



EXTRANODAL NK/T-CELL LYMPHOMA

ENT

Prithvi*	Resident, Department of ENT, Bangalore medical College and Research Institute, Bangalore *Corresponding Author
Vidya B.T	Assistant Professor, Department of ENT, Bangalore Medical College and Research Institute, Bangalore
Anila Mathew	Resident, Department of ENT, Bangalore medical College and Research Institute, Bangalore

ABSTRACT

Background: Extranodal NK/T-cell lymphoma is rare subtype of non-Hodgkins lymphoma, characterised by a universal association with Epstein-Barr virus. **Case Report:** A 24 year old male presented with history of wound over the nose since 3 months, nasal obstruction since 2 months and facial swelling since 1 month. On examination, patient had widening of root of nose and diffuse swelling of nose, ulcerative lesion with black necrotic tissue and foul smelling discharge. Computed tomography of Nose and Paranasal sinus showed significant polypoidal mucosal thickening in bilateral maxillary sinus, mucosal thickening in the bilateral ethmoid, right frontal, sphenoid sinus and significant soft tissue thickening involving bilateral nasal cavities, soft tissue swelling involving external nose, upper lip, bilateral cheek, left orbit and deossification with bony erosions of bilateral nasal turbinate was noted. Histopathological examination of the biopsy of the lesion showed as nasal NK/T lymphoma. **Discussion-** Extranodal NK/T-cell lymphoma is regarded as a rare subtype of non-Hodgkins lymphoma It often affects adults and is more common in men. Clinical outcomes have improved significantly with application of modern therapies.

KEYWORDS

Extranodal NK/T-cell lymphoma, Epstein-Barr virus,

INTRODUCTION

Lethal midline granuloma is a very rare disease. The term "lethal midline granuloma" was first described by MC Bride in 1897.¹ Also known as angiocentric lymphoma, malignant granuloma, malignant midline reticulosis, polymorphic reticulosis, Stewarts granuloma and NK/T-cell lymphoma. It's pathogenesis is complex and not understood completely. Clinical outcomes have improved significantly with application of modern therapies.²

Case Report

A 24 year old male presented with history of wound over the nose since 3 months and nasal obstruction since 2 months. He also complained of facial swelling since 1 month. Patient had no known comorbidities. On examination, patient had widening of root of nose and diffuse swelling of nose. Ulcerative lesion with black necrotic tissue and foul smelling discharge was present on the nose (left > right) (Fig. 1). On anterior rhinoscopy, necrotic tissue was present in bilateral nasal cavity. Septal defect was noted.



(Fig. 1)

Computed tomography (CT) of Nose and Paranasal sinus (PNS) showed polypoidal mucosal thickening in bilateral maxillary sinus extending through widened osteomeatal complex into middle meatus. It showed mucosal thickening in the bilateral ethmoid, right frontal and sphenoid sinus and significant soft tissue thickening involving bilateral nasal cavities extending to the vestibule. Soft tissue swelling involving external nose, upper lip, bilateral cheek left orbit and deossification with bony erosions of bilateral nasal turbinate was noted.

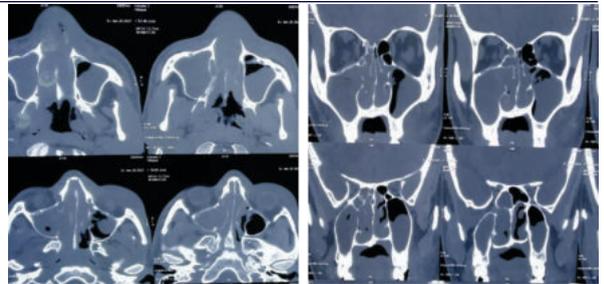


Fig. 2 CT Nose and PNS

A biopsy was taken from the lesion and sent for histopathology - extensive necrosis, mixed cellular infiltrates of atypical lymphocytes, eosinophils, plasma cells and acute inflammatory cells was seen. Case discussed with pathologist and it was diagnosed as nasal NK/T lymphoma.

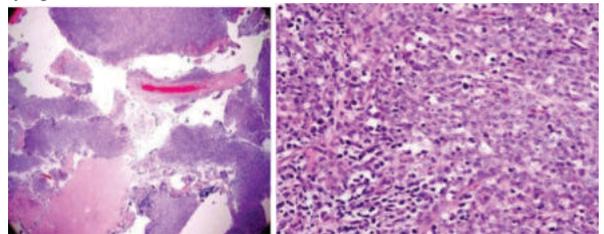


Fig.3: Microphotograph showing extensive necrosis, mixed cellular infiltrates of atypical lymphocytes, eosinophils, plasma cells and acute inflammatory cells.

Treatment

Patient treated with broad spectrum antibiotics, necrotic tissue debridement done. Patient was referred to the oncology department and underwent subsequent chemotherapy and radiotherapy.

DISCUSSION

Nasofacial NK/T cell lymphoma are aggressive locally destructive lesion of the midfacial area. Peripheral NK/T cell lymphomas constitute 10-15% of all non-Hodgkins lymphoma and out of them nasal NK/T cell lymphoma is the most common type.³ This disease occurs around the fourth decade and the male to female ratio is 8:1 to 2:1. It is associated with Epstein Barr virus.⁴ Major symptoms are nasal stuffiness with or without nasal discharge. Oral or nasal ulcers with conjunctivitis may also occur. Grossly the lesion looks like a necrotic

granuloma, which is characterised by ulceration and destruction of the nose and paranasal sinuses with soft tissue, cartilage and bone erosion of the region.⁴ Histopathology usually shows a mixed cellular infiltrates of atypical lymphocytes, plasma cells, eosinophils, histiocytes and acute inflammatory cells.^{2,4} IHC usually indicates the presence of antigen associated with the T- cell such as CD2,CD7,CD45RO,CD43 and CD56(NK cell marker).² Treatment needs combined chemotherapy and radiotherapy. Currently the most recommended treatment include CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) chemotherapy in conjunction with radiotherapy with 5 year survival rates ranging from 20% to80%. New treatment using SMILE protocol shows promising result.

CONCLUSION

Nasofacial location of NK/T cell lymphoma is rarely seen. The confusing clinical profile of this disease often creates suspicion for exact diagnosis. Diagnosis is based on the biopsy and immunohistochemistry. Early diagnosis and intervention prolongs the survival of the patients. Treatment needs combined chemotherapy and radiotherapy and multidisciplinary approach among otorhinolaryngologist, medical oncologist and radiation oncologist for best outcome. This disease carries poor prognosis even it is localized on the nasofacial area.

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