

Case Report:

Extranodal NK/T-cell Lymphoma, (ENKTL) nasal type: A Diagnostic Enigma

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ABSTRACT

Extranodal NK/T-cell lymphoma, nasal type (ENKTL) is a rare lymphoid neoplasm, with an aggressive, rapid progression. The primary presentation may be non-specific with delay in the early diagnosis and treatment. Our patient; 28 years old gentleman with unilateral nasal obstruction, nasal discharge and swelling over the face with a prior history of SARS-CoV-2 infection had a swiftly deteriorating clinical presentation. Despite repeated multiple biopsies, the extensive tissue necrosis in histopathological specimens made the diagnosis difficult. A high index of suspicion is essential with clinicopathological correlation to make appropriate diagnosis enabling early initiation of treatment.

Keywords:

Extranodal NK/T-cell lymphoma, nasal type (ENKTL), Nasal biopsy, endoscopic sinus surgery

INTRODUCTION

Extranodal NK/T-cell lymphoma, nasal type (ENKTL) is a rare extranodal lymphoma commonly associated with Epstein-Barr virus (EBV) infections involving the nasal area.¹ Patients with ENKTL occasionally presents with involvement of extra-nasal sites such as the skin, gastrointestinal tract, breast, and lungs.¹ Histopathologically, this tumor has been shown to have NK-cell and T-cell immunophenotypes. Clinicians sometimes struggle to diagnose NK/T-cell lymphoma since the initial clinical presentation resembles a variety of other diseases. Our patient a young male too presented with symptoms and signs favoring chronic rhinosinusitis masking the correct diagnosis. Involvement of the regional lymph nodes is unusual until the tumor disseminates.² The confirmation of the diagnosis requires the histopathological report which makes it even more challenging as the need to obtain a representative sample becomes very important. The histological picture is complicated at times with an intense inflammatory reaction and frequently a proliferation of histiocytes induced by the neoplastic cells requiring an expert reporting. The treatment of patients with extranodal NK/T-cell lymphoma, nasal type, is largely determined by the extent of the disease.

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CASE PRESENTATION

A 28-year-old male patient presented to the ENT outpatient department with a left-sided nasal obstruction, nasal discharge for 2 weeks and swelling over the left side of the face for 4 days. He did not give any history of fever, epistaxis, headache or trauma. On examination of the left nasal cavity slough covered mass was seen extending up to the vestibule. He had deviated nasal septum with a spur to the left with tenderness over the left nasal alae and maxillary sinus. The rest of the examination was unremarkable. Initial hemogram, renal function tests and liver function tests showed normal findings. COVID screen was negative with RTPCR. CT scan revealed heterogeneously enhancing mass involving the left nasal cavity, left maxillary sinus with widened osteomeatal complex on the left side.(Fig 1)The differential diagnosis of chronic rhinosinusitis either fungal or granulomatous were considered. Diagnostic nasal endoscopy showed a necrotic mass in the left nasal cavity involving left inferior turbinate with necrosis of left pyriform aperture, painful to touch with serosanguinous fluid found filling the left maxillary sinus. Tissue specimen sent for histopathological evaluation showed mainly necrotic tissues with focal areas of inflammatory infiltrate composed of lymphocytes and few eosinophils. Gram staining was inconclusive and GMS & Periodic acid Schiff (PAS) stains showed no fungal hyphae. The edema over the face progressed to involve the left lower eyelid with the inability to open the eye. However, vision and eye movements were normal. The patient was taken up for endoscopic sinus surgery with complete debridement.

Multiple biopsies from different regions were taken & sent separately for histopathological examination. The patient was started on Amphotericin B in view of the worsening clinical picture. Immediately post-surgery edema over the face started reducing but he developed cervical lymphadenopathy with enlargement of cervical lymph nodes; levels Ib and II. FNAC from cervical lymph nodes was suggestive of reactive hyperplasia. MRI of the nose, paranasal sinuses and orbit revealed peripherally enhancing lesion in the left nasal cavity & left maxillary sinus anteriorly with enlargement and necrosis of left level Ib lymph node. The lymph node biopsy was done; it revealed friable mass with loss of normal architecture enabling only piecemeal dissection. The patients had been treated for COVID infection, 2 weeks ago after which his nasal symptoms persisted. The clinical presentation now compelled us to think about MRSA infection or sinonasal NK T cell lymphoma; which made us liaise with the pathology department to look into the specimens sent, review the slides and especially evaluate further biopsies sent from the nasal tissues. The search concluded with the histopathological and immunostaining suggesting sinonasal NK/T cell lymphoma.

DISCUSSION

ENKTL, nasal type, occurs in the upper aero-digestive tract involving the nasal cavity, nasopharynx, paranasal sinuses, hypopharynx, and larynx.³ The etiology of this tumor is uncertain; however, it is known to have a strong association with EBV infection. The tumor is immunophenotypically mostly active NK cells and some cytotoxic T-cells.⁴ The initial diagnosis has always been a challenge for clinicians typically due to the wide array of symptoms that could be attributed to many other clinical conditions. The most common presentations include nasal obstruction, epistaxis, local swelling with redness.⁵

On imaging, ENKTL nasal type, can be seen as diffuse mucosal thickening along the nasal turbinates or as a destructive midline mass that can mimic a wide variety of diseases like chronic fungal rhinosinusitis, Wegener granulomatosis and invasive fungal infections inflicting a put off on early treatment. Therefore a high index of suspicion must be adopted.⁶ In our case CT scan nose and PNS did not show any evidence of bony involvement or extension beyond the nasal cavity. MRI is useful for visualization of soft tissue alongside the bony involvement, intracranial or orbital spread and for assessment during follow up. However, the only definitive method to diagnose ENKTL is by histological examination and immunohistochemistry.⁵ Tumor cells vary in size from small to large making the diagnosis difficult by mere histopathological examination as the pathologists find it difficult to identify the key features of neoplasia. The cytological analysis primarily based totally on small sampling can have a false report as reactive hyperplasia and is generally marked with large cells and immunoblasts often seen with

atypical forms (common in clinical conditions associated with viral infections)⁷

In our case the patient had a recent SARS COVID infection which may have led to false-negative results. The biopsy specimens sent must include specimens from each site taken separately and labeled appropriately for the histopathological examination. The neoplastic cells are dispersed among plasma cells, lymphocytes, eosinophils, and histiocytes. Invasion of vessel walls by lymphoid cells, known as angiocentricity, is characteristic of nasal type NK/T-cell lymphoma.⁸ To distinguish ENKTL from other neoplasms, infections and benign processes, deep biopsies must be obtained from multiple suspicious areas. Immunostaining along with flow cytometry could be hired for further characterization of such cases as they display the presence of T and NK cell markers (CD2, CD56, CD3), in conjunction with cytotoxic molecules, consisting of granzyme B, TIA-1, and perforins.^{3,9} COVID-19 infection might have played a crucial role in transient remission of NK/T cell lymphoma shown by the reduction of NK neoplastic cells.¹⁰

CONCLUSIONS

The extranodal natural killer/T-cell lymphoma (ENKTL) nasal type commonly manifests with nonspecific upper respiratory tract symptoms, making diagnosis difficult. An aggressive clinical course with little or no response to antibiotics and antifungals should raise the suspicion of such a deadly disease. The gold standard for diagnosis is histopathology report with IHC but owing to extensive tissue necrosis invariably multiple biopsies may be required till the representative sample is obtained and it calls for expert clinicopathological co-relation. The role of the SARS-CoV2 virus infection is yet to be uncovered. We aim to enhance the clinical awareness among the medical fraternity as the above case could be a forerunner of additional similar cases developing in the future.

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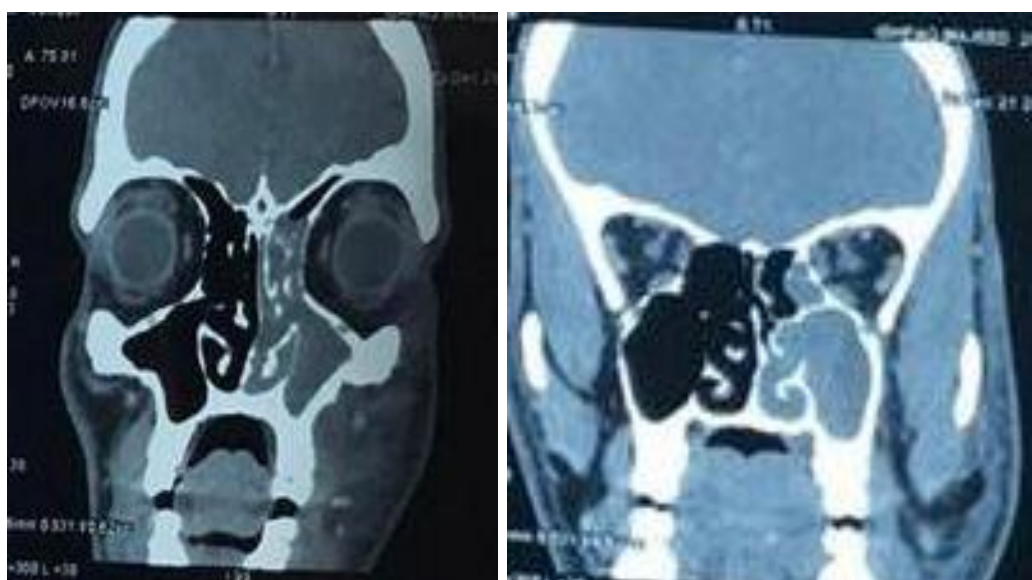


Fig 1: CT Scan Nose and PNS showing heterogeneously enhancing mass involving the left nasal cavity, left maxillary sinus with widened osteomeatal complex on the left side