Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiotherapy

Nasal NK/T Cell Lymphoma (NKTL): A Reported Case

Sara Amimi[1*], Souraya Bricha[1], C. El Alouani[1], S. Berkich[1], N. Oumghar[1], M. Darfaoui[1], A. El Omrani[1], M. Khouchani[1]

[1]Radiotherapy Department, Oncology and Haematology Hospital; Mohammed VI University Hospital, Marrakech, Morocco

DOI: https://doi.org/10.36347/sjmcr.2024.v12i09.021 | **Received:** 04.08.2024 | **Accepted:** 08.09.2024 | **Published:** 11.09.2024

***Corresponding author:** Sara Amimi
Radiotherapy Department, Oncology and Haematology Hospital; Mohammed VI University Hospital, Marrakech, Morocco

Abstract **Case Report**

Nasal NK/T cell lymphoma (NKTL), is an aggressive malignancy associated with Epstein Barr virus infection. Nasal NK/T cell lymphoma (NKTL), manifests as a necrotic process affecting nasal or upper aerodigestive structures and, rarely, extra nasal sites such as skin, and the gastrointestinal tract. The diagnosis is confirmed by histological and immunohistochemical analysis of the biopsy. Overall prognosis is poor. However, during the last two decades, advances in its clinicopathologic, genetic and molecular characterization have been achieved, as have changes in the chemotherapy regimens that, in combination with radiotherapy, are significantly improving the survival of these patients, especially in initial stages.

Keywords: Nasosinusal, NK/T-cell lymphoma, immunohistochemistry, radiotherapy.

INTRODUCTION

Nasal NK/T cell lymphoma (NKTL), also known as lethal midline granuloma, rarely manifests in the nose and paranasal sinus. It is very aggressive and carries a very poor prognosis; hence, early diagnosis and treatment determine the outcome of this disease. it's characterized by a necrotic process originating in the nasal cavity and extending to the medio-facial bone structures with centrifugal destruction of the facial bone. The advent of immunohistochemistry enabled its recognition by the WHO in 2001. NKTL is diagnosed mainly by exclusion as the clinical course and histopathology are often misleading and even the latest radiological investigations provide little help in making the diagnosis. The evolution is spontaneously fatal. Treatment Is based on radiotherapy and chemotherapy. We report one such case and discuss the specificities of this location.

MATERIALS AND METHODS

We present a case of a 34-year-old man affected by LTNK nasal treated conformational 3D radiotherapy at the oncology-radiotherapy department of the CHU Mohammed VI of Marrakech.

RESULTS

Patient aged 34, no pathological history, The early clinical history is characterized by the appearance of an inflammatory left naso-palpebral swelling with homolateral lacrimation, nasal obstruction on the left side and anosmia + agueusia. All evolving in a context of AEG made of asthenia and anorexia and febrile sensation not quantified.

Initial Examination: Irreducible inflammatory left exophthalmos with palpebral edema, Ulcerated lesion on palate, Free ganglion areas.

Nasofibroscopy: Whitish mass filling the entire left nasal fossa.

Initial Workup:
Facial CT: Lesional process centred on the left nasal fossa with left intraorbital extension measuring 56x80x61mm and blown thinning of the ethmoidal labyrinth and nasal cavity walls.

Facial MRI: Ethmoido-nasal tissue-like lesional process with bone lysis and left intraorbital and nasopharyngeal extension and meningeal invasion.

Biopsy: Location of a dense, suspicious lymphoid population suggestive of lymphomatous orig.

Immunohistochemistry: T/NK lymphoma: CD 20 CD5 CD 10 TDT negative; CD3 CD 56 and anti Granzyme B positive, Ki 67 100%.

CT TAP: No secondary localization except L5 spinal lesion and pulmonary micronodules.

Brain MRI: persistence and increase in size of locally infiltrating Eth-nasal tumour process with bone lysis and intraorbital extension, more marked on the left and nasopharyngeal, intracranial and increased meningeal invasion and appearance of frontal cerebral invasion.

Bone and bone marrow biopsy: nothing to report.

Patient started gelox chemotherapy.

Clinical Examination

General examination: WHO 2 in wheelchair, Regression of exophthalmos with persistent left palpebral infiltration, Filling of left NG sulcus, Wounds disappear at this level, Oral cavity and oropharynx nothing to report, Free lymph nodes.

The patient received locoregional external radiotherapy at a dose of 50Gy in 25 sessions. Sessions (2 Gy/fr; 5 fr/week), the patient was in complete radiological remission.

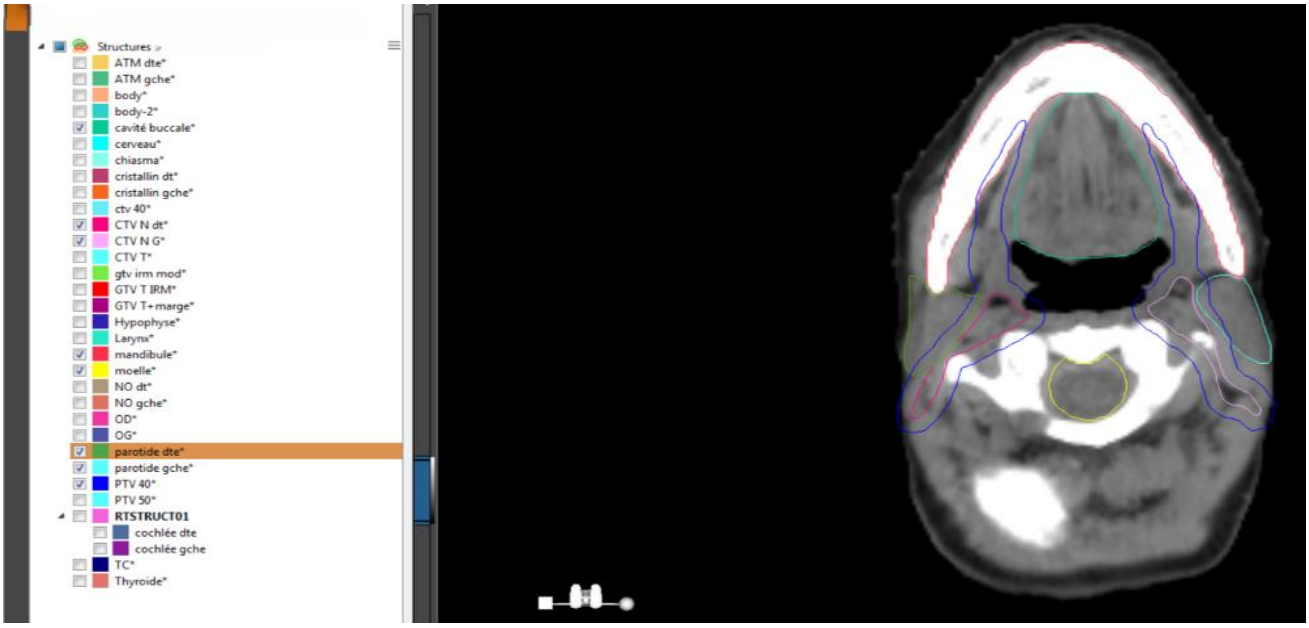

Figure 1: Contouring of nasal NKT lymphoma after chemotherapy with target volumes

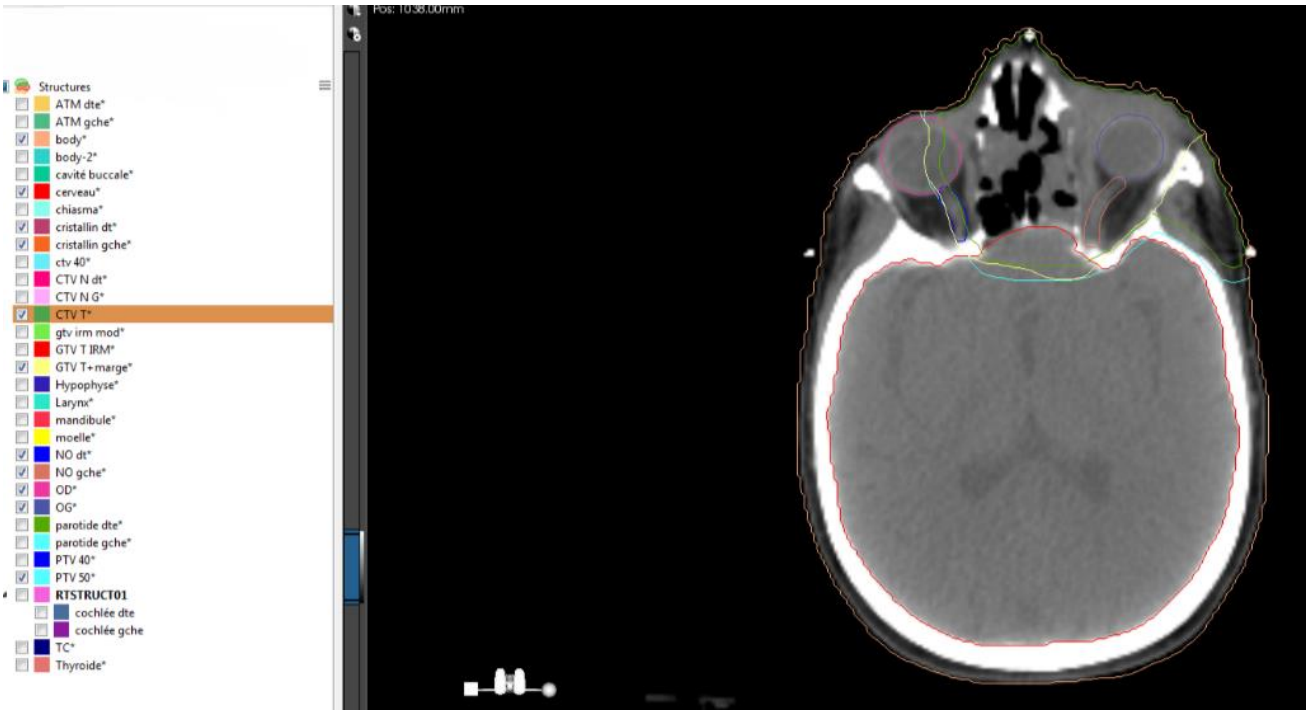

Figure 2: Contouring of nasal NKT lymphoma after chemotherapy with target volumes

DISCUSSION

Formerly known as 'centrofacial malignant granuloma'. The World Health Organization (WHO) has renamed this entity in its extranodal natural killer (NK) T-cell lymphoma type.

T/NK lymphoma is less common in Europe and North America than in Asia and South America [1]. It tends to affect young adult, which corresponds to our observations. The sex ratio is 3:1 and the median age at diagnosis is 50-60 years [2, 3].

The clinical presentation generally consists of local signs such as unilateral nasal obstruction, purulent and/or blood-streaked rhinorrhea blood, purulent and/or blood-streaked rhinorrhea, recurrent rhinorachinitis [4, 5]. General signs, such as fever, weight loss or night sweats, are rare and worrying. General signs such as fever, weight loss or night sweats are rare and of concern in the advanced stages of the disease. Signs of extension regional extension (swelling of the jaws or ophthalmic or auditory signs), pharyngeal or neurological disorders) may be the symptoms [4].

Clinical examination usually reveals an anterior lesion ulcero-necrotic lesion, bleeding on contact and filling the nasal cavity. The nasal cavity is generally located on the lateral wall [4, 6]. Involvement of the lymph nodes varies from one series to another and never exceeds 25% of cases [4, 7].

Computed tomography guides the diagnosis, often showing an appearance suggestive of a solid tumor with little or no contrast. Destruction of the bone structure is found in less than half of cases and especially in the case of large tumors.

MRI helps to assess extension to adjacent structures, differentiating tumor processes from inflammatory processes. Biological analysis systematically reveals an inflammatory syndrome.

The diagnosis is confirmed by histological and immunohistochemical analysis of the biopsy. Immunohistochemistry determines the phenotype of the lymphoid element and is essential to the diagnosis. NK etiology is confirmed by the expression of CD2, CD56 and cytotoxicity markers (T1a, Granzyme B, perforin); CD5 CD4 and CD8 are negative; CD3 expression is variable (absence of superficial CD3 marker) [6]. Association with EBV is almost systematic. In this case, only the immunohistochemical study has provided certainty. There is a problem of differential diagnosis in relation, for example, to Wegener's granulomatosis [4]

The treatment of T/NK lymphomas of the nasal cavity is not yet well codified; it depends mainly on the stage of the disease according to the ann arbor classification. For localized stages (stages I and II), external radiotherapy with a minimum dose of a round of the order of 52 Gy in conventional fractionation is recommended [8]. It results in complete remission in 40 to 80% of cases and five-year overall survival of between 40 and 59% [9]. Some teams, it generally involves anthracycline-based polychemotherapy multidrug therapy followed by external consolidation radiotherapy for patients under 60 years of age and the same with anthracycline-free multidrug therapy for patients aged over 60 [10]. Others continue to propose radiotherapy alone for patients with early-stage lesions, since the failure rate of treatment with chemotherapy is as high as 40%. Patients receiving after chemotherapy failure have a better prognosis [11]. The overall survival rate for all treatments combined, is around 37% [12].

CONCLUSION

Nasal locations for NK/T-cell lymphoma are rare. Diagnosis is founded on immunohistochemistry. Treatment associate's chemo- and radiotherapy. NKTL was characterized by its poor prognosis irrespective of clinical stage and therapy, even with treatment. The major issue at present is to standardize treatment protocols in multidisciplinary teamwork between onco hematologists and ENT physicians.

REFERENCES

1. Chim, C. S., Ma, S. Y., Au, W. Y., Choy, C., Lie, A. K., Liang, R., ... & Kwong, Y. L. (2004). Primary nasal natural killer cell lymphoma: long-term treatment outcome and relationship with the International Prognostic Index. *Blood*, *103*(1), 216-221.
2. Mestiri, S., Zeglaoui, I., Sriha, B., Belcadhi, M., Bouzouita, K., & Korbi, S. (2008, September). Les lymphomes T des fosses nasales et des sinus. In *Annales d'Otolaryngologie et de Chirurgie Cervico-faciale* (Vol. 125, No. 4, pp. 188-192). Elsevier Masson.
3. Harabuchi, Y., Imai, S., Wakashima, J., Hirao, M., Kataura, A., Osato, T., & Kon, S. (1996). Nasal T-cell lymphoma causally associated with Epstein-Barr virus: clinicopathologic, phenotypic, and genotypic studies. *Cancer: Interdisciplinary International Journal of the American Cancer Society*, *77*(10), 2137-2149.
4. Mestiri, S., Zeglaoui, I., Sriha, B., Belcadhi, M., Bouzouita, K., & Korbi, S. (2008, September). Les lymphomes T des fosses nasales et des sinus. In *Annales d'Otolaryngologie et de Chirurgie Cervico-faciale* (Vol. 125, No. 4, pp. 188-192). Elsevier Masson.
5. Lachiver, X., Salf, E., Baglin, A. C., Drewski, P., Baumelou, E., & Cudennec, P. Y. (1995). Les lymphomes non hodgkiniens primitifs des fosses nasales et des sinus: à propos de 8 cas. In *Annales d'oto-laryngologie et de chirurgie cervico-faciale*, *112*(5), 234-240.

6. Forcioli, J., Meyer, B., & Fabiani, B. (2005). Granulome malin centrofacial ou lymphome nasal T/NK. *EMC-Oto-rhino-laryngologie*, *2*(4), 390-400.
7. Chèze, S. (1996). *Lymphomes T/NK angiocentriques à localisation ORL ou pulmonaire: étude clinique, anatomopathologique et thérapeutique de 13 cas* (Doctoral dissertation).
8. You, J. Y., Chi, K. H., Yang, M. H., Chen, C. C., Ho, C. H., Chau, W. K., ... & Chiou, T. J. (2004). Radiation therapy versus chemotherapy as initial treatmentfor localized nasal natural killer (NK)/T-cell lymphoma: a single institute survey in Taiwan. *Annals of Oncology*, *15*(4), 618-625.
9. Mikhaeel, N. G., & Spittle, M. F. (2000). Nasal natural killer-cell lymphoma: A disease with very poor prognosis. *Clinical Oncology*, *12*(5), 295-297.
10. Ribrag, V., Hajj, M., Janot, F., Girinsky, T., Domenge, C., Schwaab, G., ... & Bosq, J. (2001). Early locoregional high-dose radiotherapy is associated with long-term disease control in localized primary angiocentric lymphoma of the nose and nasopharynx. *Leukemia*, *15*(7), 1123-1126.
11. Cheung, M. M., Chan, J. K., Lau, W. H., Ngan, R. K., & Foo, W. W. (2002). Early stage nasal NK/T-cell lymphoma: clinical outcome, prognostic factors, and the effect of treatment modality. *International Journal of Radiation Oncology* Biology* Physics*, *54*(1), 182-190.
12. Kim, G. E., Cho, J. H., Yang, W. I., Chung, E. J., Suh, C. O., Park, K. R., ... & Kim, B. S. (2000). Angiocentric lymphoma of the head and neck: patterns of systemic failure after radiation treatment. *Journal of clinical oncology*, *18*(1), 54-63.