:*** *Erdem Bayrakcı, Okan Can Yılmaz;* ***Materials:*** *Erdem Bayrakcı, Fakih Cihat Eravcı, Hacı Hasan Esen.*

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