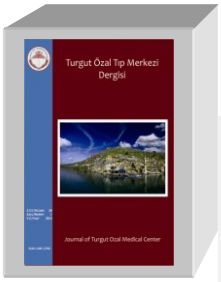

OLGU SUNUMU/CASE REPORT
J Turgut Ozal Med Cent 2013:20(3):276-278

Journal Of Turgut Ozal Medical Center
www.jtomc.org

# Extranasal NK Cell Lymphoma with Extensive Skin Involvement: A Case Report

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**Abstract**
Natural killer cell tumors are an uncommon and a heterogeneous group of disorders. These neoplasms are very aggressive, have a poor prognosis and can involve various degrees and forms of skin involvement. In contrast to the other type natural killer cell tumors, the extranasal natural killer cell lymphoma was clinically less aggressive; more localized and had a better outcome. We reported a patient with extranasal NK lymphoma, developed unexpected extensive skin involvement during the course of his disease. A-62 year old male patient admitted with swelling of the testis. Extranasal NK cell lymphoma with bone marrow involvement was diagnosed after orchiectomy and bone marrow biopsy. Extensive skin involvement developed after three cycles of CHOP chemotherapy and he died four months later from diagnosis.
**Key Words:** Ekstranazal; skin-testis; NK-cell lymphoma.

Yaygın Cilt Tululumu Olan Extranasal NK Hücreli Lenfomalı Olgu Sunumu

**Özet**
Doğal katil hücreli tümörler nadir ve heterojen bir grubu oluşturmaktadır. Bu kanserler yüksek derecede agresif ve kötü prognoza sahiptirler ve değişik derecelerde cilt tutulumu yapabilirler. Extranazal lenfomalar klinik olarak daha az agresif, daha localize ve diğer tiplere göre daha iyi sonuçlara sahiptir. Biz nadir olarak görülen yaygın cilt tutulumu ile giden bir extranazal doğal katil hücreli lenfoma olgusu sunduk. Atmış iki yaşında erkek hastaya testiste şişlik şikayetinden sonra yapılan orşektomi ve kemik iliği incelemesi sonrası kemik iliği tutulumu ile birlikte ekstranazal doğal katil hücreli lenfoma tanısı konuldu. Üç kür CHOP tedavisi sonrasında ciltte yaygın tutulum gelişti ve tanıdan dört ay sonra kaybedildi.
**Anahtar Kelimeler:** Ekstranazal, cilt-testis, NK hücreli lenfoma.

## INTRODUCTION

Natural killer (NK) cell tumors are an uncommon and heterogeneous group of disorders. NK cell neoplasms divide into immature and mature categories (1). Mature NK cell neoplasms are classified into two types: 1) Extranodal NK/T cell lymphoma, nasal type and 2) Aggressive NK cell leukemia by the World Health Organization (WHO) (1). These neoplasms are highly aggressive, and exhibit a strong association with Epstein Barr Virus (2). Extranodal NK cell lymphoma is clinically divided into two subtype; nasal, non-nasal (extranasal, nasal-type). Nasal NK cell lymphoma and extranasal NK cell lymphoma have the similar histology. The WHO classification groups evaluate both nasal NK cell lymphoma and extranasal NK cell lymphoma in the identical category but different clinical appearances, treatments, and prognoses (3).

In contrast to nasal NK cell lymphomas, extranasal NK cell lymphoma involves all of the body. Men are affected predominantly. Primary sites of involvement include the gastrointestinal tract, salivary glands, spleen, testis and skin (4). Cellulitis and ulcer are the major cutaneous manifestations. The characteristic features of the 'extranasal' compared with the 'nasal' were localized involvement of the skin, less agressive clinic course and better survival outcome (5). The diagnosis of extranasal NK cell lymphoma requires the exclusion of nasal involvement at presentation. A nasal panendoscopy with random biopsies should be performed to rule out occult involvement (6).

Evaluation of clinical presentation, morphology, immunophenotype and genotype are necessary for diagnosis of NK cell neoplasms. Expression of NK cell markers like CD56, CD16, or CD57 are positive and lack of expression of surface CD3, B-cell antigens (CD19 and CD20), myeloperoxidase, and other lineage markers are negative (7). Epstein Barr Virus status supports for diagnosis. T-cell neoplasms, myelomonocytic neoplasms, myeloid neoplasms with CD56 expression and CD41/CD56 hematodermatic neoplasm should be thought for differential diagnosis (1).

Non-nasal extranodal NK cell lymphomas have bad prognosis because the disease is clinically aggressive and chemotherapy is usually the initial choice of treatment (1). Most patients die within six months after

diagnosis. The patients applied allogeneic stem cell transplantation have long-term remission rate <10% (8).

## CASE REPORT

A-62 year old previously healthy man, an ex-smoker was admitted with a three month history of swelling right testis, fatigue, progressive weight loss. On admission physical examination revealed a well-oriented man in mild distress, a suspicious mass in the right testis.

Laboratory values were notable for leukocyte count 11.000/microL, hemoglobin 15.3 mg/dL, platelet count 123.000/microL, lactate dehydrogenase 373 U/L, peripheral blood respectively; 66% neutrophils, 26% lymphocytes, 6% monocytes, 2% eosinophils, platelets clusters. The red blood cell morphology was normal. The 3×2 cm sized mass was discovered in testis by ultrasonography. Computed tomography (CT) findings of the neck, chest and abdomen were normal. Orchiectomy was applied and non-hodgkin lymphoma was diagnosed but subgroup could not be determined due to technical inefficiency. Bone marrow examination revealed involvement of extranodal NK-cell lymphoma CD3 (-), CD19 (-), CD56 (+). Ebstein Barr Virus could not be diagnosed due to technical insufficiency. Examination of ear, nose, and throat was normal. Patient did not accept operating panendoscopik biopsy. Patient was considered Stage IVB (Ann-Arbor) extranasal NK-cell lymphoma and CHOP (cyclophosphamide 750mg/m$^2$/day, vincristine 1.3mg/m$^2$/day, adriamycin 50mg/m$^2$/day, prednisolone 100mg/day) therapy was planned because of age and performance status. Pruritic erythematous maculopapular rash which tend to coalesce with each other developed after three cycles of chemotherapy (Figure 1).

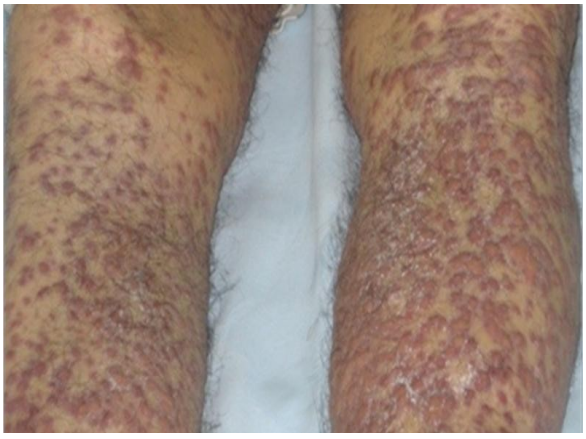

**Figure 1.** The lower extremity scattered erythematous nodules of variable size, some of which were ulcerated.

Fine needle aspiration biopsy was applied from rashes on the leg and reported as skin involvement of NK-cell lymphoma CD56 (+) (Figure 2). His clinical course worsened rapidly, pneumonia developed and he died during the fourth cycle of CHOP treatment.

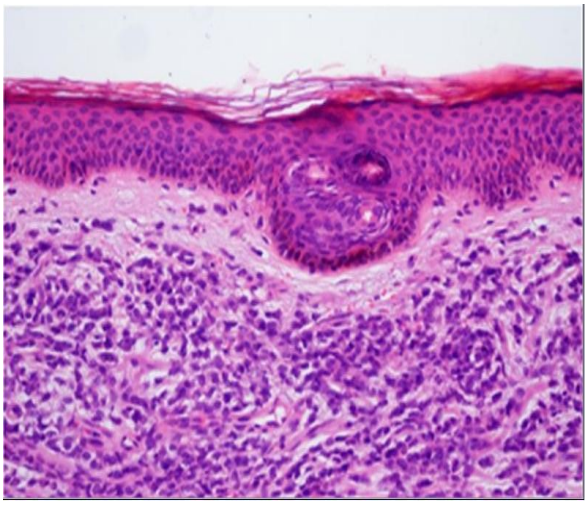

**Figure 2.** Fine needle aspiration biopsy in epidermis revealing NK cell infiltration (HE×10)

## DISCUSSION

By far the most common lymphomas' histological subtype is diffuse large B-cell lymphoma, accounting for 80-90% of primary testicular lymphoma (9). Extranasal NK cell lymphoma was diagnosed in our case's testis.

A nasal panendoscopy with random biopsies should be performed to rule out occult involvement (6). Our patient did not accept panendoscopic biopsy. We approved extranasal NK cell lymphoma because of normal ear, nose, throat examination and neck CT.

Most of the patients with non-nasal extranodal NK cell lymphoma non-nasal presented with either localized cellulitis or ulcer (5). In our patient an erythematous nodular rash covering the whole body had developed and fine needle biopsy confirmed skin involvement of non-nasal extranodal NK cell lymphoma.

Our case was very aggressive because skin rashes developed during treatment and the patient died four months after diagnosis. In extranodal non-nasal NK cell lymphoma, the long-term remission rate with allogeneic stem cell transplantation is reportedly <10% (8). We have the same opinion for patients who are eligible for allogeneic stem cell transplantation. Our patient was not eligible for allogeneic stem cell transplantation because of age and performance status.

In conclusion, extranasal NK cell lymphoma can occur in different clinical situations like extensive skin involvement, which must be kept mind.

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**Received/Başvuru: 28.02.2013, Accepted/Kabul: 08.05.2013**

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**For citing/Atıf için:**

Erkurt MA, Berber I, Karadag N, Kuku I, Kaya E. Extranasal NK cell lymphoma with extensive skin involvement: a case report. J Turgut Ozal Med Cent 2013;20(3):276-278 DOI: 10.7247/jtomc.20.3.19