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CASE REPORT: EXTRANODAL NK/T CELL LYMPHOMA, A RARE ENTITY

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ABSTRACT
Extranodal natural killer/T-cell lymphoma, nasal type (NKTL) is a rare disease with highly aggressive clinical course and poor prognosis. The most common site of presentation is upper aerodigestive tract. The disease may disseminate to various extranasal sites like intestine and other organs but bone marrow infiltration is uncommon. The treatment is largely determined by the extent of disease. Prompt recognition is essential as early treatment is necessary for survival. The efficacy of conventional anthracycline containing regimen is disappointing because of frequent expression of multidrug resistant P- glycoprotein. L-asparaginase containing regimens have significantly improved the efficacy and prognosis for patients with ENKTL. This case report presents a case of Extranodal natural killer/T-cell lymphoma, nasal type, its clinical presentation and diagnostic challenges.

KEYWORDS: Natural killer/T-cell, Anthracycline, L-asparaginase, Lymphoma.

CASE REPORT
A 52 year female presented to us in OPD with right side nasal mass for 6 months. It was gradually progressive and filling the right side nasal cavity. There was no history of mass lesions at other sites, Trauma, Epistaxis, Nasal discharge, Headache and Prior ENT surgery CD5 (T cells, naïve B cells). Except pallor, there was no lymphadenopathy and hepatosplenomegaly. Local examination showed complete right nasal cavity filled with pinkish mass along with localised facial swelling. Routine investigations showed normocytic normochromic anemia. (Fig. 1) CT-PNS was done for soft tissue swelling in the right nasal cavity. There were opacities of both maxillary sinuses with obstruction of both Osteomeatal Complexes. Patient underwent Right Medial Maxillectomy and the specimen was sent for histopathology. (Fig. 2) Dense heterogenous infiltrate of cells composed of atypical medium lymphocytes with prominent vascular invasion associated with large areas of necrosis was seen on biopsy.

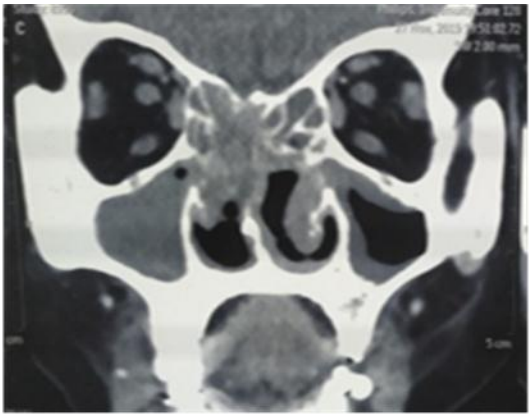

Fig. 1

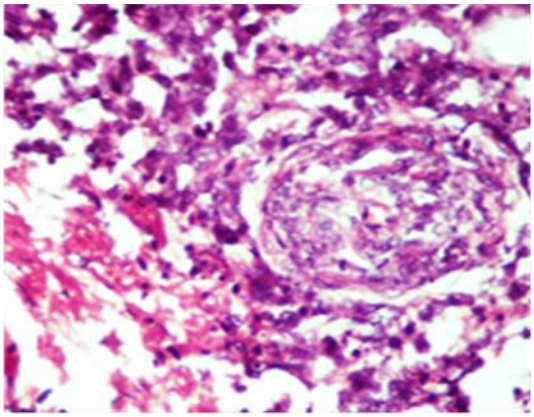

Fig. 2

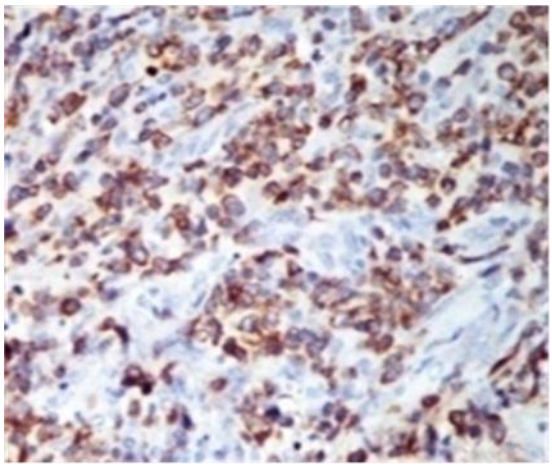

Fig. 3

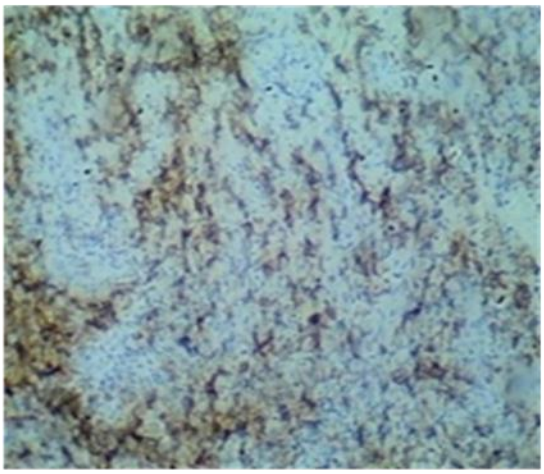

Fig. 4

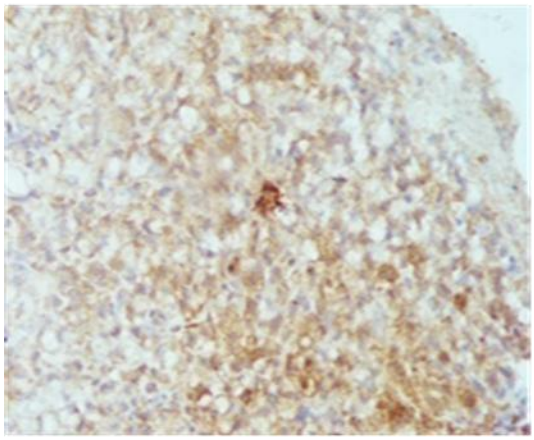

Fig. 5

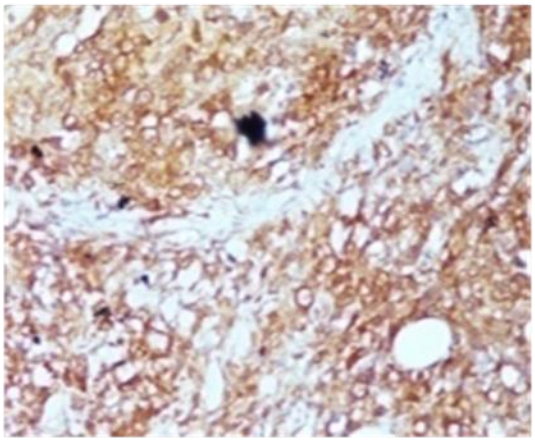

Fig. 6

IHC MARKERS results were as follows:

	RESULT
CD43 (T and NK cells)	**Positive**
CD20 {Mature B cells (except plasma cells)}	Negative
CD3 (T and NK cells)	**Positive**
CD56 (NK and activated T cells)	**Positive**
CD2 (T and NK cells)	**Positive**
CD5 (T cells, naive B cells)	Negative
CD45 (LCA)	**Positive**
CD4 (T helper cells, Tregs)	Negative
CD8 (Cytotoxic T cells)	Focal Positive
CD7 (T and NK cells)	Negative
PanCK	Negative
EMA	Negative
Ki-67	Approximately 40-45%

It was positive for CD2, (Fig .3)CD3, CD43, (Fig .4) CD56 and negative for CD 20. No extranasal lesion was identified on PET/CT. EBER(Fig. 5) and LMP(Fig .6) was positive on biopsy. She got diagnosed as case of NATURAL KILLER/ T-CELL LYMPHOMA NASAL TYPE (LIMITED- STAGE IE) and planned for GELOX regimen followed by radiotherapy and remaining cycles of chemotherapy. The outcome of which was successful.

DISCUSSION

Extranodal natural killer/T-cell lymphoma, nasal type (NKTL) is a rare disease. It is more common in men and the median age at diagnosis is 60. The Incidence in India- 0.7-1.1%.[1] The Site of presentation is the upper aerodigestive tract including nasal cavity, nasopharynx, paranasal sinuses and palate. The patients usually presents with local symptoms such as nasal obstruction, discharge and epistaxis. The disease may disseminate to various sites but bone marrow infiltration is uncommon. In advanced stages there is an extensive destructive mid-facial lesion. Stage I disease is present in 81% of patients while stage II disease in 17% of patients. B symptoms are uncommon. Extranasal sites include intestine (37%), skin (26%), testis (17%), lung (14%), eye or soft tissue (9% each), adrenal gland (6%), brain (6%), and breast and tongue (3%each). On histopathology, it usually consists of a proliferation of a mixture of small and large atypical lymphoid cells. The most characteristic features are prominent vascular invasion associated with fibrinoid necrosis of vessels walls and infarction of surrounding tissues.[2] On immunophenotyping, the cells express CD2, CD56, and cytoplasmic CD3 and are generally negative for CD4, CD8, TCR, and surface CD3. EPSTEIN-BARR VIRUS by in situ hybridization for EBV early RNA(EBER) on biopsy is gold standard investigation. EBV DNA PCR can be done to detect tumor load. The treatment is largely determined by the extent of disease. Prompt recognition is essential as early treatment is necessary for survival. NK/T cell lypmphomas are radiosensitive. This approach achieved a 74% CR rate and 96% overall response rate. The updated analysis after the long-term follow-up reported an 85% 5-year OS rate and 74% 5-year PFS rate.[3] The efficacy of conventional anthracycline containing regimen is disappointing because of frequent expression of multidrug resistant P- glycoprotein. L-asparaginase containing regimens have significantly improved the efficacy and prognosis for patients with ENKTL. NK/T-cell NHL express CD30 in about 40% cases. In such cases, anti CD-30 antibody brentuximab vedotin is shown to be effective after failure of L-asparaginase regimen.[4]

CONCLUSION

Although Extranodal natural killer/T-cell lymphoma, nasal type is a rare entity, the thorough knowledge of clinical presentation, histopathology, IHC markers and radiographic features and its association with extra nasal sites should be well understood. Early recognition and treatment drastically improves the survival rate. Also, important consideration in treatment with L-asparaginase containing regimens is mandatory due to poor efficacy of conventional anthracycline containing regimen.

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