Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/18687
DOI URL: http://dx.doi.org/10.21474/IJAR01/18687

RESEARCH ARTICLE

EXTRANODAL NK/T-LYMPHOMA, NASAL TYPE: CASE REPORT

Soukayna Lahmar, Samia Khelfi, Kawtar Sossy, Wissal Hassani, Fatima Zahraa Farhane, Zenab Alami and Touria Bouhafa

Radiation Oncology Department, Oncology Hospital, Hassan II University Hospital. Fez, Morocco.

Manuscript Info

Manuscript History
Received: 10 March 2024
Final Accepted: 14 April 2024
Published: May 2024

Keywords:-
Extranodal NK-T-Cell Lymphoma, Nasosinusal, Immunohistochemistry, Radiotherapy

Abstract

Extranodal NK/T-lymphoma, nasal type (ENKTCL-NT), is a highly aggressive malignancy linked to Epstein-Barr virus infection. This type of lymphoma primarily affects nasal or upper aerodigestive structures, resulting in a necrotic process. Despite its clinical stage or treatment, ENKTCL-NT is known for itsunfavorableprognosis,Wepresentis a case of a 57-year-old patient who presented with an ulcerated nasal swelling accompanied by nasal obstruction. Imaging revealed a tumorous process within the nasal cavity. Histological and immunohistochemical studies favored a diagnosis of NK/T lymphomanasal-type.The patient underwent primary chemotherapy followed by radiotherapy, achieving complete clinical and radiological response. This response has been maintained for three years after completing the treatment.

Introduction:-

NK/T lymphoma nasal-type was described in 1933 as a malignantmidfacialgranuloma, is a rareClinicopathologicentitycharacterizedby a necrotic process that starts in the nasal cavity and spreads to the medio-facial bone structures, causing centrifugal destruction of the facial bone,

The advent of immunohistochemistry enabled its recognition by the WHO in 2001 (1) .Evolution is spontaneously fatal, Radiotherapy and chemotherapy are the pillars of treatment (2) .

We reportcase of NKTL (Nasal NK/T lymphoma) and discuss the specifics of this location.

Case :

A 57-year-old patient with a medicalhistoryincluding type 2 diabetes and combinedsclerosis of the medullasoughtmedical attention due to an ulcerated nasal swelling, nasal obstruction, and one-sided purulent discharge. Uponexamination, an ulcerated nasal mass and retro mandibular adenopathywereobserved. A biopsyconfirmed a high-grade lymphoma, revealing CD3 positivity in tumorcells, absence of CD20 expression, low CD30 expression, no CD15 expression, intense CD6 expression in manyatypicalcells, and lack of cytokeratin expression, leading to a diagnosis of T/NK typelymphoma.

CT scans revealed a right-sided nasal tissue abnormalityextendinginto the facial soft tissues. (Figure 1)

Corresponding Author:- Soukayna Lahmar
Address:- Radiation Oncology Department, Oncology Hospital, Hassan II University Hospital. Fez, Morocco.

The case wasreviewed in a multidisciplinary cancer care meeting (PCM), resulting in a decision for chemotherapy: 4 cycles of methotrexate-L asparaginase. Subsequentevaluation scans demonstrated completeregression of the abnormality .(Figure 2)
Afteradditional discussion in the multidisciplinary cancer care meeting, the patient receivedexternal radiotherapyutilizingthe IMRT technique, receiving a total dosage of 50 Gy.(Figure3)
Threeyears post-treatment, the patient exhibitscompleteclinical and radiologicalrecovery
(Figure4)

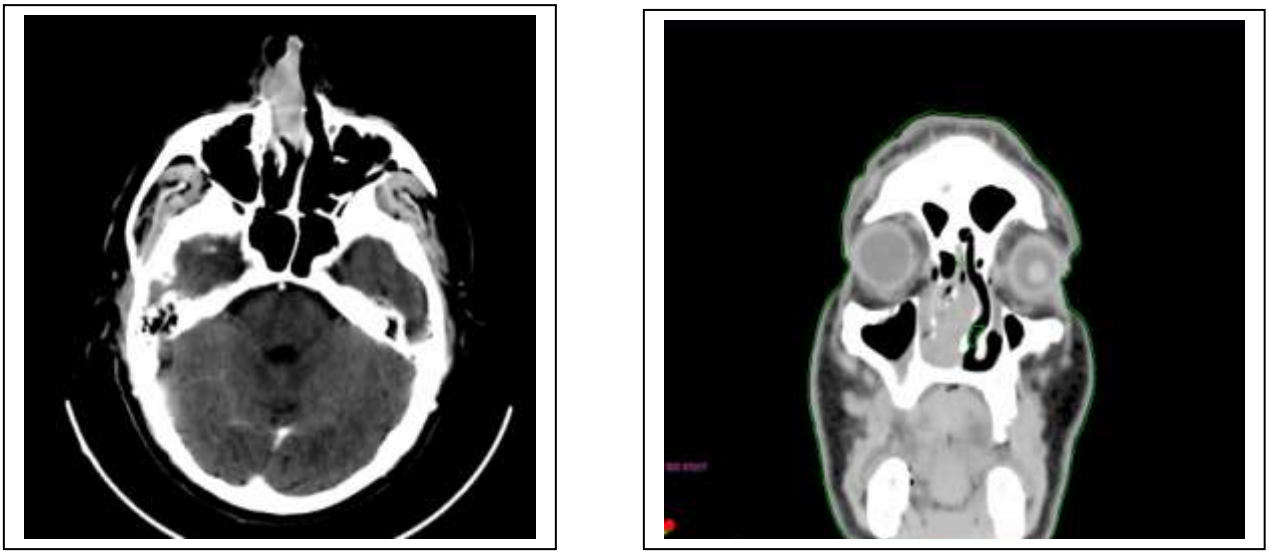


Figure 1 : Axial(A) and coronal (B) facial CT scan demonstrates a tissue processwithin the right nasal cavity, exhibitinginherent high density

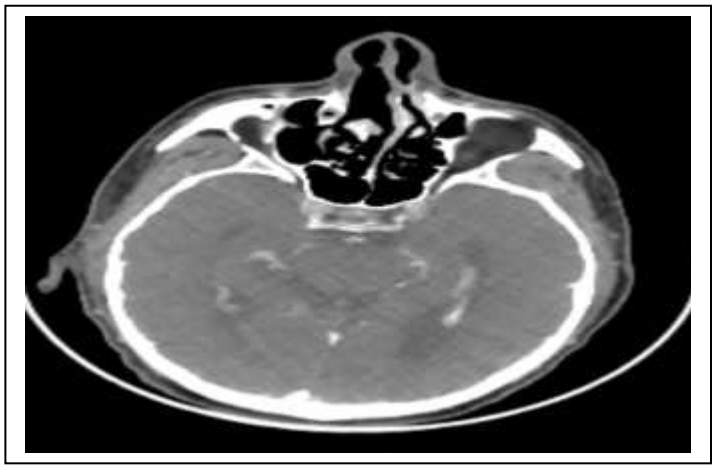


Figure 2 : Axial facila CT post-chemotherapy scanrevealingcompleteregression of thetumor process in the right nasal cavity

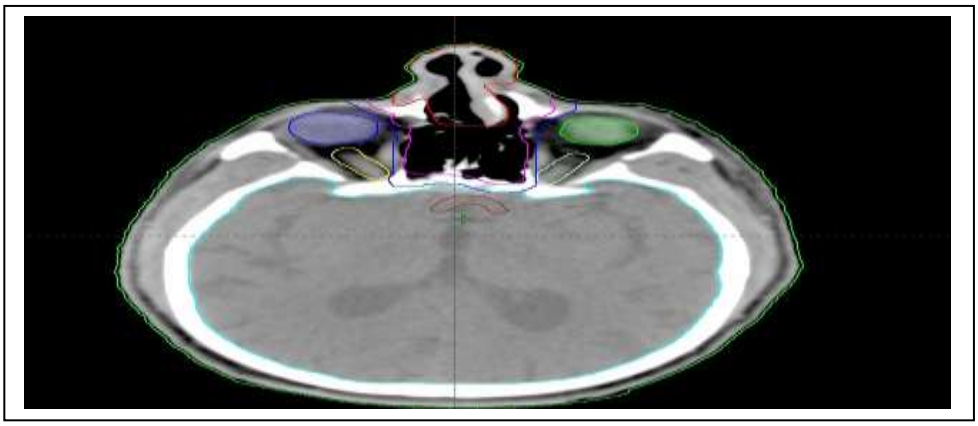
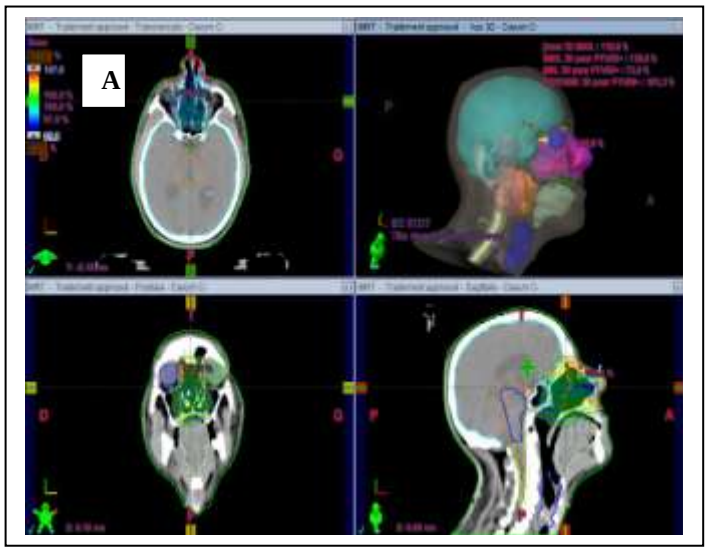


Figure 3 : Axial section of a dosimetric CT scan displaying the target volumes for radiotherapy(GTV in red, CTV in pink, and PTV in blue)



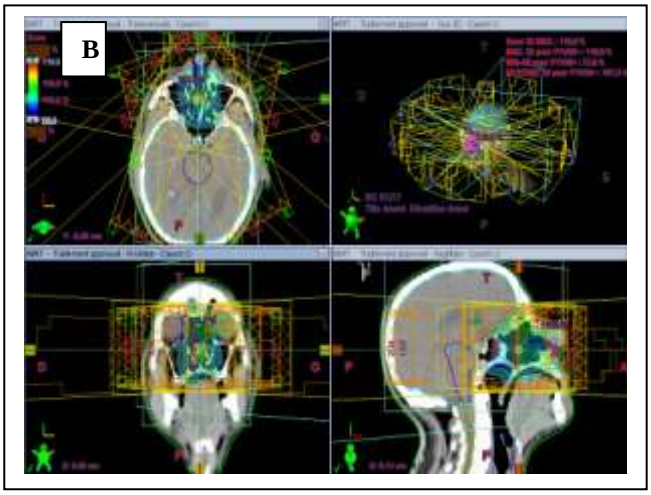


Figure 4 : Series of axial, sagittal, and coronal scans from the dosimetric CT depict the distribution of the dosagewithin thePTV (marked in blue) (A), alongside the configuration of beamsacross the three planes (axial, coronal, andsagittal) (B)

Discussion:-

Extranodal NK/T-lymphomasare a rare type of non-Hodgkin' lymphoma (NHL) that accounts for 5% 18% of all NHLs(3) .Nasal-type NK/T-lymphomasare a rare entity characterized predominantly by extranodal involvement, Unknownphysiopathologyexists. and associations exist with the Epstein Barr virus (EBV), genetics, geography, lifestyle, and environmental factors (1)

According to epidemiology, the majority of ENKTL-T patients are diagnosed in their fourth or fifth decade, with a male preponderance reported by different authors. In Asian, Central, and South American populations, there is a racial predisposition to the disease's normal distribution, althoughitisuncommon (less than 1%) in North America and Europe (4)

clinical symptoms are non-specific, including a single nasal blockage, purulent or bloody rhinorrhea,

recurrentepistaxis, or chronicsinusitis …

On the local extent of tumor involvement, conventional imaging techniques includingmagnetic resonance imaging (MRI) and CT provide helpful information. While MRI is better for soft tissue abnormalities, CT is more sensitive for bone lesions (5), ENKL is a fluorodeoxyglucose-avid lymphoma, so stagingwith positron emission tomography/computed tomography is recommended (6)

Consideringthatearlyclinicalsigns and symptoms are non-specific, diagnosis is confirmed by histologic and immunohistochemicalbiopsyanalysis, Classically, lesions demonstrate extensive angioinvasion and necrosis as well as positive staining for CD2, CD56, cytoplasmic CD3 (but not surface CD3), and cytotoxic markers. Rarely, cells may be of T-cell origin and express CD4, CD8, and/or CD7. It is essential to demonstrate the presence of EBV-encoded RNA by in situ hybridization for diagnosis (7).

Due to the rarity of the tumor and the absence of large randomizedcontrolledtrials, a diversity of therapy approaches have been adopted (8)
Initially,chemotherapy (CT) with anthracycline-containing regimens, principally CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone), was reserved for advanced stages, and radiation (RT) was thought to be the most effective treatment for ENKTCL-NT.

However, the high rate of systemic relapse (25–40%) remained an issue in patients treated with RT alone, and as a result, studies revealed a tendency toward survival in patients treated with combined CT and RT. Therefore, this combined modality is believed to bring better therapeutic outcomes than RT alone in most cases.

The preferred approach endorsed by the National Comprehensive Cancer Network (NCCN) is to recommend chemotherapy for patients who arefit to receive CT. Nevertheless, RT alone (50− 55 Gy) remains an option for patients with stage I/II localized nasal disease (demonstrated by PET/CT) who are unfit to receive CT and for elderly low-risk patients(9)

Alternative therapeuticmodalities, such as immunotherapy, EBV-specific cytotoxic T lymphocytes (CTLs) (see in "Future directions: promising therapeutic targets" section), and hematopoietic stem cell transplantation, may be useful to patients with advanced or relapsed/refractory ENKTCL-NT who continue to have poor outcomes (HSCT). (10)
Circulating EBV DNA can be measured to monitor the progression of the disease both during and after treatment. Elevated titers of this DNA are associated withadvanced disease, a bad prognosis, and a poor response to therapy. This has been confirmed for modern chemotherapies like SMILE, demonstrating a considerable effect on OS (9).

Conclusion:-

ENKL is an uncommon kind of T-lymphoma. Asparaginase-based chemotherapy and high-dose RT are frequently effective treatments for patients with limited-stage illness. The best course of treatment has not yet been established, whichisunfortunatebecause people withadvanced-stage and relapsed cancer frequently encounters an aggressive disease course.

In recent investigations, drugs including EBV- and LMP-CTLs and checkpoint blockage have demonstrated potential activity. As the data and science behind these methods develop, we anticipate that they will offer substitute curative methods, particularly for patients with severe disease.

Competing interests:
None.

Funding:
None.

Abréviations :
NKTL : Nasal NK/T celllymphoma .
PET/CT: Positron emission tomography/computed tomography
OS: Overall survival
CTLs: specific cytotoxic T-lymphocytes
LMP: latent membrane proteins
IMRT : Radiothérapie par modulation d'intensité
PCM: multidisciplinary cancer care meeting

Bibliographie:-

1. Aozasa K and Zaki MA. Epidemiology and pathogenesis of nasal NK/T-lymphoma:a mini-review. ScientificWorldJournal2011; 11: 422–428.
2. Sánchez-Romero C, Bologna-Molina R, Paes de Almeida O, Santos-Silva AR, Prado-Ribeiro AC, Brandão TB, Carlos R. Extranodal NK/T lymphoma, nasal type: An updated overview. Crit RevOncolHematol. 2021 Mar;159:103237.
3. Geller S, Myskowski PL, Pulitzer M. NK/T-lymphoma, nasal type, gdT-lymphoma, and CD8-positive epidermotropic T-celllymphomaclinical and histopathologic features, differential diagnosis, and treatment. SeminCutan Med Surg2018;37:30–8
4. Harabuchi Y, Takahara M, Kishibe K, et al. Extranodalnatural Killer/T-Celllymphoma, nasal type: basic science and clinical progress. Front Pediatr. 2019;7:141.
5. Ooi GC, Chim CS, Liang R, et al. Nasal T-cell/natural killer lymphoma: CT and MR imaging features of a new clinicopathologic entity. AJR Am J Roentgenol. 2000;174(4):1141–1145.
6. Moon SH, Cho SK, Kim WS, et al: The role of 18F-FDG PET/CT for initial staging of nasal type natural killer/T-lymphoma: A comparisonwithconventionalstagingmethods. J Nucl Med 54:1039-1044, 2013
7. Allen PB, Lechowicz MJ. Management of NK/T-CellLymphoma, Nasal Type. J OncolPract. 2019 Oct;15(10):513-520. doi: 10.1200/JOP.18.00719. PMID: 31600461; PMCID: PMC6790879.
8. se E, Kwong Y-L. The diagnosis and management of NK/Tcelllymphomas. J HematolOncol. 2017;10:85. https://doi. org/10.1186/s13045-017-0452-9.
9. Sánchez-Romero C, Bologna-Molina R, Paes de Almeida O, Santos-Silva AR, Prado-Ribeiro AC, Brandão TB, Carlos R. Extranodal NK/T lymphoma, nasal type: An updatedoverview. Crit RevOncolHematol. 2021 Mar;159:103237. doi: 10.1016/j.critrevonc.2021.103237. Epub 2021 Jan 22. PMID: 33493634.
10. Jeong SH. Extranodal NK/T lymphoma. Blood Res. 2020 Jul31;55(S1):S63-S71. doi: 10.5045/br.2020.S011. PMID: 32719179; PMCID: PMC7386895.