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CASE REPORT

Extranodal nasal non-Hodgkin lymphomas with small NK/T cells - Case report

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ABSTRACT

A 40-year-old female patient presented to our clinic for bilateral nasal obstruction, anteroposterior mucopurulent rhinorrhea, recurrent micro-epistaxis, anosmia, left otodynia, fever and sweating. The ENT findings and CT scan detected a septal tumor with predominant extension in the left nostril, with erosive character of the osteo-cartilaginous septal structure, without sinusal involvement and left lateral-cervical adenopathy. Resection of the entire septal tumor formation was performed. The histopathological outcome revealed non-Hodgkin's small-type NK/T extranodal nasal-type lymphoma – immunohistochemical tests confirmed histogenesis.

KEYWORDS: non-Hodgkin lymphomas, nasal-type lymphoma, Epstein-Barr virus.

INTRODUCTION

Malignant lymphomas are a heterogeneous group of monoclonal tumor cells of the immune system. They are divided phenotypically into T-type lymphomas and B-type lymphomas. WHO classification splits T-type lymphoma in precursor T-cell lymphomas (T-lymphoblastic lymphomas) and peripheral T-cell lymphomas (the group to which belong the extranodal non-Hodgkin lymphomas with small NK/T cells –ENKTL). NK/T cell lymphomas are a rare and distinct subtype of tumors. These are predominantly extranodal and most of them are nasal. The most common clinical features of small NK/T extranodal nasal-type include nasal obstruction or nasal bleeding due to a lesion. Appropriate histopathology and immunophenotyping are essential to confirm the diagnosis. Epstein-Barr virus infection is always present when diagnosing such pathology.

The diagnosis is based on several parameters according to the therapeutic protocol:

I). Suspected NHL (non-Hodgkin lymphoma)

The presence of a growing lymph node or a few lymph nodes in an area where they have occurred consecutively (one at a time) or a tumor formation in any organ or tissue.

II). NHL confirmation

1. General analysis of blood with platelets and reticulocytes (no specific changes – when the bone marrow is not affected. In cases of bone marrow damage in aggressive NHL, bleeding cells can be detected in the haemogram, in the indolent-prolifocyte variants, lymphocytes)
2. Fine needle aspiration (cytological confirmation)
3. Biopsy of the lymph node or tumor (histological confirmation of LNH and the morphological variant)
4. Trepanobiopsis of the bone marrow
5. Puncture of the bone marrow

III). Determining the extent of LNH spreading (clinical stage), the presence of signs, general intoxication and biological signs (fever higher than 38° C, pronounced nocturnal sweating, weight loss 10% and more in the last 6 months – the presence of one of these signs is sufficient)[1]. There are important differences between localized and disseminated disease in terms of treatment and survival. This is the reason why the detection of the sensitivity of the extranasal disease by imaging, blood tests and histology is extremely important. For staging and prognosis of the disease the Ann Arbor system and the International

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Table 1
Ann-Arbor Staging[1,3]

Stage I:	Stage II:	Stage III:	Stage IV:
Involvement of a single lymph node region or a single extralymphatic site	Involvement of two or more lymph node regions on the same side of the diaphragm; it may include localized extralymphatic involvement	Involvement of lymph node regions on both sides of the diaphragm; it may include the spleen or extralymphatic extension	Diffuse extralymphatic disease (e.g. in liver, bone marrow, lung, skin)

Prognostic Index (IPI) are widely used (Table 1)[2].

The experience is limited since disease incidence is rare, and most treatment protocols are consensus-guided[4]. Non-Hodgkin's small-cell NK/T extranodal nasal-type lymphoma is extremely aggressive, with a low survival rate and low response to therapy. Unfavourable prognostic factors are advanced stages (III - IV) of the disease as well as bone marrow impairment; secondary determinations in CNS and infection-related factors (infections caused by disease or specific treatments) are complementary exacerbation factors. It is important to discover the disease in the early stage because, with the proper treatment, it has a much higher success rate. Prognosis has improved in recent years in the cases of localized disease, but it has remained a poor one in patients with disseminated disease[5]. ENKTL is both chemosensitive and radiosensitive. Therapeutic protocols include various schemes among which the most widely anthracycline-based CHOP-like regimens (e.g., cyclophosphamide, doxorubicin, vincristine, prednisone) used for aggressive B-cell lymphoma treatment were the main factors[6,7].

CASE REPORT

A 40-year-old female patient presented to our clinic for bilateral nasal obstruction, anteroposterior mucopurulent rhinorrhea, recurrent micro-epistaxis, anosmia, left otodynia, fever and sweating for 2 months, symptomatology for which she had been consulted in other clinics in her country where she was diagnosed with "hypertrophic rhinitis".

ENT findings (Figure 1 A,B) and the CT scan detected a septal tumor with predominant extension in the left nostril (Figure 2 A,B,C,D), with erosive character of the osteo-cartilaginous septal structure, without sinusal involvement and left lateral-cervical adenopathy. Resection of the entire septal tumor forma-

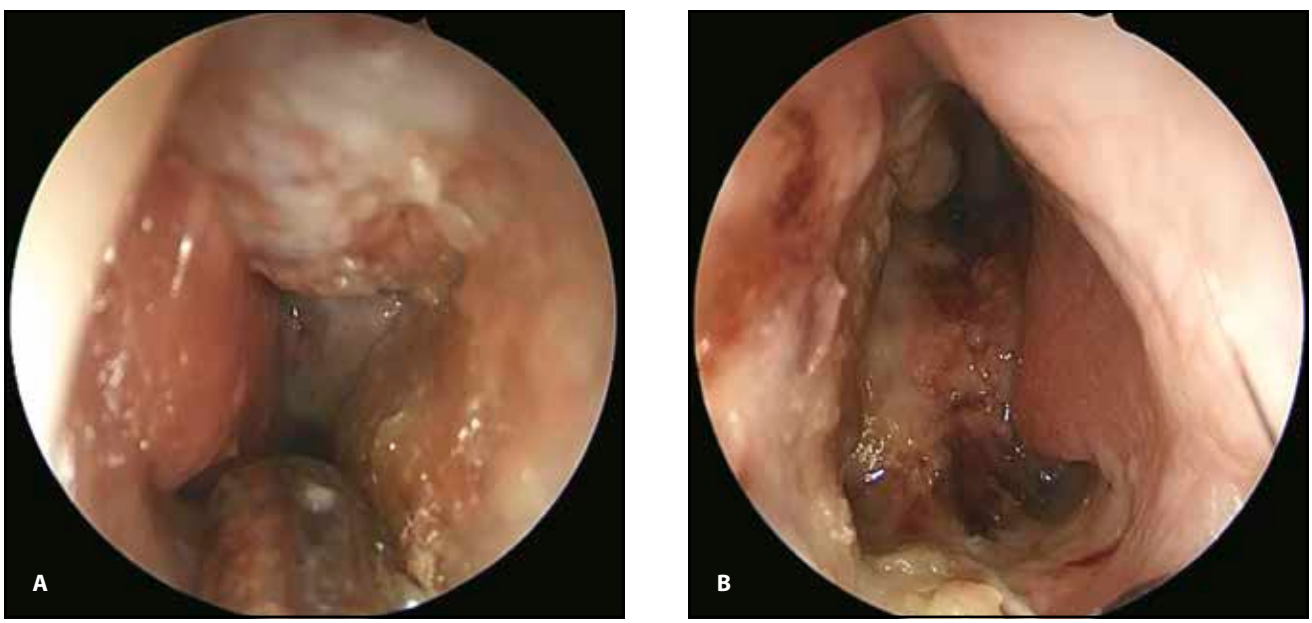

Figure 1 Nasal endoscopic examination - A. Septal tumor with extension into the right nasal fossa; B. Septal tumor with predominant extension into the left nasal fossa.

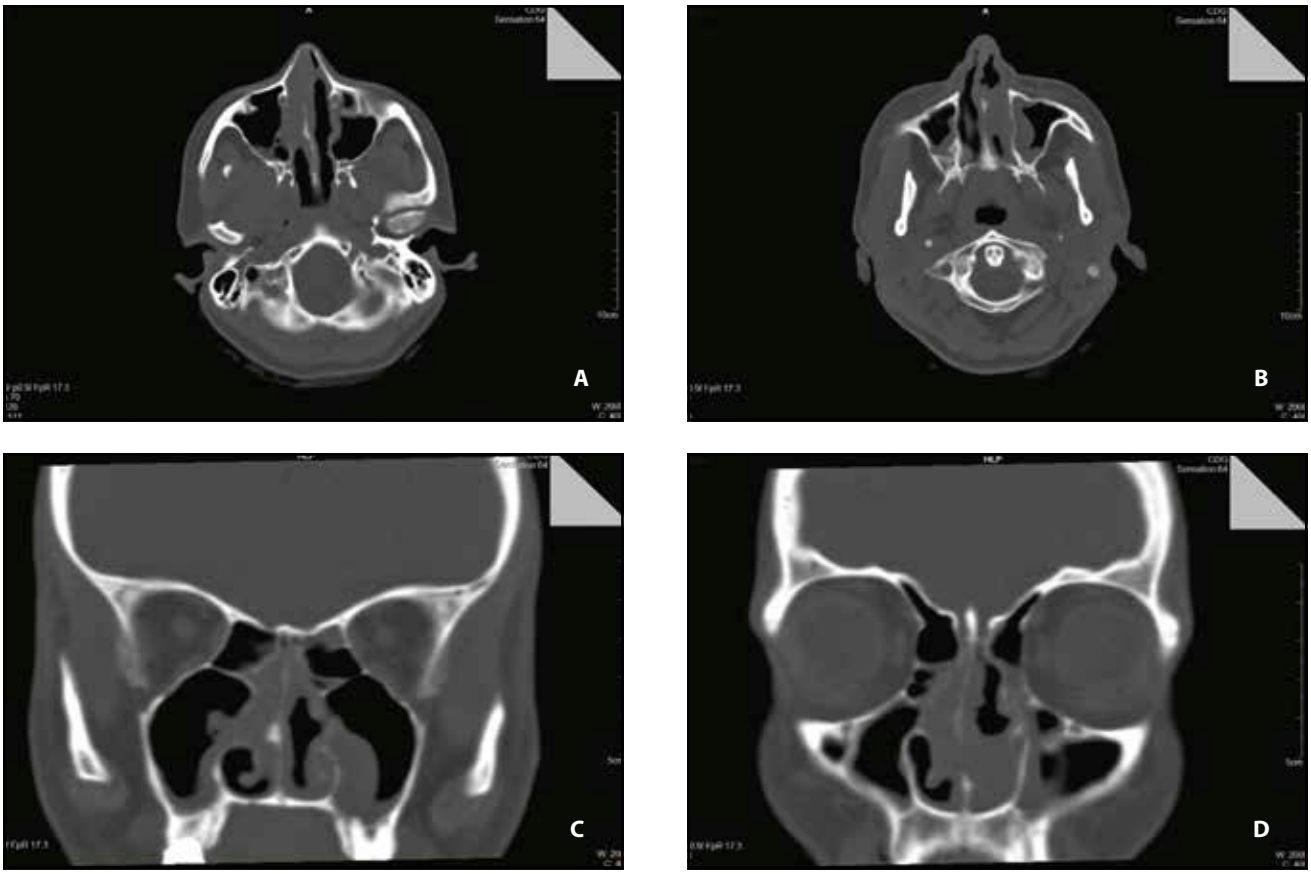

Figure 2 Cranio-facial CT Scan – axial slices (A,B), coronal slices (C,D). Nasal septal tumoral growth with extension predominantly into the left nasal cavity, having an erosive character of the osteo-cartilaginous septal structure, without sinus involvement; stagnating secretions in small quantity in the left maxillary sinus; no ethmoidal involvement, no skull base lysis.

tion was performed.

Under endoscopic control, a reddish, exophytic, brittle, fat-like appearance and slightly bleeding at instrumental palpation tumor was visualized, which occupied almost the entire left nasal cavity, originating from the nasal septum and with extension into the contralateral nasal cavity, covered with mucopurulent secretions.

With the help of the radiofrequency needle and the cold instruments (forceps, scissors and Blakesley forceps), we practiced, in a gentle manner, the quasi-total resection of the nasal septum, maintaining an anterior septal pillar, apparently with no tumor infiltration, going with a posterior excision to the sphenoidal rostrum, resecting quasi-completely superiorly the perpendicular blade of the ethmoid and inferiorly going to the floor of the nasal fossa. For the moderate bleeding, electrocautery hemostasis and bilateral anterior nasal packing with Surgicel and Merocel were performed (Figure 3).

The tumoral mass resected (Figure 4) during surgery was send to the histopathological examination which confirmed the extranodal nasal non-Hodgkin lymphoma with small NK/T cells.

Afflicting an extralymphatic organ and a lymph node on the same side of the diaphragm, the patient fell, according to the Ann-Arbor classification, in the second stage of the disease. For specialized treatment, she was directed to the haemato-oncologist.

One month after surgery, the patient showed a non-infiltrating septal tumor, without signs of tumor arrest or recurrence. The patient is currently under chemotherapy treatment (3 CHT sessions) in a good general condition. She will continue the treatment indicated by the haemato-oncologist and will return to the ENT control after 3 months.

DISCUSSIONS

Total resection of the septal tumor formation had a diagnostic purpose, a haemostatic target (recurrent epistaxis preoperatively) and nose permeability. There was no curative role regarding the background disease.

The particularity of the case resides in the complications that the tumour hides at first sight, which can endanger the patient's life, and also in the intertwining of different medical specialties, an essential point

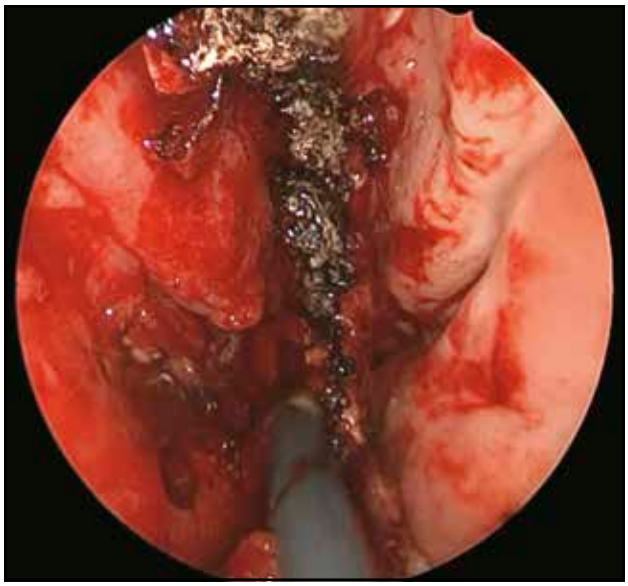

Figure 3 Intraoperative view after total tumor resection.

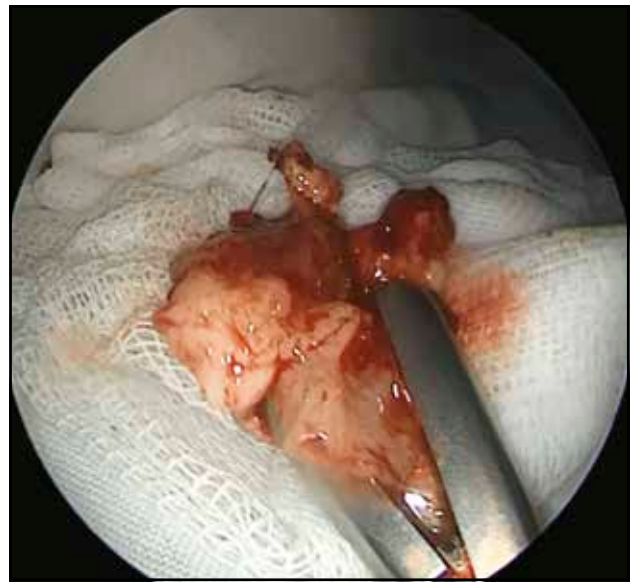

Figure 4 Tumoral mass.

when it comes to treating the patient, having in mind the desire to cure the disease. Although the surgical procedure did not have a curative role, it is important to remember that a correct diagnosis, even in the more evolved stages of disease, is crucial when it comes to making the right decision for the future surgical/oncological approach. Nevertheless, we must never forget that the quality of life is an important thing to consider while taking medical decisions.

CONCLUSIONS

NHL has an aggressive character and a poor prognosis, but with a better disease definition, staging and monitoring, the treatment of ENKTL becomes more rational. Early diagnosis is extremely important because most of the patients with localized nasal disease survive for long periods, while there is a vital negative prognosis in advanced stages. The diagnosis consists of the clinical suspicion plus the histopathological confirmation. Unlike aggressive B-cell lymphoma, in dealing with ENKTL, it should be known that some patients may run a less predictable or even chronic disease course. The main treatment is radiotherapy and chemotherapy for the lymphoproliferative neoplasm and, since late relapses are possible, a long-term vigilant follow-up is needed.

Conflict of interest: The authors have no conflict of interest.

Contribution of authors: All authors have equally contributed to this work.

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