Case Report / Olgu Sunumu

An unusual presentation of a B-cell cutaneous lymphoma mimicking as nasolabial cyst

Nazolabiyal kisti taklit eden nadir bir B hücreli kütanöz lenfoma tablosu

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Lymphoma is a cancer of the lymphocytes which leads to solid tumors in the lymphoid organs involving lymph nodes, spleen, liver, bone marrow and skin. Primary cutaneous lymphoma, a rare subtype of non-Hodgkin's lymphoma, can be classified as cutaneous T-cell or cutaneous B-cell lymphoma. These tumors are mostly T-cell origin and mainly locate on trunk, extremities and scalp or forehead. In this article, we report a 22-year-old female case without any symptoms of non-Hodgkin's lymphoma except a sign mimicking nasolabial cyst in the nasolabial fold, who was pathologically diagnosed with cutaneous B-cell lymphoma following surgery.

Key Words: Cutaneous lymphoma; large B-cell; lymphoma; nasolabial cyst; nasolabial fold.

Lenfoma lenf nodları, dalak, karaciğer, kemik iliği ve cildi tutan ve lenfoid organlarda solid tümörler meydana getiren lenfosit hücre kaynaklı bir kanserdir. Hodgkin dışı lenfomanın nadir bir alt tipi olan primer cilt lenfoması, kütanöz T hücreli ve kütanöz B hücreli lenfoma olarak sınıflandırılmaktadır. Bu tümörler çoğunlukla T hücre kökenli olup, sıklıkla gövde, ekstremiteler ve kafa derisi veya alında yerleşim gösterirler. Bu yazıda, sol nazolabial kıvrımda nazolabiyal kisti düşündüren bir bulgu dışında Hodgkin dışı lenfomanın hiçbir semptomunu taşımayan ve ameliyat sonrası patolojik tanısı büyük B hücreli kütanöz lenfoma olarak konulan 22 yaşında bir kadın olgu sunuldu.

Anahtar Sözcükler: Kütanöz lenfoma; büyük B hücreli; lenfoma; nazolabial kist; nazolabial kıvrım.

Lymphoma is a cancer originating from lymphocytes which mainly affect lymph nodes. Abnormal proliferation of malignant cells eventually causes solid tumors in lymphoid organs such as spleen, liver, bone marrow and other sites like skin. They are basically categorized into two major groups according to cell type as Hodgkin's lymphoma which shows Reed-Sternberg cell positivity and others as non-Hodgkin's lymphoma.[1,2]

Non-Hodgkin's lymphoma can also be further classified as B-cell and T-cell neoplasms. Diffuse large B-cell lymphoma (DLBCL) is an aggressive and the most common form of non-Hodgkin's

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lymphoma with a percentage of 40%.[3,4] The treatment of this disease depends on the histology and stage of disease at presentation and includes field radiation therapy, chemotherapy and immunotherapy. Although the long-term survival rate of these patients is relatively high ranging from 26-73%, only a minority are cured.[4-6]

Primary cutaneous lymphomas which primarily show skin involvement arise mostly from T-cell type origin with no extracutaneous sign or symptom at diagnosis. It is a rare subtype of non-Hodgkin's lymphoma and they are further classified according to the new World Health Organization/European Organization for Research and Treatment of Cancer (WHO/EORTC) classification as cutaneous T-cell and cutaneous B-cell lymphoma.[7-9] Mycosis fungoides is a T-cell type lymphoma that constitutes the most common form of cutaneous lymphomas at approximately 50%.[7] In addition, primary B-cell cutaneous lymphomas are mostly localized on trunk, extremities and head.

To our knowledge this is the first case that reports a patient with cutaneous B-cell lymphoma presenting only with a cystic mass beneath the nasolabial fold skin without other signs and symptoms of non-Hodgkin's lymphoma.

CASE REPORT

A 22-year-old female patient was admitted to the Department of Otolaryngology of Uludag University with the complaint of persistent swelling at the left nasolabial fold for almost one year. She did not complain of any symptoms such as nasal obstruction, epistaxis or pain. She consulted medical doctors several times but unfortunately this swelling failed to respond to antibiotics. The previous T_2-weighted magnetic resonance images (MRI) reported no mass or calibration difference between fat and muscle planes except a midfacial asymmetry on the left (Figure 1). In addition, computed tomography (CT) scans of the paranasal sinuses revealed septal deviation to the right, hypertrophy of inferior turbinate and mucosal thickening at maxillary sinus bilaterally without bony erosion (Figure 2). She underwent also an orthopantomogram for evaluation of dental pathology, and no bony erosion or deformity of both nasal floor and periapical regions of upper left maxillary incisors and canine was noted (Figure 3).

On examination, there was a semisolid, painless and immobile mass at the left alar base and nasolabial fold with a dimension of 2x1 cm. The nasal endoscopic examination showed no abnormality, but left inferior turbinate hypertrophy. She had no palpable mass in the neck and her blood count was within normal ranges only presenting mild anemia (Hct: 37.00, Hgb: 12.00). She underwent superficial tissue ultrasound which reported a cystic mass with 2 mm wall thickness and homogenous content. This cyst had a dimension of 25x4.5 mm and was considered to be a complicated cyst or

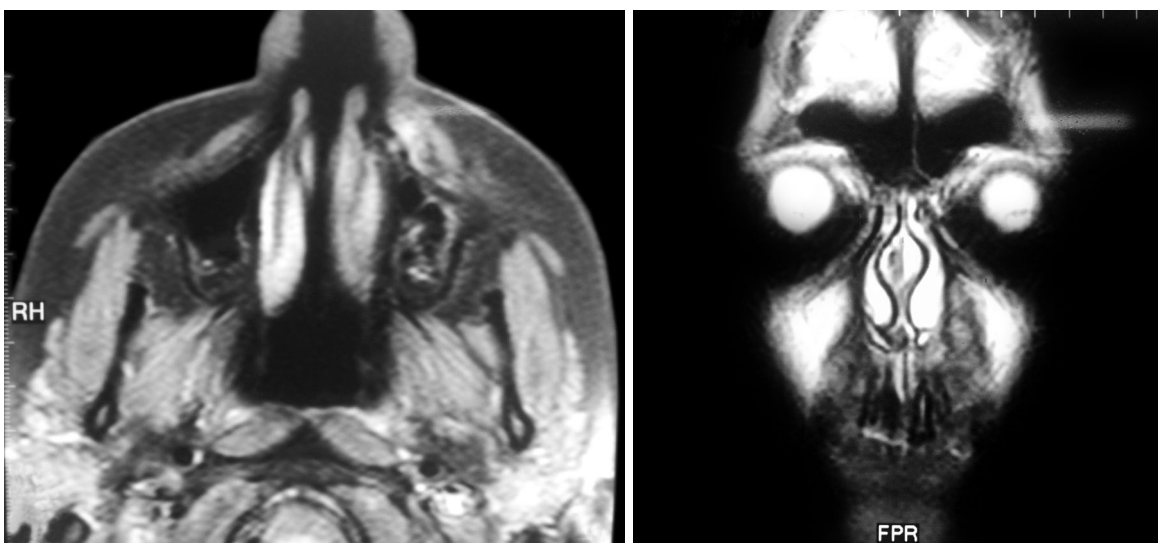

Figure 1. *The figures showing the T_2-weighted magnetic resonance imagings of a patient in axial and coronal planes in which there is a left sided subcutaneous soft tissue swelling.*

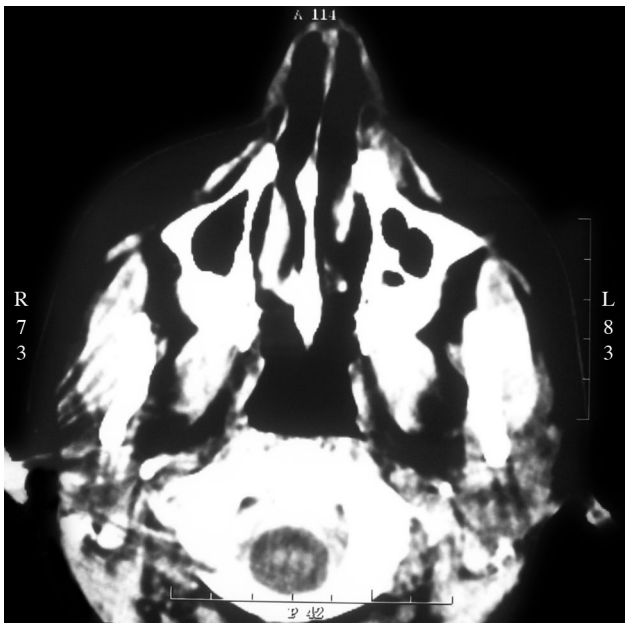

Figure 2. *Paranasal computed tomography of the patient in axial plane demonstrating hypodense soft tissue swelling on the left.*

sebaceous cyst. The patient was scheduled for surgical intervention to excise this nasolabial cyst.

The surgery was performed by a horizontal incision of 3 cm at the left gingivobuccal region. After a mucoperiosteal flap was elevated, the semisolid mass located between the canine fossa and aperture piriformis was removed. The margins of this mass were hardly defined but not invading the nasal mucosa. However the nature of this mass was not identical to an ordinary nasolabial cyst on gross examination and had

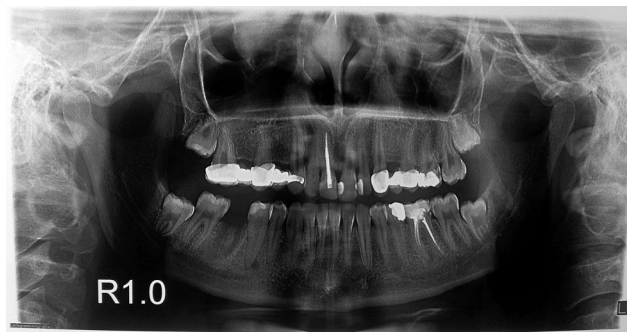

Figure 3. *Orthopantomographic image which shows no bony erosion or deformity of nasal floor and periapical regions of upper left maxillary incisors or canine.*

a dark brown color. The patient had no fever or pain following surgery and was discharged the day after.

Pathological examination of the mass reported that it was a subcutaneous fat tissue which showed high grade B-cell neoplastic lymphoid infiltration. The morphological and antigenic findings were compatible with diffuse large B-cell type lymphoma. The immunophenotype of neoplastic cells was CD20+, CD79a+, CD43+, CD23+, CD30+, Bcl2+, Vimentin+, CD4-, CD10-, CD3-, CD5-, CD8- and Epstein-Barr virus (EBV) were negative (Figure 4 and 5). Subsequently, the patient was referred to the department of hematology and oncology for further systemic assessment, staging and treatment.

DISCUSSION

The primary cutaneous lymphomas are mainly localized on trunk, extremities

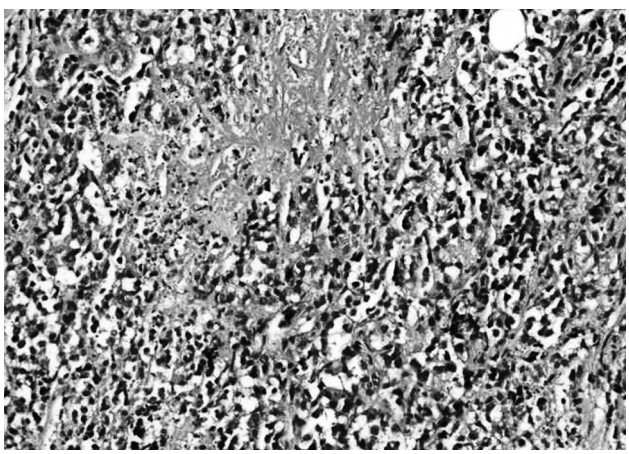

Figure 4. *Neoplastic lymphoid cells with diffuse infiltration pattern with a focal area of coagulation necrosis (H-E x 200).*

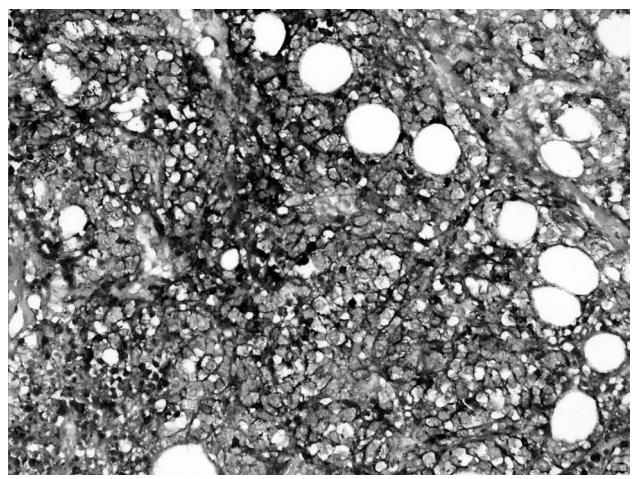

Figure 5. *Strong CD20 expression in neoplastic B lymphoid cells (CD20 x 400).*

and less commonly on scalp or forehead.[7] Histolopathologically they mostly reveal characteristics of T-cell origin.[7,10] A rare type of T-cell cutaneous lymphoma namely extranodal natural killer T (NK/T) cell lymphoma nasal type, may involve skin beside the nasal cavity, paranasal sinus or nasopharynx. They are more frequent among Asian populations such as Chinese and Japanese compared to Western countries.[11] These tumoral cells typically express CD56, CD3 and EBV positivity and are mostly originated from natural killer or T-cell lineage.[11-13] On the contrary in our case, the neoplastic lymphoid cells within the tumor were not immunophenotypically T-cell origin but identical to high grade large B-cell origin. These findings indicated a primary B-cell cutaneous lymphoma at a very rare location. To our knowledge, this is the first case which reported B-cell cutaneous lymphoma located in the nasolabial skin fold, resembling a nasolabial cyst.

Different imaging modalities such as CT and MRI are used in the assessment of extranodal lymphomas located around the nasal apparatus. Ou et al.[14] pointed out that in 75% of nasal lymphoma patients, there was bony destruction of the sinus wall, nasal turbinate, lamina papyracea, orbital wall and hard palate. In addition, they mentioned that although CT and MRI were nonspecific, one should suspect nasal lymphoma in the presence of soft tissue of nasal cavity with bony erosions and subcutaneous or nasolabial fold soft tissue infiltration. Kim et al.[15] also assessed the CT findings in patients with nasal-type NK/T-cell lymphoma. They reported that in 48% of patients, tumor spread to adjacent structures of the nasal cavity and in 17% of patients there was bone destruction. Extranodal NK/T-cell lymphoma nasal type is a different clinical entity with aggressive behavior compared to primary B-cell cutaneous lymphoma, so that the radiologists did not report any abnormality of the nasal bony apparatus in our case. But unfortunately, they also did not even mention of any mass lesion in CT and MRI. However, in the retrospective evaluation of these imagings postoperatively, an asymmetrical soft tissue swelling under the skin of the left nasolabial region was quite obvious in both MRI and CT. This finding indicated the necessity of special attention by radiologists to these soft tissue lesions in the head and neck.

In general, any space occupying lesion located between the nasal apertura and upper lip that causes unilateral swelling of the nasolabial fold in a young adult is considered either an odontogenic or nasolabial cyst. Nasolabial cysts are developmental cysts which remain silent unless infected and they can eventually cause alveolar bony erosion.[16-18] The standard treatment of these cysts is surgical excision via a sublabial incision[17] Since radiological and clinical findings of a nasolabial cyst may resemble other skin or dental lesions, the differential diagnosis of such lesions at the nasolabial fold should include histopathological confirmation.

In conclusion, although very rare in western countries, otolaryngologists should be highly suspicious and be aware of cutaneous lymphoma around the nasal apparatus especially in the presence of any mass obstructing the nasal airway, epistaxis, nasal discharge or swelling. We also should consider the possibility of malignancy if the lesion causes bony destruction and shows skin involvement. In addition, those lesions which highly mimic benign tumors or cysts in the head and neck region should not be underestimated until diagnosed histopathologically.

Declaration of conflicting interests

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