Natural Killer/T-Cell Non-Hodgkin Lymphoma of the Maxilla: Case Report

Maksillanın Natural Killer/T-Hücreli Non-Hodgkin Lenfoma Olgusu

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ABSTRACT

NK/T Cell lymphoma involving nasal cavity, paranasal sinuses and nasopharynx is a kind of Non-Hodgkin lymphoma which also known as midline lethal granuloma. This rare lymphoma mostly involves midfacial region and has a destructive clinical course with poor prognosis. Clinical presentation includes non-specific sinusitis-like symptoms such as rinorrhea, nasal obstruction, nasal crusting, epistaxis and midfacial swelling. A 62-year-old female patient presented with left sided face swelling, hyperemia and nasal obstruction is discussed in this paper. Left intranasal and nasopharyngeal located mass is determined during routine ENT examination. Paranasal Computerized Tomography (PNCT) and Magnetic Resonance Imaging (MRI) of the paranasal sinuses revealed a mass located to the left maxillary sinus, nasal cavity, frontal sinus and extended into periorbital fat tissue. Endoscopic biopsy of the mass reported to be NK/T Cell lymphoma.

Keywords
Non-Hodgkin lymphoma; maxilla

ÖZET

Sinonazal yerleşimli NK/T hücreli lenfoma nazal kaviteyi, paranazal sinüsleri ve nazofarenksi tutan nadir bir hastalıktır. Midline Letal Granüloma olarak ta bilinen bu hastalık genel olarak yüzün orta hattına yerleşen, destrüksiyonla seyreden kötü prognozlu bir Non-Hodgkin Lenfomadır. Bu yazıda 62 yaşında 6 aydan beri sol yanak üzerinde şişlik, kızarıklık ve burun tıkanıklığı şikayeti olan bayan hasta sunuldu. Hastanın fizik muayenesi ve radyolojik incelemeler neticesinde sol maksiller sinüsü, sol nazal kaviteyi, sol frontal sinüsü tutan sol orbita içine uzanan NK/T hücreli lenfoma tespit edildi. Sinonazal yerleşimli NK/T-hücreli Non-Hodgkin lenfoma nadir görülen, klinik olarak kronik sinüzit, Wegener Granülamatozisi gibi inflamatuar hastalıkları taklit eden agresif seyirli bir tümördür. Burun akıntısı, burun tıkanıklığı, burunda kabuklanma, burun kanaması, yüzde şişlik gibi non spesifik semptomlar ile seyretmesi nedeni ile kronik sinüzit, Wegener Granülamatozisi, Tersiyer Sifiliz, yanak cildinde sellülit gibi hastalıklar ile ayırıcı tanısı yapılmalıdır. NK/T- hücreli lenfoma tedavisinde kemoradyoterapi protokolleri uygulanmaktadır.

Anahtar Sözcükler
Non-Hodgkin lenfoma; maksilla

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INTRODUCTION

Extranodal nasal/para-nasal natural killer T cell lymphoma (NK/T Cell) is a rare and aggressive Non-Hodgkin Lymphoma which originates from Natural Killer or Cytotoxic T cells and involves nasal cavity or paranasal sinuses.[1] It accounts for 0.4-2.2% of all Non Hodgkin Lymphoma cases at non-endemic countries and 6-7% at endemic regions like South- East Asia and South America and has a relationship with Ebstein Barr Virus infection (EBV).[2] Initial symptoms includes nasal obstruction, facial fullness, epistaxis and rinorhea which also may be seen in non-neoplastic diseases. Midfacial destruction, necrosis, facial swelling pain and hemorrhage may also be seen during the advanced disease.[1]

Midfacial necrosis and destruction may also be seen in the clinical course of tertiary syphilis, epithelial neoplasms, invasive fungal infections of paranasal sinuses and Wegener Granulamatosis.[3]

Nasal cavity and paranasal sinus located hemorrhagic mass and widespread mucosal ulceration is mostly determined at the endoscopic examination of the nasal cavity. Endoscopic biopsy gathered from the mass is the cornerstone of the diagnosis. Mucosal thickening of the paranasal sinuses on the PNCTmay be determined in the clinical evaluation of the NK/T Cell lymphoma. Diagnosis may be delayed due to non spesific symptoms and PNCT findings of the disease.[4]

Chemotherapy and radiotherapy is widely used at the treatment of NK/T Cell lymphoma.[5] Whereas local disease may be treated only with radiotherapy, chemotherapy should be added in the management of the advanced disease. Despite aggressive treatment modalities 50% local recurrence has been reported. Local invasion, involvement of lymph nodes, elevated lactate dehydrogenase levels, night sweatings, fever and loss of weight are poor prognostic evidences and in the light of these parameters 5-year survival is reported to be 38-85%.[1,6]

NK/T Cell Lymphoma of the maxilla is discussed with the literature due to its rarity, mimicking inflamatory diseases, conflicts in the differntial dignosis and aggresive clinical course.

CASE REPORT

A 62-year-old female patient presented with left sided facial swelling, hyperemia and nasal obstruction with a duration of 6 months (Figure 1). The patient had not a chronic disease history and routine laboratory evaluation was insignificant. ENT examination revealed left sided facial swelling, hyperemia and a mass originated from left nasal cavity and nasopharynx. PNCT and MR imaging exams revealed a mass involving left maxillary sinus, left nasal cavity, left frontal sinus and left periorbital fat tissue (Figures 2,3). The result of an endoscopic biopsy was reported to be nasal type NK/T Cell lymphoma. On macroscopic examination of the biopsy material two pieces of grey-white irregular tissue sized 0,5*0,4*0,3 cm observed. On serial sections in the microscopic examination, it was found that the entire specimen contained neoplastic lymphoid infiltration. On mucosal site there was an ulceration. The lymphomatous infiltrate was diffuse, but an angiocentric and angiodestructive growth pattern was frequently present. In our case lymphoma was composed of medium-sized cells, scattered small and large cells. The cells had irregular nuclei. There was an admixture of inflammatory cells, including lymphocytes, plasma cells, eosinophils (Figures 4,5). Immunohistochemically neoplastic cells were CD2+, CD56+, Granzyme B+, cd7+, but surface CD3- (Figures 6-8). Thus the case was diagnosed as 'Extranodal NK/T-cell lymphoma"

Patient was refferred to the medical oncology clinic for staging and treatment. On 4th month of treatment the patient died of chemotherapy complications.

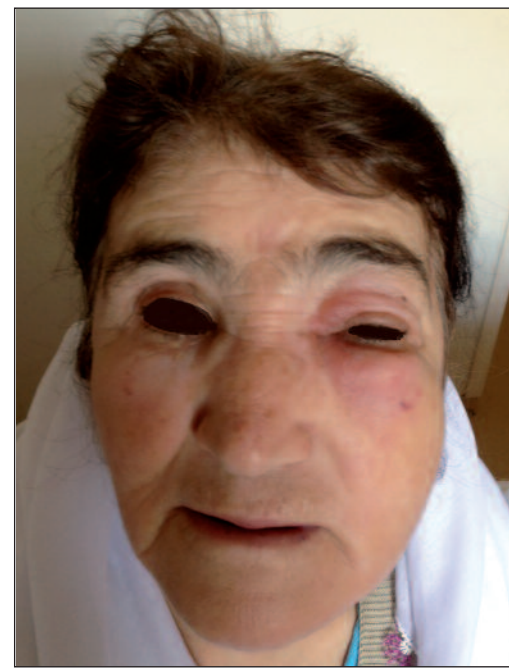


Figure 1. Congestion and swelling of the left cheek and left periorbital region.

DISCUSSION

NK/T Cell lymphoma of the nasal cavity and paranasal sinuses is a kind of rare Non-Hodgkin lymphoma originated from natural killer cells (NK) or Cytotoxic T-Cells. It consist 5-6% of all Non-Hodgkin lymphomas in South-East of Asia.[7,8] Patients are generally at their 4th or 5th decade with a mild male predominance.[9] Nonspecific symptoms like facial pain, swelling, nasal obstruction, rinorhea and epistaxis are usually seen during the clinical course of this malignancy. Due to this non spesific symptoms correct diagnosis may be delayed and tertiary syphilis, invasive fungal sinusitis, wegener's granulamatosis, other types of lymphomas and epithelial malign tumors of the paranasal sinuses should be ruled out during the diagnostic workshop.[2]

Our patient has presented with facial pain, swelling and rinorhea. Detailed endoscopic examination, radiologic investigation and advanced age of the patient is raised a concern of malignancy. Correct diagnosis is reached by endoscopic biopsy of the mass. Delaying of the correct diagnosis may lead fatal clinical course. Nasal culture for fungal and bacterial in-

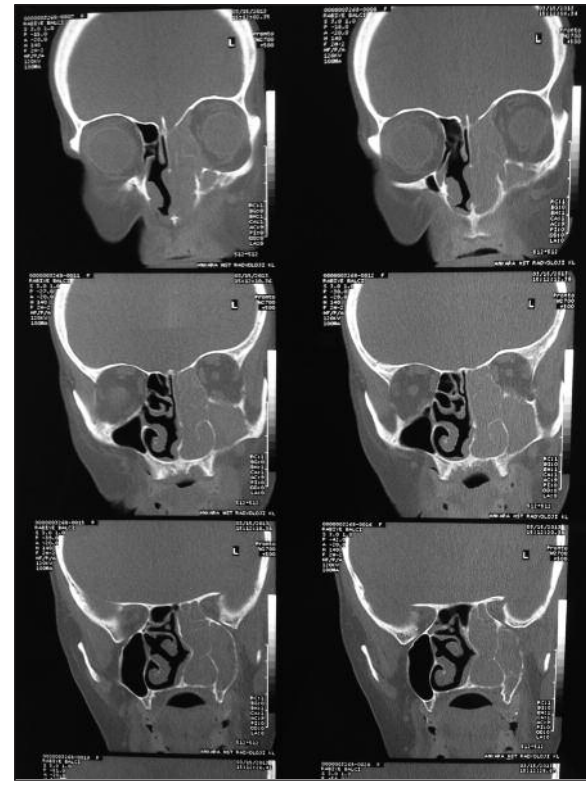


Figure 2. Coronal Computerized Tomography images of the patient.

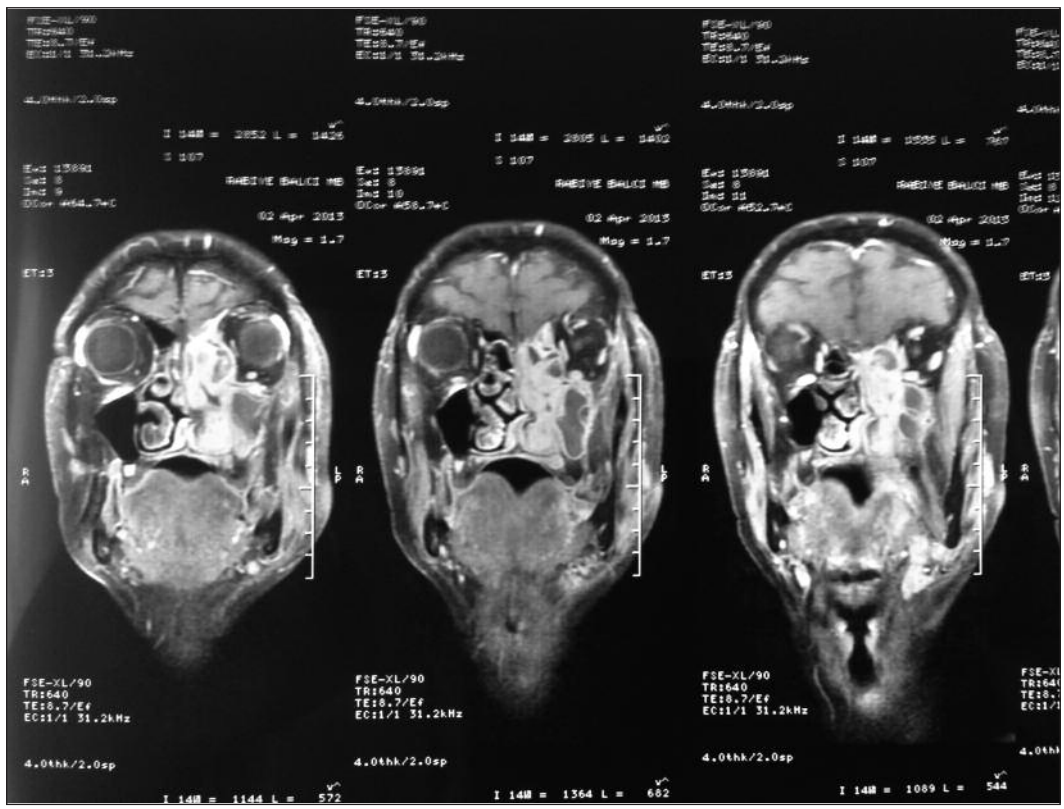


Figure 3. Coronal T1 Magnetic Resonance images of the patient.

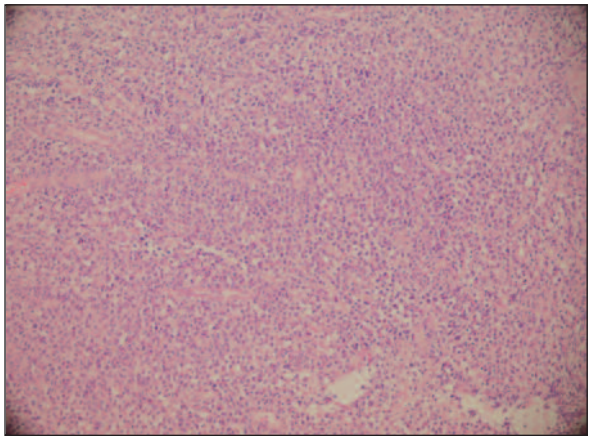


Figure 4. The lymphomatous infiltrate had diffuse, angiocentric growth pattern, the cells had irregular nuclei, some of them were elongated (x10 hematoxilen eosin).

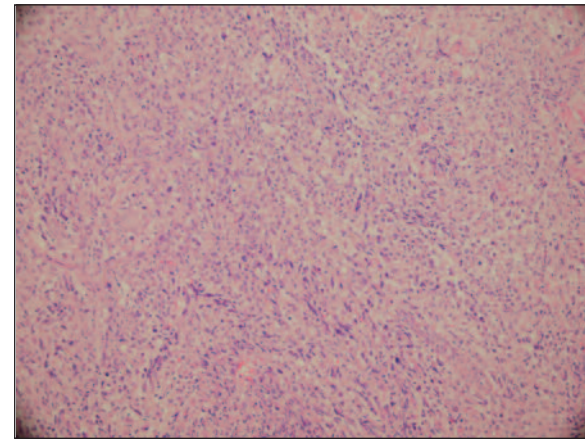
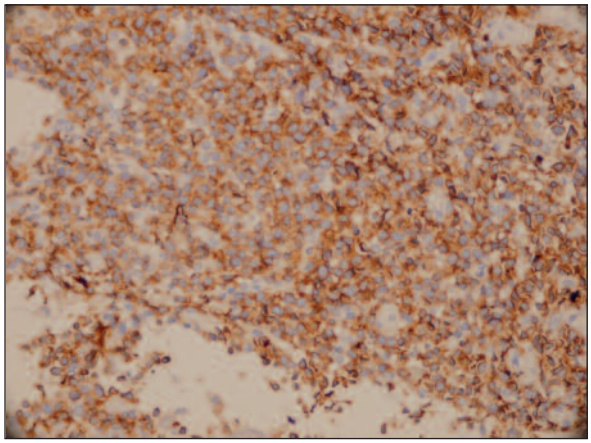


Figure 5. Mucosa was diffusely infiltrated by neoplastic cells admixture with small lymphocytes, histiocytes eosinophils (x10 hematoxilen eosin).



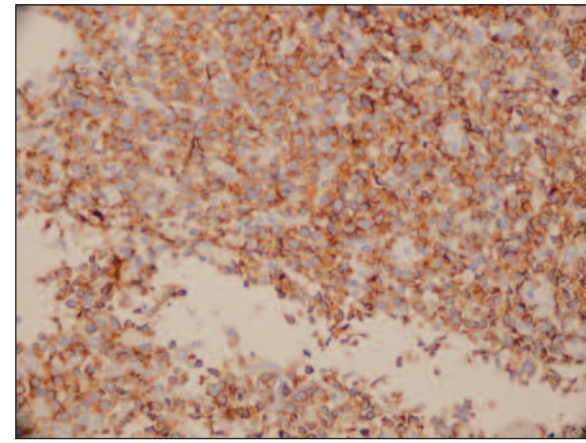


Figure 6,7. The neoplastic cells show strong staining for CD2, CD56 (x40).

fectious disease and histopathological investigation is essential for diagnostic study.[3] Angioinvasion and occlusion of the vascular network leads to have some difficulties during the histopathological evaluation. Wu et al. Reported %44 misdiagnosis and %22,5 repeating biopsies during the diagnostic study of the NK/T Cell lymphomas because of the angioinvasion property of the disease.[10] In our case we could be able to diagnose the malignancy through the second biopsy. Multiple biopsies should be taken to manage the misdiagnosis in the diagnostic studies of the paranasal sinus diseases.

Although nasal type NK/T Cell Lymphoma mostly seen in the nasal cavity and paranasal sinuses; it can also

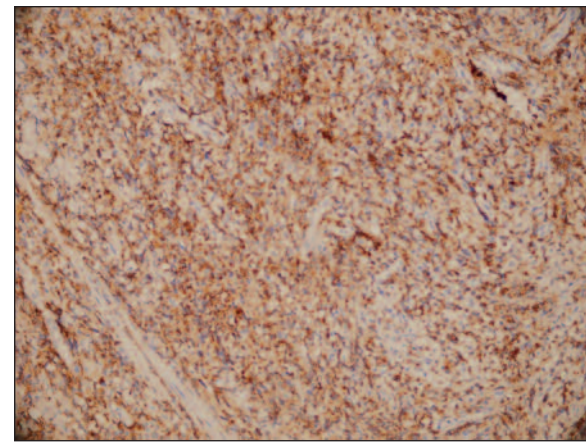


Figure 8. The neoplastic cells show strong granular staining for granzyme B.(x40).

be seen in the nasopharynx, oropharynx, oral cavity, hypopharynx and tonsils. Skin, liver and spleen are other places where NK/T Cell Lymphoma may also be seen.[2] Nasal cavity, maxilla and nasopharynx was invaded in our case.

There is no consensus on the treatment due to lack of reported large series. According to the limited series of the disease radiotherapy and chemotherapy seems to be effective in the treatment. Cyclophosphamide, doxorubicin, vincristine and prednisone are the mostly used chemotherapeutic agents with external radiotherapy.[2,10] Diseases limited to paranasal region are more amenable to treatment and has better prognosis. Patients who has locoregional involvement, metastatic diseases and type B symptoms (night sweating, loss of weight, fever) have poor prognosis. 5-year survival is reported between 35-85%. The patient presented in this paper has died 4 months after the treatment has started.

CONCLUSION

Nasal type NK/T Cell lymphoma is a rare and aggressive kind of Non-Hodgkin Lymphoma. Differantial diagnosis should be done carefully due to mimicking non-neoplastic diseases. Delaying in the diagnostic studies may lead fatality.

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