

Primary Nasopharyngeal Lymphoma in a 36 Year Old Male Patient: Diagnostic And Treatment Challenges

¹Dr. Angelina Shiny.P, ²Dr. BV Subramanian, ³Dr. Pranabandhu Das, ⁴Dr. AY Lakshmi

⁵Dr.TC Kalawat, ⁶Dr. N Rukmangadha, ⁷Dr. V Raga Bhavana

^{1,7}Junior Resident, ^{2,4,5,6}Professor, ³Associate Professor.

^{1,2,3}Department of Radiation Oncology, ⁴Department of Radiology,

⁵Department of Nuclear Medicine, ⁶Department of Pathology,

Sri Venkateswara Institute Of Medical Sciences(SVIMS), Tirupati

***Corresponding Author:**

Dr. Pranabandhu Das

Associate Professor , Department of Radiation Oncology

Sri Venkateswara Institute Of Medical Sciences(SVIMS), Tirupati

Type of Publication: Original Research Paper

Conflicts of Interest: Nil

Abstract

Nasopharyngeal lymphoma is a rare primary malignancy of the head and neck originating from the nasopharynx. The signs and symptoms of nasopharyngeal lymphomas and carcinomas are often found to have similarities. Histopathologically undifferentiated nasopharyngeal tumors lack the evidence of lineage differentiation between lymphoma and undifferentiated carcinoma on the basis of routine light microscopic morphology. Distinction between these lineages is important for treatment and prognostication. We present here a case of 36 year old male patient who presented with neck swellings and was initially diagnosed as undifferentiated nasopharyngeal carcinoma, but later confirmed to be of lymphoma following functional imaging with whole body 18FDG PET-CT and immunohistochemistry.

Keywords: NIL

Introduction

Lymphomas are the second most common neoplasms in nasopharynx after squamous cell carcinoma. Incidence is 2.5%. Oral and parapharyngeal regions constitute the second most affected sites by extra nodal lymphomas⁽¹⁾ Nasopharyngeal lymphoma is a rare extranodal lymphoma with various histological subtypes. Diffuse large B cell lymphoma and NK/T cell lymphoma are the two most common types^(2,3). Waldeyer's ring is the most common site for the occurrence of lymphomas⁽²⁾. Immunohistochemistry plays an important role in distinguishing lymphoma from undifferentiated nasopharyngeal carcinoma.

Case Report:

A 36 year old male patient presented with complaints of neck swelling which gradually increased in size

over a duration of 6 months, associated with pain, dyspnoea on exertion and epistaxis. On examination bilateral lymphadenopathy was noted with large conglomerate node involving left level Ib,II,III,IV,V largest m/s 10x10cms, on right side level Ib,II,III region m/s 3x3cm. MRI neck contrast study showed irregular ill defined T2 hyperintense, T1 hypointense lesion noted involving the roof, posterior wall, right and left walls of nasopharynx extending into bilateral parapharyngeal fat abutting bilateral CCA, ICA and deep lobe of right parotid gland, the lesion is extending into left foramen ovale, m/s 5.6x3.1x7.8cms. Multiple enlarged bilateral level II, left level III, IV lymph nodes noted largest m/s 7.2x4.1cm causing compression over trachea and CCA, as shown in figure 1. Biopsy from the nasopharyngeal growth showed Non keratinizing

undifferentiated nasopharyngeal carcinoma, as shown in figure 2

Figure 1- MRI contrast study images A (sagittal) & B (axial) sections showing the lesion in nasopharynx with cervical lymphadenopathy

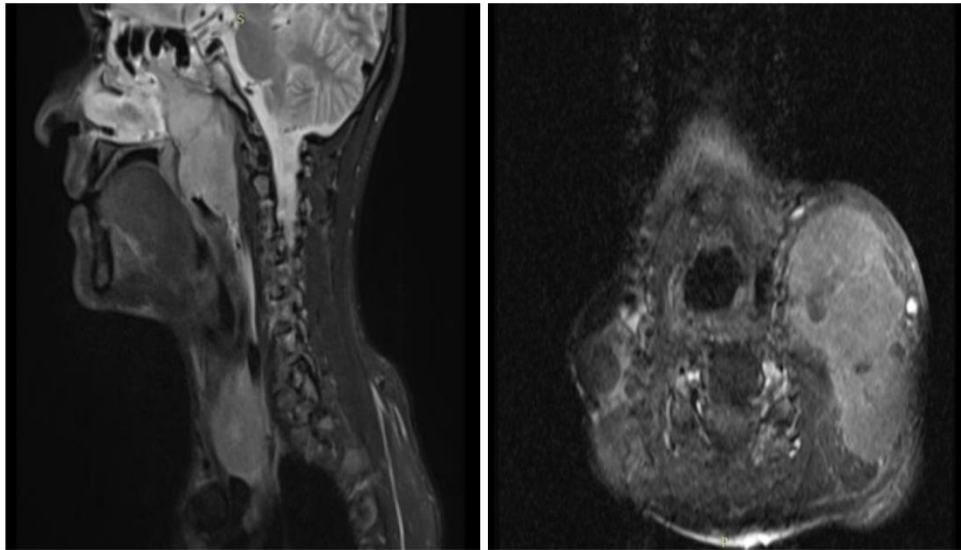
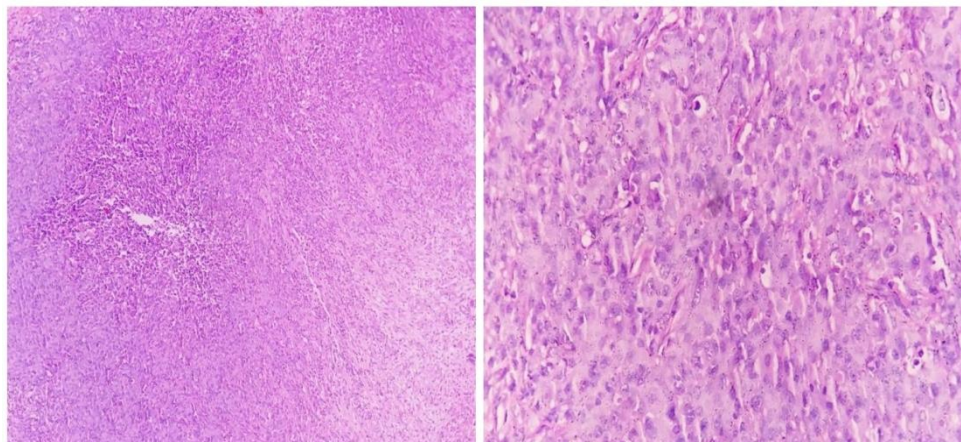
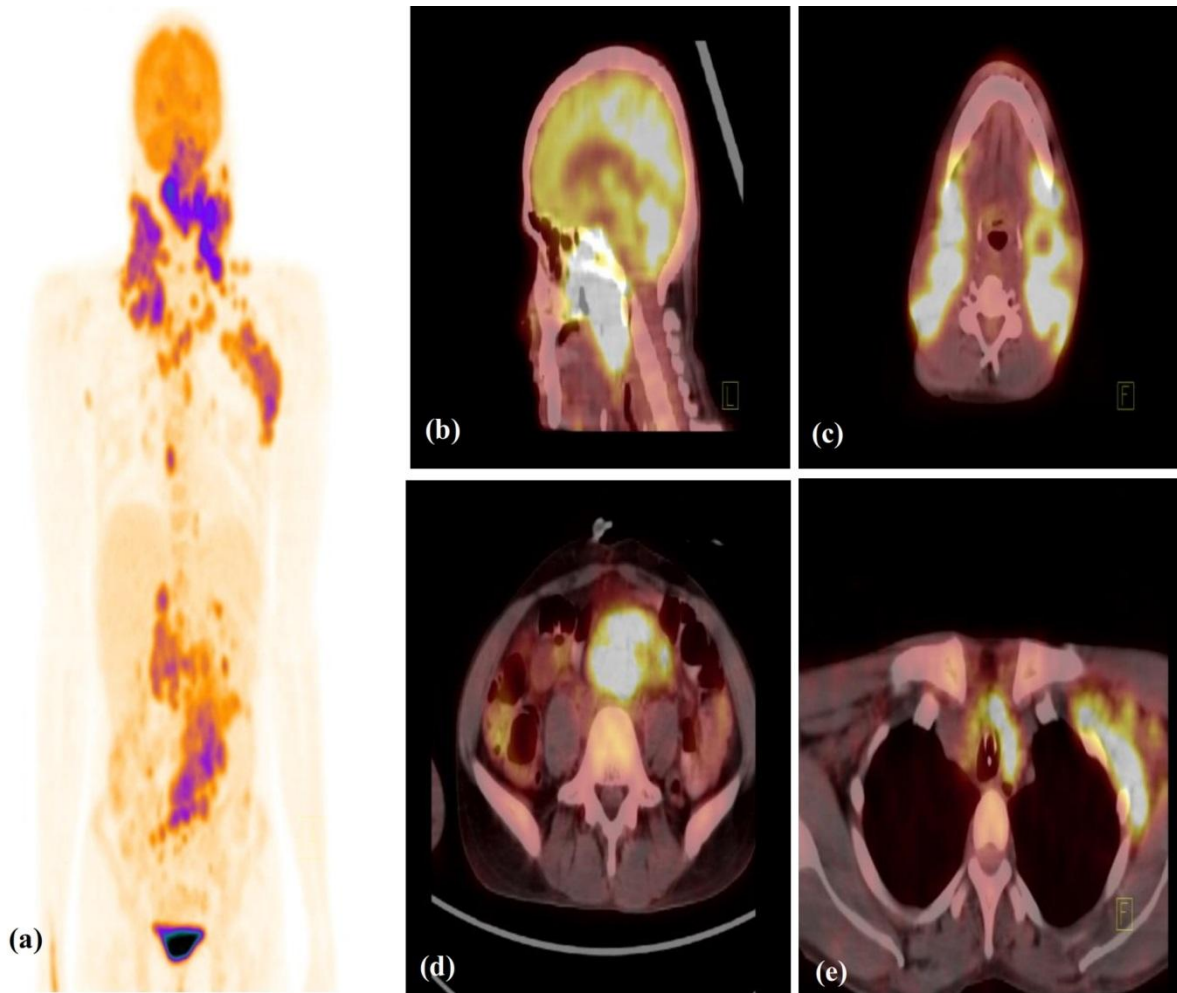


Figure 2 -H&E images A (100X) & B(400X) : Showing sheets of pleomorphic lesional cells with vesicular nucleus and prominent nucleoli showing Non keratinizing undifferentiated nasopharyngeal carcinoma



Patient received 3 cycles of three weekly neoadjuvant chemotherapy regimen of cisplatin and paclitaxel. He later presented with progressive disease with bilateral supraclavicular lymphadenopathy and left axillary lymphnode measuring 2x2 cms. Post chemotherapy PETCT was done which showed Residual Metabolically active lesion in nasopharynx measuring 5.8 x 3.4 x 7.3cms with intracranial extension with maximum SUV of 14.4. Metabolically active bilateral cervical lymphadenopathy, largest measuring 3.0 x 2.6cms with max SUV: 13.3. and axillary region, largest measuring 3.3 x 2.5cms with max SUV of 9.0. Metabolically active Mediastinal, abdominal, pelvic lymphnodes, as shown in figure 3.

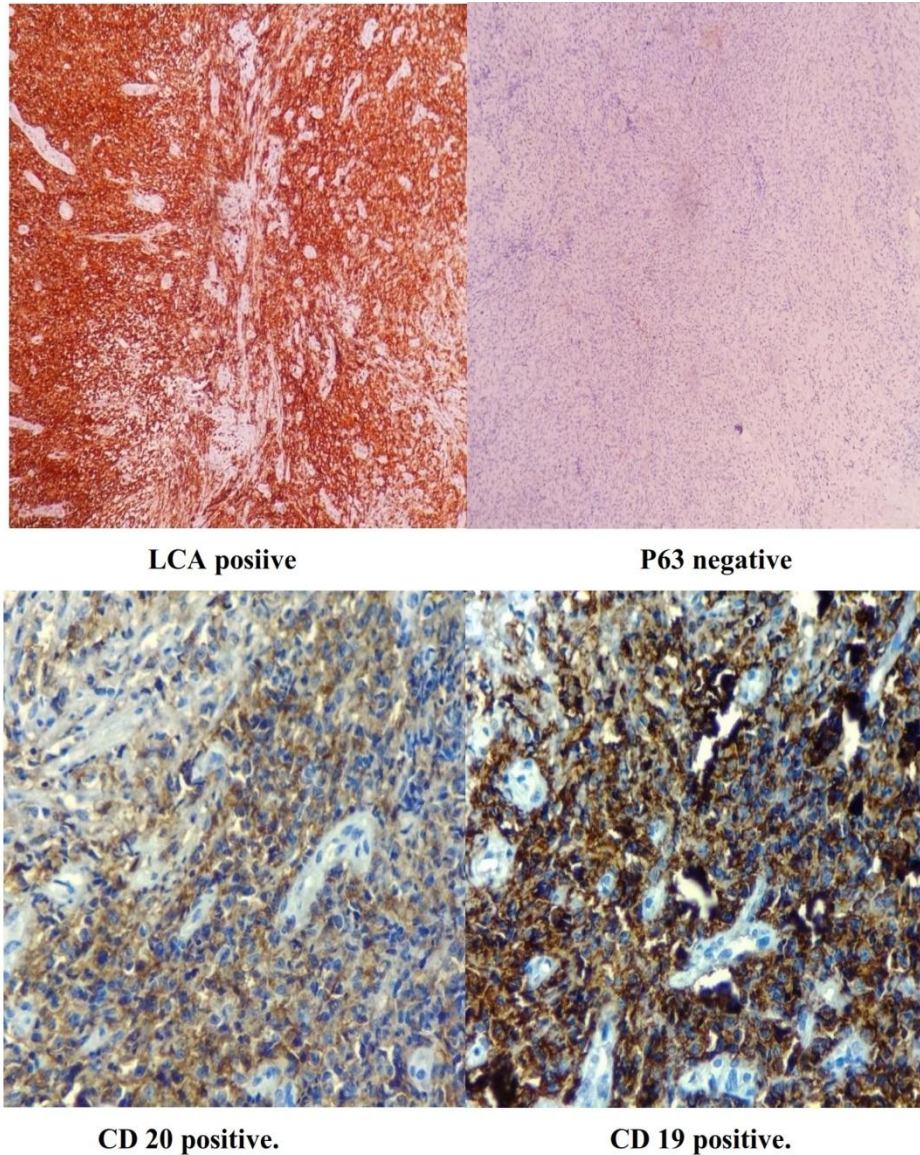
Figure 3-PETCT images- (a)- MIP , (b)-metabolically active lesion in nasopharynx, (c)-uptake in B/L cervical lymphnodes , (d)-uptake in pelvic lymphnodes, (e)- uptake in mediastinal lymphnodes.



IHC was suggested which showed LCA diffuse intense positivity, P63 negative, CD20 positive, CD 19 positive and the diagnosis was reviewed to be nasopharyngeal B-cell lymphoma, as shown in figure 4.

Patient was given palliative RT to neck and abdominal LN to manage localized pain due to tumor infiltration. Patient was treated with a total dose of 30Gy in 10 fractions @ 3Gy/#, further treatment at medical oncology could not be undertaken because of non compliance from patient's side

Figure 4- IHC showing LCA- diffuse intense positivity, P63- negative, CD20- positive and CD 19- positive



Discussion:

The undifferentiated tumors are a heterogenous group of tumors with lack of differentiation. The histological features of tumors of undifferentiated nasopharyngeal carcinoma resemble large cell or immunoblastic types of Non-Hodgkins lymphoma. Symptoms and signs of Nasopharyngeal lymphoma include neck mass, nasal discharge, and nasal obstruction, epistaxis, hearing loss.^[4,5] In areas where there is high incidence of nasopharyngeal carcinoma, Nasopharyngeal lymphoma should always be considered in the differential diagnosis in a patient with a nasopharyngeal mass. Immunohistochemical studies are often used to differentiate nasopharyngeal lymphomas from

carcinoma. It is crucial for the treatment planning and prognosis of the disease as lymphomas have better outcome than nasopharyngeal carcinoma^(8,9,10) Various case reports have demonstrated the possibility of lymphoma with a background of undifferentiated nasopharyngeal malignancy.^(6,7) In this case the patient presented with large neck mass. MRI showed growth in the nasopharynx with cervical lymphadenopathy. Biopsy from nasopharynx was reported as undifferentiated nasopharyngeal carcinoma and patient was started on neoadjuvant chemotherapy for 3 cycles which apparently showed minimum to no response. Post chemotherapy Whole body PETCT was done which showed residual disease in nasopharynx with cervical, mediastinal, abdominal and pelvic

lymphadenopathy . Because of diffuse lymphadenopathy detected on functional imaging, case was suspicious of lymphoma. Immunohistochemical studies were advised to rule out lymphoma. later IHCs revealed B cell lymphoma showing LCA , CD19 and CD 20 positivity and P63 ,CD 5, CD 3 negative. In view of pain, Radiotherapy was offered to neck and abdominal nodes , a dose of 30Gy in 10# was given with Palliative intent. Hence an undifferentiated nasopharyngeal carcinoma should always be further evaluated with IHCs for accurate diagnosis as there is a possibility of it being a lymphoma^(6,7)

Conclusion :

In conclusion Lymphoma being second most common malignancy in nasopharynx, treatment work up using Immunohistochemical studies should be done in case of histomorphological description of undifferentiated carcinoma nasopharynx. This could help in diagnosing the disease in early stage before the commencement of adequate treatment and there by avoiding the possibility of inappropriate treatment.

References

1. Epstein JB, Epstein JD, Le ND, Gorsky M. Characteristics of oral and paraoral malignant lymphoma: a population-based review of 361 cases. *Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology*. 2001 Nov 1;92(5):519-25.
2. Wu R.Y., Li Y.X., Wang W.H., Jin J., Wang S.L., Liu Y.P., Song Y.W., Fang H., Ren H., Liu Q.F., et al. Clinical disparity and favorable prognoses for patients with Waldeyer ring extranodal nasal-type NK/T-cell lymphoma and diffuse large B-cell lymphoma. *Am. J. Clin. Oncol*. 2014;37:41–46. doi: 10.1097/COC.0b013e318261084b.
3. Han A.Y., Kuan E.C., Alonso J.E., Badran K.W., St John M.A. Epidemiology of nasopharyngeal lymphoma in the United States: A population-based analysis of 1119 cases. *Otolaryngol. Head Neck* *Surg*. 2017;156:870–876. doi: 10.1177/0194599817695808.
4. Laskar S., Muckaden M.A., Bahl G., de Sandeep R.N., Gupta S., Bakshi A., Prabhaskar K., Maru D., Gujral S., Parikh P., et al. Primary non-Hodgkin's lymphoma of the nasopharynx: Prognostic factors and outcome of 113 Indian patients. *Leuk. Lymphoma*. 2006;47:2132–2139. doi: 10.1080/10428190600733531.
5. Allam W., Ismaili N., Elmajjaoui S., Elgueddari B.K., Ismaili M., Errihani H. Primary nasopharyngeal non-Hodgkin lymphomas: A retrospective review of 26 Moroccan patients. *BMC Ear Nose Throat Disord*. 2009;9:11. doi: 10.1186/1472-6815-9-11.
6. Gatter KC, Abdulaziz Z, Beverley P, Corvalan JR, Ford C, Lane EB. Use of monoclonal antibodies for the histopathological diagnosis of human malignancy. *J Clin Pathol*. 1982 Nov; 35(11): 1253-67.
7. Gatter KC, Heryet A, Alock C, Mason DY. Clinical importance of analyzing malignant tumours of uncertain origin with immunohistological techniques. *Lancet*. 1985 Jun 8; 1(8441): 1302-5.
8. Zong YS, Zhang RF, He SY, Qiu H. Histopathologic types and incidence of malignant nasopharyngeal tumors in Zhongshan County. *Chin Med J (Engl)*. 1983 Jul;96(7):511 – 6.
9. Ensani F, Karimi SK. Nasopharyngeal Carcinoma: The role of immunohistochemistry in differentiation between undifferentiated carcinoma and malignant lymphoma: Report of 10 cases and review of literature. *Acta Med Iran*. 38 (1); 55-60: 2000.
10. Senba M, Zhong XY, Itakura H. Immunohistochemical investigation of nasopharyngeal carcinoma using keratin, EMA, laminin, fibronectin, collagen type IV, laminin receptor, and laminin/collagen receptor antibodies. *Acta Med Nagasaki*. 1993; 38(2): 182-185.