International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

A Comprehensive Case Study on Extranodal NK/T - Cell Lymphoma, Nasal Type: Clinical Insights and Challenges

Loong Jie Jun[1], Priyanka Menon[2]

[1]MD, Medical Officer, Department of Otorhinolaryngology, Hospital Selayang, Lebuhraya Selayang Kepong, Selangor, Malaysia
Email: *jun_jiejun[at]hotmail. com*

[2]MS ORL – HNS, Otorhinolaryngology Specialist, Department of Otorhinolaryngology, Hospital Selayang, Lebuhraya Selayang Kepong, Selangor, Malaysia

Abstract: ***Extranodal NK/T - cell Lymphoma of nasal type is a rare and highly aggressive subtype of non - Hodgkin lymphoma, closely associated with the Epstein - Barr virus. It exhibits a poor prognosis and limited responsiveness to available treatments irrespective of clinical stage and therapy. We report a case of a 54 years old gentleman who presented to the hospital after being unwell for one week. Histology study revealed lymphoma cells which are polymorphous, admixed with small lymphocytes, angiocentric growth invasion and positive immunohistochemistry for CD2, CD 56, cytoplasmic CD3+, surface CD3−ve. Patient was referred for further treatment however, died while ongoing treatment. This case study adds valuable insights into the clinical presentation, diagnosis, and challenges in managing Extranodal NKT - Cell Lymphoma, emphasizing the need for increased awareness and research in this area.***

Keywords: Extranodal NK/T - cell lymphoma, nasal type, non - Hodgkin lymphoma, Epstein - Barr Virus Infections

1. Introduction

Extranodal Natural Killer/T - cell lymphoma (ENKTCL), Nasal Type is an uncommon and aggressive form of Non - Hodgkin's Lymphoma that is strongly associated with the Epstein - Barr Virus [1] [3] [5] [6]. It is more prevalent in South - East Asia and Central and South America [5] [6]. Most commonly it occurs at the upper aerodigestive tract [1] [3] [4] and can also be primarily or secondarily involving the extranasal sites, like the skin, termed extranasal ENKTCL [5] [6]. Due to the non - specific clinical presentation of ENKTCL, it can be misdiagnosed as non - neoplastic conditions. A definitive diagnosis is made based on histopathological, immunological, and molecular study findings. We report a case of a male patient presented with an advanced stage of ENKTCL, nasal type with cutaneous involvement.

2. Case Report

A 54 - year - old male patient, a smoker, with no known medical illness prior presented with complaints of bilateral nasal pain, nasal block, clear nasal discharge and diplopia for one week. He also complained of palpable reddish skin lesions scattered over the chest, upper back, neck and face. He otherwise denies constitutional symptoms, high - risk behaviours, tuberculosis and leprosy contact.

On examination, the patient had left oculomotor nerve palsy, and other cranial nerves were intact. There was no neck, axillary, inguinal nodes or hepatosplenomegaly. Skin examination noted erythematous nodules and patches over the chest, upper back neck and face, a few with violaceous centre, non - blanchable with firm to hard in consistency (Figure 1). A rigid nasoendoscopy performed noted a smooth, reddish and vascularised mass occupying the entire right nostril (Figure 2) and a smooth mass seen beneath the left inferior turbinate.

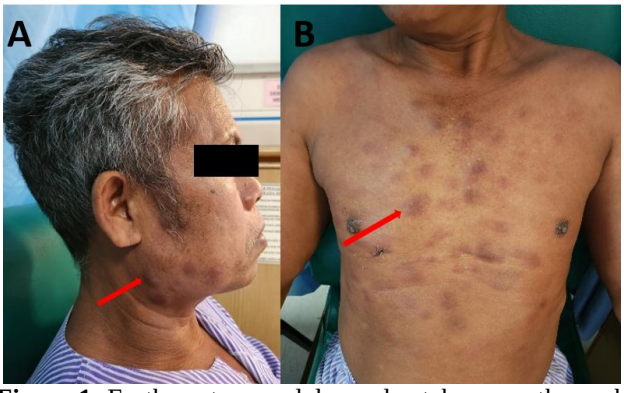

Figure 1: Erythematous nodules and patches over the neck and face (A) And chest (B)

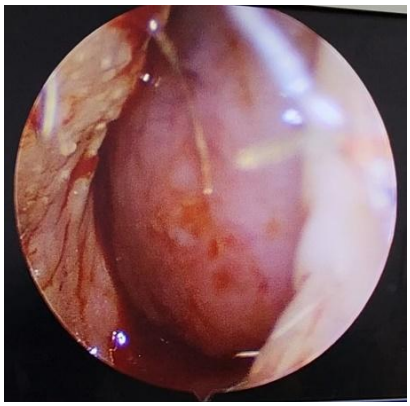

Figure 2: Rigid nasoendoscopy findings of smooth, reddish and vascularised mass occupying the entire right nostril

The patient was not anaemic, renal and liver profile was normal, LDH and CRP were not raised, HIV, hepatitis B, hepatitis C and syphilis were non - reactive, and sputum for acid - fast bacilli was negative.

Contrast Enhanced CT Scan of the Brain, Orbit, Paranasal sinus and Neck demonstrated right posterior nasal cavity and right nasopharyngeal ill - defined poorly enhancing lesions with the erosion of the floor of the right ethmoid sinus.

Intraoperatively, an irregular friable mass was seen occupying the left nasal cavity and an irregular mass was seen arising from the right side of the septum, right inferior end of the inferior turbinate, middle turbinate and fossa of Rosenmuller. A skin biopsy was taken from the chest skin lesion.

He was diagnosed with extranodal NK/T cell lymphoma with cutaneous involvement. The patient was referred to the oncology department, administered one cycle of CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) but he died soon after.

3. Discussion

Extranodal NK/T - cell lymphoma, nasal type (ENKTCL) is a rare and aggressive peripheral T - cell lymphoma. The median survival is 4.2 years [6] but varied greatly by stage, with only 7 months for stage IV disease [7]. It is more common in the middle age with male predominance. ENKTCLis almost exclusively extranodal in presentation [7] and the typical presenting features are limited to local symptoms such as nasal obstruction, epistaxis, and rhinorrhea [1] [2] [4] [6] which mimics sinusopathy leading to delay in the arrival of correct diagnosis. Most often present as localized disease, however, a subset presents with widespread dissemination to the skin, gastrointestinal tract, bone marrow and peripheral blood involvement [6] [7]. The disease is locally invasive in approximately 50% of patients. Differential diagnosis includes other lymphomas, nasopharyngeal squamous cell carcinoma, tertiary syphilis, Wegener granulomatosis and fungal infections [5]. A biopsy is necessary to confirm the diagnosis using microscopy, immunohistochemistry, and Epstein - Barr encoding region (EBER) in situ hybridization [8]. Histopathologically, ENKTCL features variable sizes of atypical lymphocytes with plasmocytes, eosinophils, histiocytes [2] [3] [4] [7] and the presence of angiodestruction and angiocentricity [1] - [5] [7] [9]. The diagnosis is further reinforced with the presence of EBV by in situ hybridization as EBV has been implicated in disease pathogenesis [2] [5] - [9]. High proliferation rate (Ki - 67>60%) predicts a poorer prognosis. Immunohistochemistry, the natural killer (NK) cell marker CD56 and cytotoxic markers granzyme B and TIA - 1 are expressed and detectable. T - cell markers demonstrates positive for CD2, cytoplasmic CD3+ and surface CD3 - ve [2] - [5] [7] [9]. T - cell markers CD4, CD8, CD5 and CD 7 are usually negative [2] [7] [9]. In this case, a biopsy of the nasal lesion and skin biopsy showed lymphoma cells which are polymorphous, medium to large in size, hyperchromatic, admixed with small lymphocytes, histiocytes, plasma cells and angiocentric growth invasion. The patient's immune - phenotyping was positive for CD2+, CD 56+, CD3 negative surface (-), CD3 positive cytoplasmic (+), CD4 - , CD5 - CD 8 - , CD7+, cytotoxic molecule TIA1+, positive EBER insitu hybridization and Ki - 67 proliferative index more than 70%. Conventional imaging techniques such as computed tomography (CT) and magnetic resonance imaging are useful for assessing local tumour invasion, but not sensitive enough to detect subclinical metastases [8] [9]. Extranodal NK/T - cell lymphoma, nasal type (ENKTL) isfluorodeoxyglucose (FDG) - avidin nearly 100% of cases and it is important to note that the maximum standardized uptake values in ENKTL tend to be lower when compared to diffuse large B - cell lymphoma [9]. Therefore, positron emission tomography (PET) is recommended to be performed at diagnosis in order to stage the disease [8] [9]. Additional studies, such as bone marrow biopsy is performed for staging purpose [8] [9]. A combination of radiotherapy and chemotherapy is considered the gold standard [3] [8] and currently ongoing further studies on new protocols and therapies such as targeted therapy, stem cell transplantation, and immunotherapy targeting the EBV.

4. Conclusion

Extranodal NK/T - cell lymphoma, nasal type is a rare but aggressive tumour. This case underscores the aggressive nature of Extranodal NKT - Cell Lymphoma, nasal type, and highlights the challenges in its diagnosis and management. It emphasizes the importance of early recognition and the need for further research in treatment strategies.

References

[1] Marks JA, Ghandour Z, Collins V, Schmieg J, Atkinson T. Extranodal NK/T - Cell Lymphoma, Nasal Type: A Case Series. J Cancer Treatment Diagn. (2022); 6 (3): 1 - 5
[2] Neves Cavada M, Silveira Martha A, Sgarabotto Pezzin L, Mazzaferro Krebs J, Mazzini Steffen L, Schultz Maahs G. Extranodal NK/T - Cell Lymphoma, Nasal Type—Case Report of 2 Cases. Ear, Nose & Throat Journal.2020 Sep; 99 (8): NP91 - 3.
[3] Lopes[1] LN, de Oliveira Sato[1] RA, Pinto[1] CA, de Souza Lima JA, Ferreira[1] B, de Moraes CA. NK/T cell lymphoma, nasal - type (rare, rapidly evolving, mutilating, and highly lethal lymphoid neoplasm): a case report.
[4] Varshney AN, Patidar R, Malhotra M, Behera M, Nagar M, Kataria SP. Extranodal NK/T - cell lymphoma of the nasal type with predominant T - cell markers: A rare subtype of rare disease entity. International Journal of Medicine and Public Health.2015; 5 (4).
[5] Tlholoe MM, Kotu M, Khammissa RA, Bida M, Lemmer J, Feller L. Extranodal natural killer/T - cell lymphoma, nasal type: 'midline lethal granuloma. 'A case report. Head & face medicine.2013 Dec; 9: 1 - 5.
[6] Li S, Feng X, Li T, Zhang S, Zuo Z, Lin P, Konoplev S, Bueso - Ramos CE, Vega F, Medeiros LJ, Yin CC. Extranodal NK/T - cell lymphoma, nasal type: a report of 73 cases at MD Anderson Cancer Center. The American journal of surgical pathology.2013 Jan 1; 37 (1): 14 - 23.
[7] Allen PB, Lechowicz MJ. Management of NK/T - cell lymphoma, nasal type. Journal of Oncology Practice.2019 Oct; 15 (10): 513 - 20.
[8] C. Sánchez - Romero *et al.*, "Extranodal NK/T cell lymphoma, nasal type: An updated overview, " *Critical Reviews in Oncology/Hematology*, vol.159. Elsevier

Ireland Ltd, Mar.01, 2021. doi: 10.1016/j. critrevonc.2021.103237.

[9] J. A. Van Doesum, A. G. H. Niezink, G. A. Huls, M. Beijert, A. Diepstra, and T. Van Meerten, "Extranodal Natural Killer/T - cell Lymphoma, Nasal Type: Diagnosis and Treatment, " *HemaSphere*, vol.5, no.2. Wolters Kluwer Health, Feb.12, 2021. doi: 10.1097/HS9.0000000000000523.