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Sinonasal Burkitt Lymphoma Related Palatal Ulceration

Sinonazal Burkitt Lenfoma İlişkili Palatal Ülserasyon

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ABSTRACT There are numerous reasons for the etiology of ulcerative lesions seen in the palatal area such as infection, trauma, systemic granulomatous diseases and neoplasms. The patients usually have non-specific symptoms such as headaches, nasal congestion and nasal flow similar to chronic rhinosinusitis. Although infective reasons are in the foreground in this type of lesions, sinonasal lymphomas which are rarely seen malignities in particular in the head-neck area should be dealt with in differential diagnosis. In cases in which it is considered in the beginning to be related to infective reasons, proven microbiologically and given treatment but the treatment does not work and the condition progresses negatively, there might be an underlying malignity as in this case. This may lead to delayed diagnosis and treatment for the patient.

Keywords: Palate; palate hard; paranasal sinuses; Burkitt lymphoma

ÖZET Damak bölgesinde rastlanan ülseratif lezyonların etiyolojisinde enfeksiyon, travma, sistemik granülomatöz hastalıklar ve neoplazmlar gibi birçok neden mevcuttur. Hastalar sıklıkla kronik rinosinüzit benzeri baş ağrısı, burun tıkanıklığı, burun akıntısı gibi nonspesifik semptomlara sahiptir. Enfektif nedenler bu tarz lezyonlarda ön planda olsa da özellikle baş-boyun bölgesinde nadir rastlanan maligniteler olan sinonazal lenfomalar ayırıcı tanıda ele alınmalıdır. Bu olguda da olduğu gibi başlangıçta enfektif bir nedene bağlı olduğu düşünülen ve mikrobiyolojik olarak kanıtlanıp tedavi verilen fakat tedaviden fayda görmeyen ve kötü seyirli ilerleyen olgularda altta yatan bir malignite söz konusu olabilir. Bu durum, hastanın gecikmiş tanı ve tedavi almasına sebep olabilir.

Anahtar Kelimeler: Damak; sert damak; paranazal sinüs; Burkitt lenfoma

Palatal ulceration is a rarely seen pathology and can emerge as a result of local or systemic pathologies related to congenital or acquired reasons. Among the primary acquired reasons which play a role in its etiology are; traumas which penetrate the palate, iatrogenic reasons, systemic granulomatous diseases, malignities and infections. Lymphomas which are among oral cavity malign lesions are the most frequently seen tumors following squamous cell carcinoma and salivary gland neoplasms observed in the oral cavity and their incidence has been reported as 3-5%.[1]

In oral cavity lymphomas and sinonasal lymphomas, generally extranodal involvement is in the foreground rather than primary involvement. While the hard palate, gingiva and vestibule are involved more frequently in the oral cavity, maxillary and ethmoid sinuses can be involved in the sinonasal region.[2] As a result of the effect of the mass and impairment of mucociliary function, simple nonspecific rhinosinusitis symptoms such as aches, facial edema and nasal congestion can be seen in patients. In high degree lymphomas, serious symptoms such as ulcers which cannot be treated, cranial nerves involvement and bleeding can be seen.[3] In this case study, a patient with complaints of pain in his palatal region and head area for the past 6 months who was found to have an ulcer lesion in his palate and diagnosed with Burkitt lymphoma in the sinonasal region as a result of advanced assessments is presented.

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CASE REPORT

Seventy-nine year old male patient applied to our clinic with complaints of pain in his palatal area, nasal congestion and headaches which he had been experiencing for 6 months. The patient has hypertension and Type 2 diabetes mellitus among his comorbid diseases. During the patient's oral cavity examination on the day of his visit to our clinic, a 1*1 cm ulcer lesion was observed in the intersection of his hard palate and soft palate (Figure 1). In the patient's nasal endoscopic examination, edema in nasal mucosa and purulent discharge and nasal polyp tissues around the bilateral middle concha medial were observed. His laryngeal endoscopic examination was considered to be natural. Incisional biopsy was performed under local anesthesia on the lesion in his palate with oral cavity malignity pre-diagnosis. Following the biopsy, empiric antibiotherapy and topical nasal steroid treatment were prescribed. The patient's pathology result was reported as severe chronic active inflammation due to actinomyces colonization. An infectious disease consultation was requested and the patient was referred to the infectious diseases unit. An oral amoxicilline 3 g/day was initiated by the infectious diseases unit and follow-up was started for the patient. During this process, the patient's follow-up was done with 2 week intervals. On the patient's examination 2 weeks after the biopsy, a 2*1 cm perforation was observed in the patient's biopsy area and the patient started having problems with respiration and eating (Figure 2). In the nasal endoscopy performed during this second examination of the patient, it was observed that the nasal polyps had regressed and a suspicious mass which is different from the polyp tissues in the nasal cavity was observed. Upon these findings, contrast-enhanced nasopharynx and diffusion magnetic resonance imaging (MRI) examinations were performed on the patient. In the MRI examination, a lesion of 40×25 mm dimensions which extended from the right maxillary sinus base towards the nasal cavity and in its posterior, a lesion of 2 cm of mass appearance in the naso-oropharyngeal area were found (Figure 3 a,b,c). Incisional biopsies were performed on the suspicious areas observed in the MRI on the hard palate and nasal cavity and

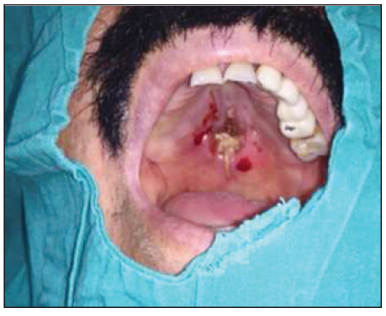

FIGURE 1: 1*1cm ulcerous lesion in the hard palate.

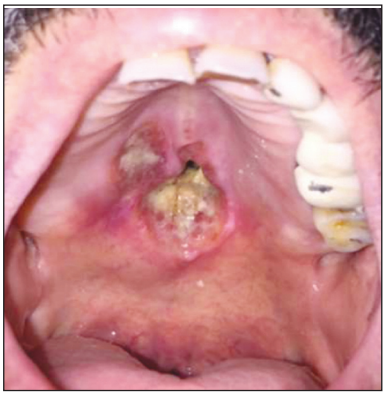

FIGURE 2: 2*1cm hard palate perforation found in the follow-up examination on the second week after the biopsy.

tracheostomy was performed simultaneously to alleviate the patient's respiration problems. The result of the incisional biopsies taken from the patient's hard palate, right upper gingiva, left upper gingiva and right nasal cavity were reported histopathologically as B-cell lymphoma. While Bcl-2 (-) was identified as a result of additional immunohistochemical analyses, the Ki-67 proliferation index was determined as >95% (Figure 4 a,b,c).

The patient was diagnosed with Burkitt lymphoma in the light of these findings. As a result of the hematology consultation, the patient was accepted as 1E stage according to Ann Arbor staging system (Lugano classification). The patient whose treatment was arranged by the hematology unit, was followed-up in monthly periods in terms of perforation and clinical findings. While the patient's headache and palate pain decreased with the help of the treatment, his oral eating problems continued due to the size of

the perforation. Therefore, the patient was fed with naso-gastric catheter and surgical percutaneous endoscopic gastrostomy (PEG) needed to be opened. In the examination after 6 cycles of chemotherapy treatment, it was observed that the size of the right maxillary sinus based mass decreased to 20×16mm. In the otolaryngological examination performed after the treatment, it was observed that the diameter of the perforation on the palate of the patient reached 4*5 cm (Figure 5).

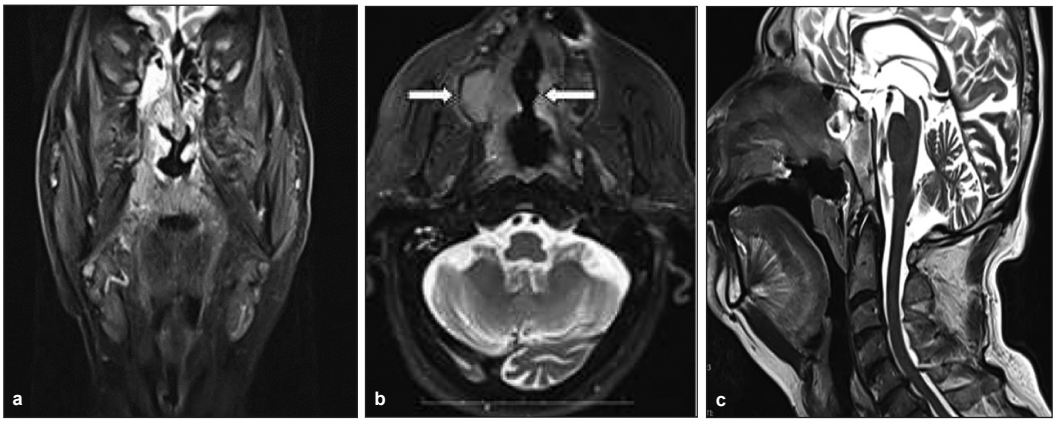

FIGURE 3: a) Computerized tomography imaging, the mass which filled the right maxillary sinus and the right nasal vestibule in the coronal section, **b)** Computerized tomography imaging, the mass which filled the right maxillary sinus and palate perforation in the axial section, **c)** Computerized tomography imaging, the mass which extends to the naso-oropharyngeal area, hard palate perforation in the sagittal section.

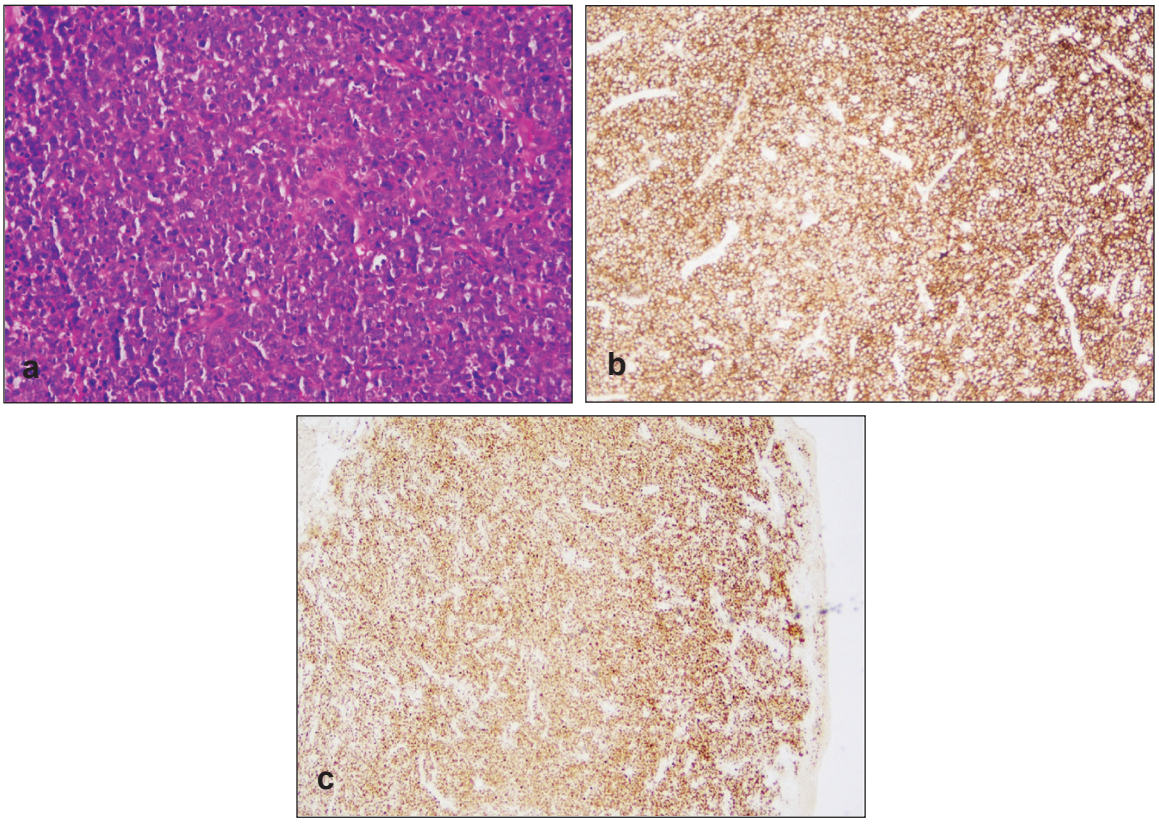

FIGURE 4: a) H&E, x400, diffusely distributed, "starry sky" pattern, hyperchromatic large nucleus, locally prominent nucleoli, cytoplasmic retractions and apoptotic bodies, very high mitotic activity, atypical lymphoid proliferation, **b)** CD20 expression in tumor cells Immunohistochemistry for CD20, x200, **c)** Ki-67 proliferation index 100% Immunohistochemistry for CD20, x200.

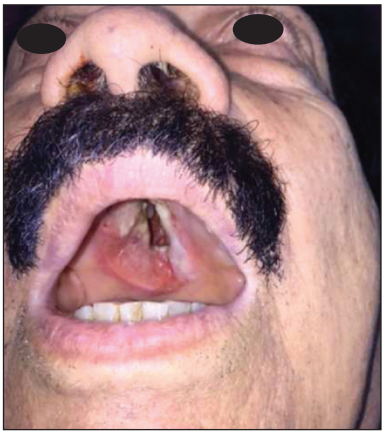

FIGURE 5: 4*5cm hard palate perforation in the follow-up after the treatment.

The patient was suggested an examination by the periodontal diseases and prosthesis unit due to his palate perforation and eating problems. The patient's check-ups were continued in the following period and the patient was lost due to the lung enfection related immundeficiency. Informed consent was obtained from the patient during this period.

DISCUSSION

There are numerous factors in the etiology of the destructive lesions seen in the palate area and the most frequently seen problems among these are some infectious periods and neoplasms. Among the most frequently seen pathogens in infectious processes are fungi such as *Zygomycosis*, *Aspergillus*, *Histoplasmosis;* viruses such as human herpesvirus 8, cytomegalovirus, Epstein-Barr virus and bacterial factors such as extrapulmonary *Mycobacterium tuberculosis*, *Treponema pallidum*, *Actinomyces*.[4]

Actinomyces family consists of opportunistic saprophytic pathogens with low virulence which are usually found in the oral cavity and the gastrointestinal system. In cases where anatomical barriers are damaged or host defense mechanisms are suppressed, they may cause actinomycosis.[1]

Invasion of *Actinomyces* in the palate is a rarely seen situation and only four cases have been reported in the literature so far. The actinomycosis diagnosis put in patients' first consultation carried less than 10% accuracy. In clinical findings in which palate perforation is observed, firstly malign processes should be eliminated.[3] In this respect, extensive examination and imaging methods for malignity analysis are quite important. In this case, a primary malignancy developing on the palate in the oral cavity was considered and an incisional biopsy was performed from the lesion in order to make a rapid diagnosis. Nasal endoscopic examination of the patient was also performed, considering that there may be a pathology originating from the nasal cavity. However, in nasal endoscopic examination, the difficulty of endoscopic examination of nasal polyps caused the actual pathology to be overlooked. Reporting of the pathology result as chronic inflammation and *Actinomyces* infection, no suspicious lesion seen in the nasal examination, and the patient's existing comorbidity that may lead to opportunistic infections, took us away from the priority of malignancy for a short time and caused a delay in the diagnosis.

It is difficult to diagnose sinonasal masses by clinical approach. The diagnosis must be clarified histopathologically with biopsies to be made from the mass. Repeated biopsies are often needed. Sinonasal lymphomas and other sinonasal malignancies can mimic benign diseases, biopsy materials from disease-free living tissue should be included in the pathological examination.[3]

Among the most frequently seen tumors of paranasal sinuses; epithelial tumors such as squamous cell carcinoma, adenoid cystic carcinoma and adenocarcinoma can be listed.[2] Primary sinonasal lymphomas are extremely rare tumors and constitute approximately 2% of extranodal non-Hodgkin lymphomas. Extranodal natural killer (NK)/T cell lymphomas, also called "nasal type lymphoma", are the most common in the sinonosal region. By advanced immunohistochemical phenotyping and molecular genetic studies, it has been revealed that most of the midfacial destructive lesions are caused by non-Hodgkin lymphoma or Wegener's granulomatosis developing from the sinonasal region. Most of the lesions that are macroscopically resembling necrotic granulomas, characterized by erosion and destruction of bone, cartilage and soft tissue in the palate, nose and paranasal sinuses, are extranodal NK/T-cell lymphoma.[5]

Burkitt lymphoma is a speedily growing and aggressive high-grade B-cell non-Hodgkin lymphoma type. Burkitt's lymphoma in the head and neck usually presents as lymphadenopathy, whereas primary involvement of the nasal cavity and paranasal sinuses is uncommon. In paranasal Burkitt's lymphoma, maxillary sinus is most commonly involved and spheno-ethmoidal sinuses are less commonly involved.[6]

No intraabdominal or cervical involvement was observed in the defined patient and the diagnosis was Burkitt lymphoma classified as Ann Arbor stage 1E which displayed extranodal involvement. The aggressive chemotherapy used in the treatment of Burkitt lymphoma is curative in the rate of 60%. In the treatment of the defined patient, R-CHOP treatment protocol (Rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone) was applied and significant remission was achieved with this treatment.

As a conclusion, destructive lesions such as palate ulcers/perforations can be caused by infections, firstly the malign processes should be eliminated. Lymphomas are rarely seen among the sinonasal area malign lesions. The clinical findings related to sinonasal lymphomas are non-specific symptoms such as pain, nasal congestion and nasal discharge and the lack of a specific finding in the beginning can cause delays in diagnosis. Therefore, in the differential diagnosis of ulcerative/destructive lesions seen in the oral cavity and sinonasal area, malignities should be considered before infectious processes. Early diagnosis and the implementation of early treatment are extremely important in terms of the patient's prognosis. The infectious process seen in the defined patient has led to delays in malignity diagnosis and treatment.

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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: *Kerimcan Çakıcı, Ozan Gökdoğan;* ***Design:*** *Kerimcan Çakıcı, Ozan Gökdoğan, Mahmut Demirtaş;* ***Control/Supervision:*** *Kerimcan Çakıcı, Harun Üçüncü;* ***Data Collection and/or Processing:*** *Kerimcan Çakıcı, Ozan Gökdoğan;* ***Analysis and/or Interpretation:*** *Ozan Gökdoğan, Mahmut Demirtaş, Harun Üçüncü;* ***Literature Review:*** *Kerimcan Çakıcı, Ozan Gökdoğan;* ***Writing the Article:*** *Barış Can Arıcı, Kerimcan Çakıcı, Ozan Gökdoğan;* ***Critical Review:*** *Kerimcan Çakıcı, Ozan Gökdoğan, Mahmut Demirtaş, Harun Üçüncü;* ***References and Fundings:*** *Kerimcan Çakıcı, Ozan Gökdoğan, Mahmut Demirtaş, Harun Üçüncü;* ***Materials:*** *Kerimcan Çakıcı, Ozan Gökdoğan, Mahmut Demirtaş, Harun Üçüncü.*

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