

Nasal T Cell Lymphoma: A Rare Entity

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ABSTRACT

Nasal type of Natural Killer (NK)/T cell lymphoma manifests in the nasal cavity. Approximately 95% of them are associated with EBV (Epstein Barr Virus) with a strong predilection for the Asian population. It has certain systemic and localized symptoms which aid in diagnosis of the condition. However, the histological criteria is pivotal in confirming the diagnosis as well as aiding in confirming the association of EBV. Nasal type of NK/T cell lymphoma has a guarded prognosis. Treatment plan include radiotherapy with concurrent chemotherapy. Despite all this, the 5-year survival rate ranges from 15 - 75%. A 35 years old male presented with an ulcerative nasal lesion. Diagnosed as nasal type of NK/T cell lymphoma via a tissue biopsy, it was managed by chemo-radiotherapy leading to complete resolution of symptoms and disease free on his follow-up 6 months later.

Key Words: *Nasal type of NK/T cell lymphoma. Nasal ulcer. Midface destruction. Radiation therapy. Chemotherapy. CHOP regimen.*

INTRODUCTION

The nasal type of NK/T (natural killer/T) cell lymphoma is a rare but a slowly progressive destructive entity. The term nasal type of NK/T cell lymphoma was first introduced by Williams in 1949. With time, it came to be known by multiple synonyms including Stewart's granuloma, polymorphic reticulosis and other more obscure terms like progressive lethal granulomatous ulceration, malignant granuloma, midline granuloma, non-healing granuloma, and midline malignant reticulosis.¹ Nasal type of NK/T cell lymphoma occurs most commonly in the fourth decade of life, with a female preponderance (2:1 - 8:1).

Described below is the case of a middle aged male presenting with nasal type of NK/T cell lymphoma and subsequent management.

CASE REPORT

A 35 years old male, with no prior co-morbid conditions, presented with a necrotic defect over the left ala of nose and bridge. He had initially developed the lesion 3 months prior that had been un-responsive to multiple antibiotic therapies. He subsequently underwent a biopsy for it, but since he was travelling abroad, he left before receiving the final biopsy report and hence received no treatment for the same. As a result, the necrotic area continued to increase in size and soon involved the dorsum, eroding the septum, with induration

around the malar area and inferior orbital region on the left side (Figure 1).

Upon presentation to our department, he already had a disfigured nose, with almost complete necrosis of the lateral alar wall. A small part of the cartilaginous nasal septum remained which was visible through the necrotic area and blackened. On examination of the oral cavity, the hard palate had blackened mucosa with a fistulous communication in the nasal cavity. Patient denied any history of fever or myalgias.

The patient was admitted, started on intravenous antibiotics and his baseline investigations including complete blood picture, electrolytes, creatinine and blood sugars were carried out, which came out normal. Computed tomography (CT) scan of the para nasal sinuses was performed; it showed the disease to be superficial, not involving the sinuses, and localized to the midface. There was no evidence of bony destruction. A biopsy under local anaesthesia was performed at bedside. It showed dense lymphocytic infiltration with angiocentric pattern of growth, characteristic of nasal type of NK/T cell lymphoma. Further immunochemistry performed was positive for CD56.

The gentleman was then referred to radiation and medicine oncology. He received chemo-radiation therapy, with radiation of 50Gy (2 Grays/fraction) given in 5 weeks and 3 cycles of chemotherapy (CHOP regimen: cyclophosphamide, hydroxy doxorubicin, vincristine, prednisolone). It led to gradual but complete resolution of the disease (Figure 2).

He remained well after completing his treatment and on regular follow ups-of 6 months he was disease free. He had a 3 cm x 3 cm sized defect along the left lateral wall of his nose. He was advised to get treated by a plastic surgeon for the muco-cutaneous defect of his nose for which he opted to receive treatment at local hospital in his city.

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Figure 1: Figure showing necrotic edges of the lesion with loss of the septum.

Figure 2: Small defect caused due to the destructive effect of the lesion, requiring a local flap.

DISCUSSION

Nasal type NK / T cell lymphoma was first recognized more than a century ago, in 1897 by McBride. He reported a case of rapid destruction of face and nose and facial swelling.² The condition gained popularity in 1922, when Stewart described a case series of 10 patients with similar mid-face destructive lesions.¹

Rarely reported in all races, it is more common in Mongoloids, Indonesians and Latin Americans.³ Despite the fact that the etiology is unknown, approximately 95% of the cases are associated with Epstein Barr Virus (EBV).⁴

Diagnosis of nasal type of NK/T cell lymphoma is based on history, clinical examination along with pathological findings. Symptoms may be general or with head and neck manifestations. Constitutional symptoms like fever, malaise, chills, rhinorrhea and weight loss usually precede the midface destruction, none of which were apparent in this patient. Other symptoms include diplopia, proptosis and cranial nerve paralysis due to involvement of local structures. Histopathology shows invasion and destruction of blood vessels along with superimposed bacterial infection. The histological criteria considered in confirming the diagnosis includes: angiocentric infiltration with or without necrosis, extra nodal infiltration into the surrounding tissues, immunophenotype of CD56 or CD3 as well as association with EBV.⁵ T/NK cell lymphoma with an association to EBV is resistant to conventional chemotherapy and has an immensely aggressive clinical course.⁶ EBV test was performed in our patient after his initial diagnosis, which turned out to be negative.

Studies have suggested imaging as non-specific and only beneficial to see the extent of destruction.⁶ It has a series of prodromal symptoms of persistent rhinorrhea, stuffiness and symptoms of sinusitis. In its active phase the nasal discharge becomes purulent with progressive destruction of nasal framework and mid-face. It all ends with gross mutilation as well as systemic metastasis.

On radiological images, nasal T / NK cell lymphoma has to be distinguished from a number of different entities causing sinonasal destruction such as Wegener's granulomatosis, other non-Hodgkin's lymphoma, granu-

omatous infections, adenoid cystic carcinoma, olfactory neuroblastoma, malignant melanoma, and squamous cell carcinoma.⁴ Although, it leads to bony erosion of the palate, alveolar bones and orbit, which are easily picked up on computed tomographic scans, the fact remains that these findings also correlate to a malignant neoplasm. Magnetic resonance imaging is helpful in evaluation of the disease extent but again it is non-specific.⁷

Untreated, nasal type of NK/T cell lymphoma is fatal. Maeda *et al.* have reported that one year survival rate of 50% and 5 years survival rate of 23% for an untreated condition.⁸ Chemotherapy (CHOP regimen) with radiation therapy is the proposed treatment option. Despite radiation and chemotherapy the overall survival rate remains low.⁹

Discrepancies persist in the rates of local control rates using radiation therapy as the modality in different series. The favoured dose of radiation is at least 50 Gy. Studies have shown the 5-year overall survival after radiation therapy to be around 15 - 75%.¹⁰

Nasal type of NK/T cell lymphoma is a slow growing condition, following a waxing and waning course. It follows a downhill course in case of relapse or systemic spread. Good cure and overall survival rates have been observed, once diagnosed early and treated promptly.

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