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CASE REPORT

Non-Hodgkin's lymphomas of the nose and paranasal sinuses: a case report

Burun ve paranazal sinüslerin Non-Hodgkin lenfoması: Olgu sunumu

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Primary Non-Hodgkin Lymphoma (NHL) of the nasal cavity is a rare neoplasm. They account for about 0.17-2% of all cases of NHL and 5.8% of all malignant neoplasms of the sinonasal region in the adult. A 37- year old male patient presented with left sided nasal obstruction and headache for a couple of weeks before the admission. Nasal examination revealed a massive poliposis. After medical therapy with corticosteroid, neither the polyposis nor his complaints showed any regression thus he underwent a nasal polypectomy operation. Biopsy results indicated an inflammatory nasal polyp. 2 weeks later, a rapidly progressive facial swelling, especially in left periorbital area and blurred vision occured. Second biopsy, which was also taken from left nasal cavity suggested again an inflammatory nasal polyp. To rule out any possible malignancy a third biopsy was conducted on the left maxillary sinus which indicated "T/NK cell lymphoma" of the sinonasal tract.

Key Words: Lymphoma, Non-Hodgkin/pathology/diagnosis/therapy; nose neoplasms/pathology/diagnosis/therapy; paranasal sinus neoplasms/pathology/diagnosis/therapy.

Nazal kavitenin primer Non-Hodgkin lenfomaları (NHL) nadir izlenen neoplazmlardandır. Bunlar tüm NHL olgularının %0.17-2'sini ve sinonazal bölgenin tüm malign neoplazmlarının %5.8'ini oluşturur. Otuz yedi yaşındaki erkek hasta bir kaç haftadır devam eden sol taraf burun tıkanıklığı ve baş ağrısı yakınmaları ile başvurdu. Nazal muayenede masif polipozis izlendi. Kortikosteroid tedavisine rağmen hastanın polipozisi ve yakınmaları düzelmedi ve hastaya nazal polipektomi yapıldı. Biyopsi sonucu enflamatuvar nazal polip olarak geldi. İki hafta sonra, hastada hızlı ilerleyen özellikle sol periorbital bölgede yoğunlaşan yüzde şişme ve görme bozukluğu yakınmaları başgösterdi. Sol nazal kaviteden alınan ikinci biyopsi sonucu da tekrar enflamatuvar nazal polip olarak belirtildi. Olası bir maligniteyi ekarte etmek için üçüncü biyopsi sol maksiller sinüsten alındı ve biyopsi sonucu sinonazal bölgenin "T/NK cell lymphoma"sı olarak geldi.

Anahtar Sözcükler: Lenfoma, Non-Hodgkin/patoloji/tanı tedavi; burun neoplazileri/patoloji/tanı/tedavi; paranazal sinüs/patoloji/tanı/tedavi.

Primary Non-Hodgkin Lymphoma (NHL) of the nasal cavity is a rare neoplasm that can be defined as an unusual site of origin for extranodal lymphoma in the presence of symptoms that can be initially referred to the nasal region.[1-8] They account about 0.17-2% of all cases of NHL and 5.8% of all malignant neoplasms of the sinonasal region in the adult.[9] Although lymphomas may be most common nonepithelial malignant tumors of the nose, an obstructive mass in this location is far more likely to

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be a carcinoma than a lymphoma. It is this rarity that may cause clinicians to dismiss them as a benign inflammatory infiltrate.[10]

CASE REPORT

A 37 year old male patient with no history of allergy had complaints of nasal obstruction and headache for a couple of weeks. Initial head and neck examination revealed out a left-sided nasal polyposis. After a three-week long medical therapy with corticosteroid neither the polyposis nor his complaints showed any regression thus he underwent a nasal polypectomy operation. Biopsy results indicated an inflamatory nasal polyp. About 2 weeks after the operation, rapidly progressive facial swelling, especially in left periorbital area and blurred vision occured in addition to his initial complaints, nasal obstruction and headache (Fig. 1). As an in-patient CT scan of the paranasal sinuses (Fig. 2) and a biopsy was taken from left nasal cavity. Again, the results suggested an inflammatory nasal polyp. To rule out any possible malignancy a secondary biopsy was conducted on the left maxillary sinus which indicated "T/NK cell lymphoma" of the sinonasal tract. The patient was then staged according to Ann Arbor classification as IIE.

DISCUSSION

The nasal cavity and surrounding paranasal sinuses are anatomically adjacent to Waldeyer's ring, which is recognised as a potential primary site for the origin of lymphomas. The main sites of involvement of NHL in the sinonasal region are: the intersinonasal wall; the middle hom; the nasal septum; the ethmoid often associated involvement of the maxillary sinus; the frontal sinus; and the sphenoid sinus, which is rarely involved. In addition there is important extension to neighbouring structures, such as the subcutaneous tissues of the cheek; the oral cavity, palatine tonsils and larynx; the orbit and pterygomaxillary fossa; and the central nervous system.[2,11] The most common initial signs of NHL of the sinonasal tract can be interpreted as a benign disease with a recent history. They are classified in Table I.[2,12]

Lymphomas are usually submucosal and on gross appearance differ from squamous cell carcinomas, which are usually ulcerative.[13]

The T/NK cell lymphomas have unique features that help to distinguish them from B-cell lymphomas. They are characterized by progressive, unrelenting ulceration and necrosis not seen in B-cell lesions. There is often severe destruction of the nasal septum and facial midline structures and are frequently associated with angioinvasion, coagulative necrosis, and epiteliotropism.[10]

The distinction betwen T and B cell lymphomas is based on the morphological characteristics of the neoplastic cells. With advances in immunohistochemistry, a significant proportion of these lymphomas were found to be T- cell phenotypes. This is especially true in Asian populations, in which greater than 90% of sinonasal lymphomas are T- cell. This high incidence of T- cell fenotypes has been

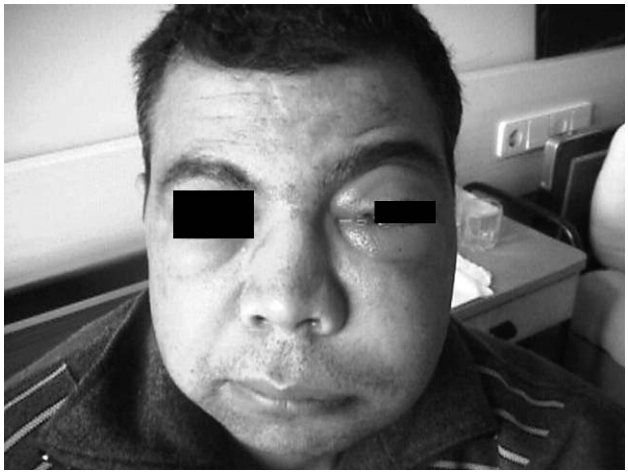

Fig. 1 - *Patient, frontal view; facial swelling, especially in left periorbital area.*

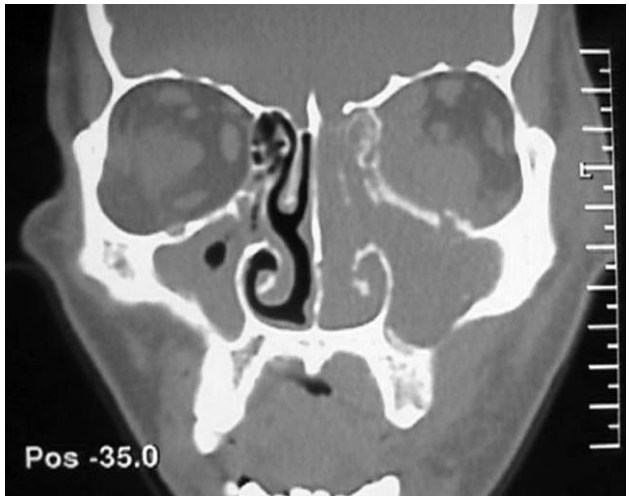

Fig. 2 - *CT scan of the paranasal sinuses.*

TABLE I

THE CLASSIFICATION OF SYMPTOMS IN NHL OF THE SINONASAL TRACT

Local symptoms	Nasal respiratory subobstruction; spontaneous epistaxis; mucosal or mucoprulent rhinorrhea; dry nasal mucosa;
Regional symptoms	Facial pain and toothache; headache; paraesthesia of the cheek; sense of fullness in the ear; epiphora; dyplopia; diminished visual acuity;
General symptoms	(Related to the evolution of the disease) sudden high fever; weight loss (>10%); nocturnal sweating; anorexia.

found to be unrelated to infection with human T-cell leukemia virus (HTLV-1).[10] In Western countries, B-cell lymphomas seem to predominate, reportedly constituting 55% to 85% of the sinonsal lymphomas.[14]

NHL is one of the most common malignancies in patients infected with human immunodeficiency virus (HIV); it occurs 25-60 times more frequently in HIV- infected patients than in the general population.[15] Numerous studies have demonstrated that both T-cell and NK cell lymphomas of the sinonasal region have a high incidence of Epstein-Barr virus (EBV) infection.[14,16-18] Unlike primary NHL of Waldeyer's ring, in which B cell phenotype expression is predominant,[19] NHL of the nasal cavity is a different disease[1,20,21] characterised by the predominance of the T Cell phenotype[14] and with poor prognosis.[1,4,6,7,16,19,20,22-24]

The poor prognosis for NHL of the nasal cavity reported by many authors has been linked to the presence of an agressive lymphoma in these patients. Prognosis for nasal lymphomas is worse than that for tumours originating in the paranasal sinuses and Waldeyer's ring.[9]

A particular characteristic of NHL of the nasal cavity is that it appears to be more resistant to the effects of chemotherapy than are other forms of NHL. Its resistance to the conventional chemotherapy may be associated with a frequent expression of glycoprotein p and with the loss in function of p53.[9]

Rare tumors, such as malignant melanomas and schwannomas can also originate from nose and paranasal sinuses.[25,26] In histopathological examination of the biopsies taken from this region, these rare tumors also have to be taken into consideration.

Although lymphomas are rare sinonasal tract neoplasms, it should be kept in mind, that biopsy should be taken from at least 2 different locations in massive polyp-like lesions of the nose and paranasal sinuses to rule out any malignancy.

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